Kawasaki Disease

ABSTRACT BOOK -

Oral Abstracts

Title: "Recurrent Kawasaki disease or something more? Unraveling the mystery of autoinflammatory disease."

Authors: Ali S,Maheshwari A, Mahto D

Division of Immunology & Rheumatology, Department of Paediatrics, Lady Hardinge Medical College & Hospital

Introduction: Autoinflammatory syndrome is a group of disorder characterized by recurrent episodes of fever rash joint pain and recurrent episodes of inflammation often affecting multiple organs and systems.Also autoinflammatory syndromes are recurrent in nature and are chronic disorder involving dysregulation of innate immune pathway and these disorder are also associated with genetic mutations and can run in families. In contrast, Kawasaki disease (KD) is a self-limiting febrile illness characterized by medium vessel inflammation, predominantly affecting coronary arteries and leading to acquired heart disease in children.

Case Description: A 6 years old female child with a significant past history of recurrent episodes of Kawasaki like Disease from the age of 3 years. Child multiple episodes of respiratory illness for which the child was taking treatment on OPD basis.child also received IvIg in the context of Kawasaki like presentation but her 2D echo was normal and NT pro bnp was normal and child also found to have periapical gingival abcesses. In the light of recurrent episodes of Kawasaki like Disease and ruling out other probable causes genetic testing was done and it revealed SOCS1 mutation consistent with autoinflammatory syndrome.The younger sibling also developed similar complain at the age of 3 year and genetic testing reveals the same mutation. The child was started on colchicine and was doing well without recurrence of symptoms

Discussion and Conclusion: Recurrent Kawasaki disease (KD) episodes have been linked to underlying autoinflammatory syndromes, suggesting a shared pathophysiological mechanism. Studies have identified genetic mutations, such as SOCS1, MEFV, and NLRP3, in patients with recurrent KD, indicating an overlap with autoinflammatory disorders. The recurrent KD phenotype often presents with atypical features, including prolonged fever, skin rashes, and joint involvement, which are characteristic of autoinflammatory syndromes. Furthermore, some patients with autoinflammatory syndromes, such as FMF and CAPS, have developed KD-like episodes. The association between recurrent KD and autoinflammatory syndrome highlights the need for genetic evaluation and consideration of autoinflammatory disorders in patients with recurrent or atypical KD presentations, enabling early diagnosis and targeted therapy to prevent long-term complications.

KAWASAKI DISEASE WITH LEFT ABDUCENS PALSY: A RARE NEUROLOGICAL MANIFESTATION

RASHMI GUPTA1, GAYATHRI C V1, PRABAL BARMAN1, RAKESH KUMAR PILANIA1, SURJIT SINGH1

1. Pediatric Allergy Immunology Unit, Department of Pediatrics, Advanced Pediatric Centre, Postgraduate Institute of medical education and Research, Chandigarh

Introduction: Neurological manifestations reported in KD include extreme irritability, aseptic meningitis, sensorineural hearing loss and rarely cranial nerve palsies. We report an unusual clinical finding: a toddler with complete KD and abducens nerve palsy (ANP).

Case report: A 3-year-old boy came with 9 days of persistent fever and 7 days of inward deviation of the left eye. He had conjunctival injection, a diffuse rash and cracking of lips with red tongue. Physical examination showed sheet-like periungual peeling of skin, left cervical lymphadenopathy and irritability. There was restriction of left eye abduction on attempted left gaze suggesting left lateral rectus palsy. Investigations revealed neutrophilic leukocytosis, thrombocytosis raised ESR and CRP. 2D ECHO revealed normal coronary arteries. Lumbar cerebrospinal fluid(CSF) analysis showed lymphocytic pleocytosis. Fundus examination, Magnetic Resonance Imaging brain and CSF infective workup was normal. Based on the above features, a diagnosis of complete KD with an atypical manifestation of left ANP was proffered. He was treated with IVIG 2g/kg along with aspirin at antiplatelet doses at day 9 of illness. Though he had a defervescence of fever, abducens palsy persisted. Hence, oral prednisolone started with gradual tapering. He had complete resolution of lateral rectus palsy at 6 weeks follow-up.

Results: Neurological manifestations of KD can occur in up to 30% of patients. ANP is a rare manifestation of KD with only 4 cases reported so far. It is postulated to be due to peri neuritis associated with aseptic meningitis seen in KD. Including the index child, 80% of cases with ANP required steroids to reverse the clinical features.

Conclusion: This case highlights that cranial nerve palsy, including ANP, in KD may require treatment intensification with corticosteroid. It also underscores the need to identify the atypical manifestations of KD promptly to offset delays in diagnosis and unnecessary investigations.

CALCIFICATIONS IN CORONARY ARTERY ANEURYSMS: OUR EXPERIENCE WITH CTCORONARY ANGIOGRAPHY (CTCA) OVER THE LAST 10 YEARS

Ahmed Zaid Jamal, Rakesh Kumar Pilania, Munish Arora, Manphool Singhal, Surjit Singh

Allergy Immunology Unit, Advanced Pediatrics Center, PGIMER, Chandigarh
Contact details for correspondence: Dr Ahmed Jamal, Senior Resident, Allergy Immunology Unit, Advanced Pediatrics Center, PGIMER, Chandigarh.
Mob No: 9969551769
Email: ahmed5690.jamal@hotmail.com

Introduction
Coronary artery abnormalities (CAAs) of KD mandate long-term follow-up. CAAs can be complicated by calcification, thromboses, and/or stenoses. Computed Tomography Coronary angiography (CTCA) on present-day CT scanners with radiation optimization capabilities has enabled the comprehensive evaluation of coronary arteries. This study pertains to CTCA performed during follow-up in 12 patients with calcifications in CAAs.

Methods:
This study was carried out in the Allergy Immunology Unit, Advanced Pediatrics Centre, Post Graduate Institute of Medical Education and Research, Chandigarh, India, from 2014 to 2024. Diagnosis of KD was based on standard criteria. CTCA was carried out on dual-source CT scanners: 128-detectors Definition Flash/194-detectors-Force (Siemens, Erlangen, Germany). We have carried out CTCA in 197 KD patients during follow-up. Calcifications were noted in 21 arteries in 12 patients.

Results:
There were 10 boys and 2 girls in this cohort. The mean age at occurrence of KD was 4 years (range 2 months - 8 years). The most common artery involved in calcification was the Left anterior descending artery (12 calcifications), followed by the right coronary artery (7 calcifications). The left main coronary artery was involved in 2 patients. No calcification was seen in the left circumflex artery. Three patients revealed a thrombus in LAD. Two patients had stenosis in LAD, while one patient had stenosis in RCA.

Conclusion:
Children with KD and CAA require prospective long-term follow-up as they may develop complications like calcifications, thromboses, and stenoses. CTCA provides a more detailed and comprehensive evaluation of calcifications. This is the first study of its kind from the Indian subcontinent.

ASSOCIATION OF SINGLE NUCLEOTIDE POLYMORPHISMS OF TGF-b- PATHWAY IN NORTH INDIAN CHILDREN WITH KAWASAKI DISEASE: THE FIRST STUDY FROM THE INDIAN SUBCONTINENT.

BACKGROUND:
KD is the most common vasculitis in children. The phenotype of KD in India appears to be different when compared to reports from the Western hemisphere as well as Japan. Although single nucleotide polymorphisms in the TGF-b pathway have been associated with development of KD, there are no studies from the Indian subcontinent.

OBJECTIVES:
We aimed to study 7 single nucleotide polymorphisms (SNPs) of 4 genes of the TGF-b pathway putatively involved in the pathogenesis of KD: TGFB2: rs2796817; TGFBR2: rs1495592, rs795430; SMAD3: rs12901071, rs1438386, rs6494633; ADAM17: rs670540.

METHODS:
The study was conducted in a tertiary care hospital from July 2022-December 2023. Diagnosis of KD was based on standard criteria. We enrolled 47 patients with KD and 46 healthy controls. Genotypes for different SNPs genes of TGF-b pathway were compared between cases of KD and controls. Similarly, a comparison of the SNPs was made between KD patients with and without CAAs by KASP assay.

RESULTS:
Our results indicated that SNP rs1438386 of the SMAD3 gene exhibited higher odds for both KD (odds ratio 1.71) and for development of CAAs (odds ratio 1.46), suggesting a potential role in disease susceptibility, but not statistically significant. SNP rs2796817 of the TGFB2 gene correlated with the development of CAAs- the GT/GG genotype showing 8 times higher odds for CAAs, compared to the TT genotype. Conversely, SNP rs6494633 of the SMAD3 gene did not demonstrate a significant association with KD or development of CAAs. However, the odds ratio suggested a lower likelihood of KD in individuals with the A allele, while the same allele showed a higher, though statistically insignificant, odds ratio for the development of CAAs. SNP rs1495592 of TGBR2 gene revealed a notable odds ratio of 0.31, implying a 69% lower chance of developing KD in individuals with the CT/TT genotype. However, this polymorphism did not show a significant association with CAAs. SNP rs795430 of TGFBR2 gene did not emerge as a significant risk factor for KD or development of CAAs. SNP rs12901071 of SMAD3 gene also did not exhibit a significant association with the risk for KD or development of CAAs.

CONCLUSION:
Our results suggest that SNPs rs1438386 of SMAD3 gene and SNP rs2796817 of TGFB2 gene may have a role in the pathogenesis of KD and development of CAAs. However, this is a single centre study and results need to be replicated from other centres before definitive conclusions can be drawn.

Title: Low-density neutrophils in Patients with Kawasaki disease-new kid on the block.

Manpreet Dhaliwal, Reva Tyagi, Madhubala Sharma, Kanika Arora, Aditya Dod, Taru Goyal, Rakesh Kumar Pilania, Saniya Sharma, Surjit Singh

Allergy Immunology Unit, Advanced Pediatrics Centre, Postgraduate Institute of Medical Education and Research

Introduction

KD is a medium vessel vasculitis that is the leading cause of acquired heart disease in children the world over. Although the etiopathogenesis of this condition is still poorly understood, innate immunity appears to play an important role. A subset of neutrophils, viz. low-density neutrophils (LDNs) have been associated with inflammatory disorders such as lupus and rheumatoid arthritis. Their role in the pathogenesis of KD, however, has not been studied. In this study, we have attempted to characterize the role of LDNs, measure their oxidative capacity, and correlate disease activity with the proportions of LDNs.

Methodology

The objective of this study was immunophenotyping and functional assessment of LDNs in patients with KD, febrile controls (FC), and healthy controls (HC). We studied the association between disease activity and the proportion of LDNs. LDNs were analyzed on BD LSR Fortessa X 20 5 Laser Flow Cytometer using monoclonal antibodies (mAbs) CD3-PerCP-Cy5.5, CD14-BV421, CD15-APC, CD33-PE (BD Pharmingen, USA). Statistical analysis was performed on SPSS (ver. 29) using the Kruskal-Wallis test, Wilk-Shapiro test and Unpaired T-test.

Results

Median (IQR) percentage of LDNs (%gated) in KD patients was elevated when compared to HC but the difference was not statistically significant (p=0.07). Similarly, there was no statistical difference in %gated LDNs between KD patients and FC (p=0.18). Statistical significance was obtained when %gated LDN in patients, FC, and HC were compared (p=0.05) (Fig-1). We found that the proportion of LDNs was elevated in patients with conjunctival injection compared to those without conjunctival injection (p=0.2). We also performed oxidative reduction assay in KD patients, FC, and HC. Comparison of delta MFI (mean fluorescence index) in KD patients with FC and HC showed a p-value of 0.85 and that of SI (stain index) in patients with FC and HC showed a p-value of 0.34.

Conclusion

Our study demonstrates that the proportion of LDNs in patients with KD is elevated compared to healthy controls, suggesting a potential association between LDNs and the disease. Additionally, the oxidative capacity assays suggest that further functional studies could provide deeper insights into the role of LDNs in KD pathogenesis.

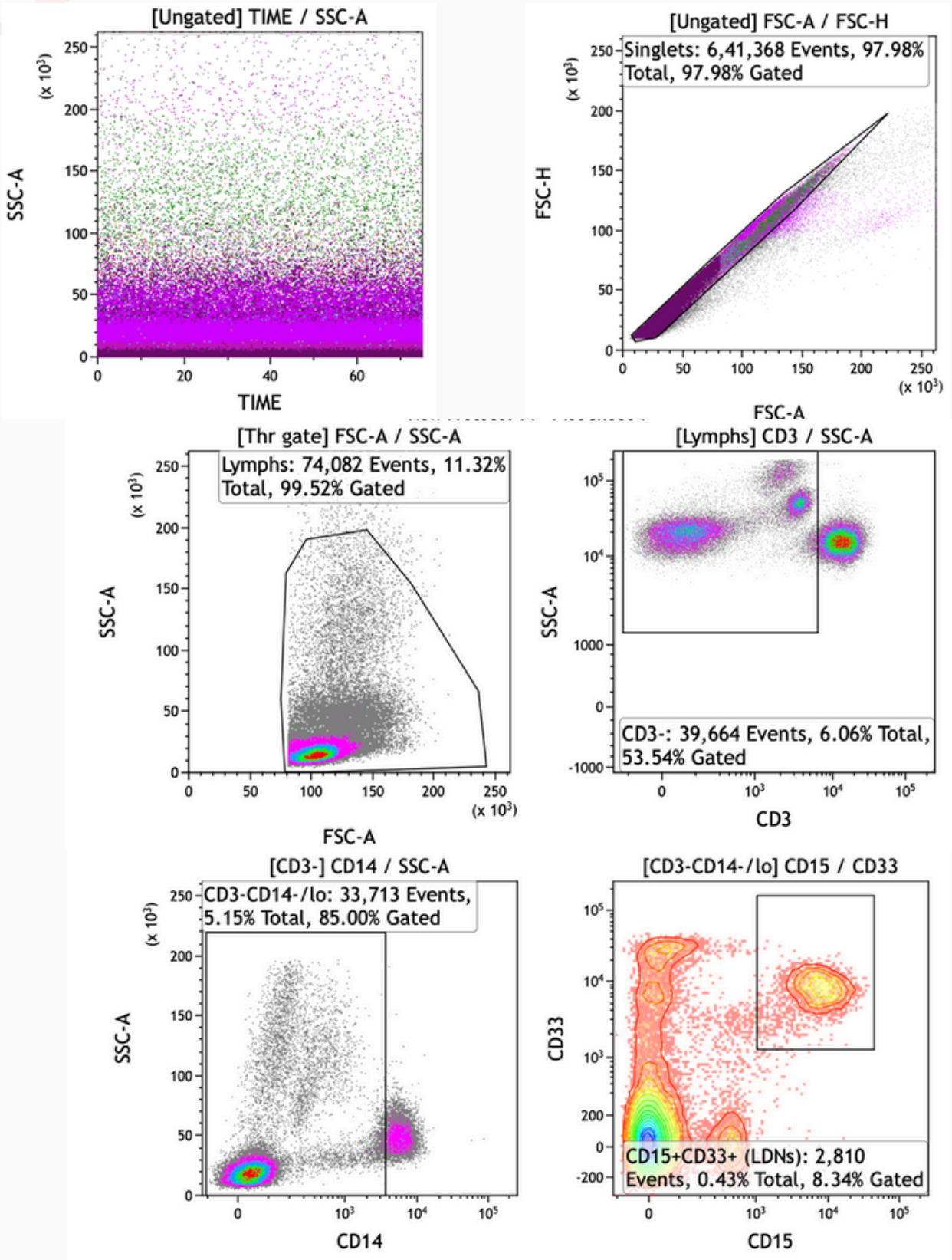


Figure 2): Characterisation of LDNs. Representative image of surface staining of LDNs in patient 4 a) Quality control during sample acquisition using time parameter b) FSC-A and FSC-H plot gate singlets c) Lymphocytes gated on FSC-A and SSC-A d) Gating of CD3- negative cells e) The CD3 negative cells further gated using CD14 marker to exclude monocytes f)The CD3 and CD14 negative population was further studied using the CD 33 and CD 15 markers and were characterised as LDNs

Title: Neutrophil Subsets and Extracellular Trap Formation in Kawasaki Disease-Insights into the Development of Coronary Artery Aneurysms.

Manpreet Dhaliwal*, Ayush Srivastava*, Rakesh Kumar Pilania*, Vinod Sharma#, Mohinder Kaur#, Arunima Sharma*, Saniya Sharma*, Amit Rawat*, Surjit Singh*

*Advance Paediatrics Centre, Postgraduate Institute of Medical Education and Research
#Department of Dermatology, Postgraduate Institute of Medical Education and Research

Introduction
Kawasaki disease (KD) is an acute systemic vasculitis of unknown etiology, primarily affecting children and leading to significant complications such as coronary artery aneurysms (CAA) and myocarditis. Neutrophils, key components of the innate immune system, are among the first cells to infiltrate the vascular endothelium during the acute phase of KD. Neutrophils produce Neutrophil Extracellular Traps (NETs) which may play a role in the pathogenesis of KD, especially in forming CAA. In this study, we aimed to investigate the formation of NETs and the neutrophil subsets in patients with acute KD and incomplete KD (ICKD) and compare the findings to those of healthy controls.

Methodology - The study enrolled 23 KD patients (aged 4.31 ± 2.97 years) and 10 healthy controls. Blood samples were collected, and polymorphonuclear leukocytes (PMNs) were isolated using Lymphoprep™ (Stemcell®). NETs formation was evaluated in both PMA-stimulated and unstimulated neutrophils using ImageJ software. Flow cytometry was performed to analyze neutrophil subsets using antibodies against CD3-APC, CD62L-PE, CD11b-FITC, and CD184-PEcy7.

Results - We found a higher percentage of male children in the KD group, with a mean platelet count of 4.14 ± 1.03 lac/µL and a total leukocyte count (TLC) of 14,000 ± 3,000/µL, significantly higher than in healthy controls. NETs formation in both stimulated and unstimulated neutrophils was significantly higher in KD/ICKD patients compared to controls ($p = 0.027$ for PMA stimulation, $p = 0.0423$ for unstimulated). Specifically, unstimulated KD/ICKD neutrophils showed a NETs formation rate of 25-26%, with normal coronary arteries (z-score <2) in most cases. Notably, two KD patients with aneurysms in the left anterior descending (LAD) artery (z-score ⩾2.5) showed a significantly higher NETs formation (33.53 ± 17.3%). Flow cytometry revealed an increased proportion of mature neutrophils (CD184/CXCR4+/CD62L+) in KD patients, although this difference was not statistically significant ($p = 0.939$).

Conclusion - This study provides evidence of increased NETs formation in both stimulated and unstimulated neutrophils in KD/ICKD patients. This suggests a potential role for NETs in the pathogenesis of KD, particularly in the development of coronary artery aneurysms. The observed association between elevated NETs and aneurysms in the LAD artery warrants further investigation to explore the therapeutic potential of targeting NETs in KD.

Title: LOW DOSE DUAL-SOURCE CT CORONARY ANGIOGRAPHY AND CALCIUM SCORING AT 10 YEARS OF FOLLOW-UP IN CHILDREN WITH KAWASAKI DISEASE AND CORONARY ARTERY ABNORMALITIES AT PRESENTATION

Authors: Bipin Dhakal, Rakesh Kumar Pilania, Manphool Singhal, Surjit Singh

Background: Kawasaki disease (KD) is an acute vasculitis that predominantly affects children and can lead to significant coronary artery complications. It is crucial to understand long term status of coronary artery abnormalities (CAAs) in children with KD.

Objective:
The study aims to evaluate the long-term outcomes of CAAs in children with KD with CAAs at presentation, using 2D Echocardiography (2D ECHO) and low-dose dual-source CT coronary angiography (CTCA) at 10-year follow-up.

Methods:
All children diagnosed as KD with CAAs at presentation were enrolled on follow-up and underwent CTCA and 2D echocardiography (ECHO) on the same day to assess coronary artery diameters, aneurysms, and the presence of thrombosis, stenosis, or calcification. Statistical analysis was conducted using SPSS v26 software. Intraclass correlation coefficient and Bland-Altman analysis were used to evaluate the agreement between CTCA and 2D ECHO findings.

Results:
The study included 32 patients with a mean follow-up duration of 12.56 years. Of 32 patients, coronary artery dilatation (2Z to <2.5Z) was noted in 11 patients, small aneurysms (2.5Z to <5Z) in 6, medium aneurysms (5Z to <10Z) in 9 and giant aneurysms (^{3}10Z or absolute diameter ^{3} 8mm) in 6 patients. Follow-up 2D ECHO revealed persistence of CAAs in 6 patients, of which dilatation of the coronary artery was found in 1 patient, small aneurysms in 3, and medium-sized aneurysms in 2 patients. There were 57 baseline CAAs present at the time of diagnosis. During follow-up, CTCA revealed 15 aneurysms as compared to 12 that was visualised in 2D ECHO - 3 aneurysms [2 in left anterior descending coronary artery (LAD) and 1 in left circumflex coronary artery (LCx)] were missed in 2D ECHO. None of the patients had giant aneurysms on follow-up.

Regression rates were 88.23% for small aneurysms, 66.66% for medium aneurysms, and 44.4% for large aneurysms. The regression rates of left main coronary artery (LMCA), LAD, right coronary artery (RCA) and LCx were 89.2%, 66.6 %, 57.14% and 50% respectively. Calcification was observed in three patients, confined to the aneurysmal segments, that were detected by CTCA only. The intraclass correlation coefficients for LMCA, LAD, and RCA were 0.636, 0.648, and 0.776, respectively, indicating good agreement between CTCA and 2D ECHO. However, CTCA demonstrated superior visualization capabilities, particularly for the LCx and distal segments of the coronary arteries, and identified 3 more CAAs and calcifications that were not visible on 2D ECHO.

Conclusion:

CTCA is a valuable non-invasive imaging modality for the long-term assessment of coronary artery abnormalities in KD patients. It provides superior visualization of coronary arteries, including distal segments, and can detect calcifications and stenosis that may be missed by 2D ECHO. The complementary use of both imaging techniques enhances diagnostic accuracy and patient management. Further validation through multicenter cohort studies is recommended to generalize these findings.

Keywords:

Kawasaki Disease, Coronary Artery Abnormalities, CT Coronary Angiography, Calcium Scoring, Long-Term Follow-Up, 2D Echocardiography

Title: Long-Term Coronary Artery Outcomes in Infants with Kawasaki Disease: A 7-Year Follow-Up Using 2D Echocardiography and CT Coronary Angiography

Authors: Pragati Bagh, Rakesh Kumar Pilania, Manphool Singhal, Surjit Singh
Affiliation: Pediatric Allergy Immunology Unit, Department of Pediatrics, Advanced Pediatric Centre, Postgraduate Institute of medical education and Research, Chandigarh

Background: Kawasaki disease (KD) is an acute vasculitis that predominantly affects children and can lead to significant coronary artery complications. Long-term follow-up of these patients is crucial for understanding the progression and regression of CAAs, as well as for the detection of coronary artery calcification and stenosis.
Objective: The study aims to evaluate the status of the coronary arteries in infants diagnosed with KD irrespective of the presence of CALs, using 2D echocardiography (2DE) and low-dose dual-source CT coronary angiography (CTCA) at a 5 year follow-up period.

Methods: This observational study, conducted at an Advanced Pediatric Centre in North India from Jan 2023 to June 2024, enrolled patients were diagnosed with KD in infantile period. Infants with KD were enrolled, irrespective of CALs. Participants underwent CTCA and 2DE on the same day to assess coronary artery diameters, aneurysms, and the presence of thrombosis, stenosis, or calcification. Statistical analysis was conducted using SPSS v26 software. Intraclass correlation coefficient and Bland-Altman analysis were used to evaluate the agreement between CTCA and 2DE findings.

Results: The study included 33 infants with KD with a mean follow-up duration of 7.06±2.20 years. The results indicated that 86.8% CALs showed regression over the follow-up period. Regression rates were 100% for small aneurysms, 55.1% for medium aneurysms, and 33.3% for large aneurysms. Regression seen in LMCA (95%), LAD (85.7%), RCA (84.2%), and LCA (100%), respectively. None of patents showed coronary artery calcification. Intraclass correlation coefficients for LMCA, LAD, and RCA were 0.801, 0.739, and 0.895, respectively, indicating good agreement between CTCA and 2DE. However, CTCA exhibited superior visualization capabilities, of LCx and distal segment of coronary arteries, and identified aneurysms and calcifications that were not detectable on 2DE.

Conclusions: After more than 7 years of follow-up, complete resolution of CALs was observed in 89.5% of infants with KD, whereas 10.5% had persistent CALs, mainly medium and large aneurysms. Most small aneurysms resolved over time. One patient experienced aneurysm enlargement, highlighting the need for long-term follow-up. CTCA proved to be a critical tool in identifying coronary abnormalities and enhancing diagnostic precision when paired with 2DE. Therefore, integrating CTCA in the long-term management of KD patients, particularly those with CAAs, is essential for optimizing care.

Keywords: Kawasaki Disease, Infants, Coronary Artery Lesions, CT Coronary Angiography, Calcium Scoring, Long-Term Follow-Up, 2D Echocardiography

Title: New Horizons in IVIG -resistant Kawasaki Disease: The promise of Anakinra

Authors: Dr. Kumar Abhinav 1, Dr. Suma Balan2, Dr. Sabin George 1
Affiliations:
1.Senior Resident, Department of Clinical immunology and Rheumatology, Amrita Institute of Medical Sciences, Kochi
2.Professor and HOD, Department of Clinical immunology and Rheumatology, Amrita Institute of Medical Sciences, Kochi

Introduction: Kawasaki Disease (KD) is a medium- to small-vessel vasculitis primarily affecting children under five, diagnosed based on clinical criteria. Serious complications, such as coronary artery aneurysms and ectasia, occur in about 25% of patients and can be fatal. Intravenous immunoglobulin (IVIG) and aspirin are the first-line treatments, but 10-20% of patients show resistance to IVIG, increasing the risk of giant aneurysms and fatal sequelae. Interleukin-1α (IL-1α) plays a crucial role in the development of KD and coronary artery aneurysms. Anakinra, an IL-1 receptor antagonist, inhibits both IL-1α and IL-1β. Real-world evidence of anakinra's effectiveness in IVIG-resistant KD stays scarce.

Case Report:
We present three clinically confirmed KD cases referred to our hospital after receiving IVIG and steroids. One patient was referred in the first week of illness, while the other two were referred in the third week and had also received infliximab. All three patients presented with high-grade fever, rashes, significantly elevated inflammatory markers, and worsening coronary dilatations. One patient developed macrophage activation syndrome, another had a giant aneurysm, and the third had a large aneurysm. Due to persistent inflammation, anakinra was started as a second-line agent in all three cases Two patients showed marked improvement in clinical symptoms, aneurysm size, and inflammatory markers. However, one patient experienced re-emergence of aneurysms after stopping anakinra, necessitating its restart, which again resulted in a reduction in aneurysm size.

Conclusion:
Our experience highlights that IL-1 blockade can significantly mitigate systemic inflammation in KD. Despite the small sample size of three cases, our findings emphasise the crucial role of IL-1 in KD-related coronary artery disease. With the literature review and the KAWAKINRA study, only 38 patients have received anakinra to date, with reported clinical benefit. These results suggest that anakinra is a promising, cost-effective choice for IVIG-resistant KD when used early in the disease course.
.

CLINICAL CASE ABSTRACT

Atypical Kawasaki Disease and Its Complications: A Case Report

Arun Soni, Avinash Sharma, Sandesh Guleria
Dr Rajendra Prasad Government Medical College Kangra at Tanda

Background: Kawasaki disease (KD) is a common childhood vasculitis. KDSS (Kawasaki disease shock syndrome) is characterized by hemodynamic instability. A rare association of KD with appendicitis is known. Atypical KD is difficult to diagnose and has remained a challenge for physicians.

Objective: To report a case of KD with shock syndrome presenting as acute appendicitis, associated with pulmonary tuberculosis and complicated with giant coronary artery aneurysms (CAAs).

CaseAn 8-year-old previously healthy boy presented with fever, irritability, nausea, and vomiting for 3 days. He was admitted to a hospital elsewhere and started on IV antibiotics. He then developed acute abdominal pain, localized to the periumbilical region, along with shock, so referred here. On examination, He was lethargic. He had tachycardia, tachypnea, hypotension, cracked lips, a strawberry tongue, bilateral non purulent conjunctivitis, and unilateral cervical lymphadenopathy. Abdominal examination revealed marked tenderness with rigidity and guarding in right iliac fossa.

Laboratory investigations showed, elevated white blood cell count (21,110/µL) with neutrophil predominance (78.8%), high C-reactive protein (163.33 mg/dL), prolonged INR (2.38), PT (26.7), and APTT (35s), with an ESR of 68 mm/h and low platelet count (105,000/µL). His renal function tests indicated severe impairment (BUN 127, serum creatinine 2.17, eGFR 25 mL/min/1.73 m^2). He had elevated serum ferritin (415.38), NT-pro BNP (>35,000), LDH (655).

Possibility of KD with KDSS with ? acute appendicitis was considered and he was kept nil per oral, nasogastric tube was inserted. After fluid resuscitation for shock, IV adrenaline infusion was initiated. IV piperacillin+tazobactam and intravenous immunoglobulins (IVIG) @ 2 gm/kg was given over next 16 hours. Ultrasound abdomen revealed acute appendicitis and he underwent appendicectomy. Histopathological examination of appendix also showed features of acute appendicitis.

On day 6, he continued to have fever with elevated inflammatory markers. His sepsis work up was negative. His 2-D echocardiogram revealed aneurysms in right coronary artery (RCA) and left main coronary artery (LMCA) (6 mm and 7 mm, with Z-scores of 8.9 and 10.9, respectively) and CT coronary angiography showed giant CAAs in RCA, LMCA and left anterior descending artery (LAD). In view of persistent fever, possibility of IVIG-resistant KD was considered and was given IV infliximab (7 mg/kg). His fever subsided but had cough with persistent inhomogeneous opacities on chest X-ray. Work up for tuberculosis revealed sputum CBNAAT positive for

tuberculosis, leading to the initiation of anti-TB treatment. He was discharged on oral aspirin (3 mg/kg/day), injection enoxaparin (1 mg/kg/day, 12 hourly) and ATT. On follow up, he is doing fine.

Conclusion This atypical case of KD, presenting with KDSS and a rare manifestations as acute appendicitis, may be an impedance in early diagnosis and management. Furthermore, the presentation of pulmonary tuberculosis raises the question of it being an infectious trigger for KD. Further studies on this aspect may help in early diagnosis and prevention of morbidities.

CLINICAL PROFILE OF COMPLETE AND INCOMPLETE KAWASAKI DISEASE WITH CORONARY ARTERY ANEURYSM AT DIAGNOSIS : A RETROSPECTIVE OBSERVATIONAL STUDY AT A TERTIARY CARE CENTER IN EASTERN INDIA

Introduction: Kawasaki disease (KD), an acute systemic vasculitis of childhood is an important cause of acquired heart disease in children. Diagnosis of complete KD (cKD) is based on clinical signs and symptoms, whereas diagnosis of incomplete KD (iKD) is challenging.

Objectives: To study the clinical spectrum of cKD and iKD patients with coronary artery aneurysm (CAA) at diagnosis and to compare the parameters between the two groups.

Material and methods: A retrospective observational study of KD patients, admitted at Institute of Child Health, Kolkata from January 2020 to July 2024. Children with congenital heart disease and those who received treatment for KD prior to admission, were excluded from the study. Diagnosis was based on American Heart Association 2017 guidelines. Coronary artery lesions were diagnosed by echocardiography using Z score classification.

Results: Among 130 KD patients included, 64 (49.23%) were cKD and 66 (50.76%) were iKD. Number of patients with CAA at diagnosis was 49/130 (37.69%). Number of cKD with CAA was 25/64 (39.06%). Number of iKD with CAA was 24/66 (36.36%). Median age at diagnosis of cKD and iKD was 24 omonths (range12-36 months) and 10 months (range 4.75-16.25 months) respectively (p= 0.0021). Median duration of fever at diagnosis of cKD and iKD was 8 days and 7 days respectively. Median value of clinical parameters noted in cKD and iKD group respectively: C-reactive protein (CRP) (mg/L) 106 and 51.2, platelet (/cmm) 4,61,000 and 4,46,000, hemoglobin (g/dL) 8.9 and 9.35, Total leucocyte count (TLC) (/cmm) 20,300 and 17,875, serum albumin (g/dL) 3 and 3.35. Hemoglobin and serum albumin were significantly lower in cKD group (p=0.04 and p=0.0019 respectively). CRP was significantly high in cKD group (P=0.0090). There was no significant difference in duration of fever at diagnosis, TLC, platelet counts between the two groups.

Conclusion: High number of CAA (37.69%) was noted in KD patients at diagnosis. Median age at diagnosis of iKD was significantly lower than that of cKD (p=0.0021). . Number of cKD patients with aneurysm at diagnosis was comparable to number of iKD patients with aneurysm at diagnosis. Hemoglobin and serum albumin were significantly lower whereas CRP was significantly higher in cKD as compared to iKD.Thus occurrence iKD was more in infants, with lower CRP at diagnosis but with similar occurence of CAA as cKD.

Clinical Profile of Kawasaki Disease- Experience from a Tertiary Care Center in South India

1Jyothi Janardhanan, 2Bharath A P, 1Sagar Bhattad

Affiliation:
1Division of Pediatric Immunology and Rheumatology, Department of Pediatrics, Aster CMI Hospital, Bangalore, India.
2Division of Pediatric Cardiology, Department of Cardiology, Aster CMI Hospital, Bangalore, India

Contact information of presenting and submitting author:
jyothi428@hotmail.com
Phone no. +91 9096437793

Introduction:
Kawasaki disease (KD) is an acute multisystem inflammatory disease involving medium-sized blood vessels with a predilection to coronaries, that most commonly affects infants and young children.

Objective:
To study the clinical profile of Kawasaki Disease in patients at a tertiary care center in Bangalore, India.

Methods:
A retrospective review of clinical records was performed and patients with KD, diagnosed during the study period (Feb 2017 to October 2024) were included. Clinical and laboratory profiles and clinical outcomes were reviewed. Factors contributing to intravenous immunoglobulin (IVIg) refractoriness and coronary artery abnormalities (CAA) development were assessed. A p-value <0.05 was considered as significant.

Results:
During the study period, 109 children with KD presented to the center. The mean age at presentation was 42.77 ± 29.73 months. Infantile Kawasaki disease was seen in 29 patients (27 %). Males were predominantly affected (n=73, 67 %). The mean duration of fever was 9.66 ± 4.47 days. Delayed diagnosis (>10 days) was seen in 35 patients (32%). Fever was noted in all patients (n=109,100%). Eye changes (n=74, 68 %) were the most commonly associated clinical findings, followed by rashes (n=67, 62 %), mucosal involvement (n =59,54 %), extremity changes (n= 37, 34%), and lymphadenopathy (n= 27, 25 %). Incomplete KD was seen in 47 patients (43%). Overall, 45 patients (42 %) developed CAA. Systemic artery aneurysms were noted in two infants. 97 % of patients received intravenous immunoglobulin (IVIG) along with aspirin as the first line of treatment, while 42 % needed treatment intensification. Steroids were used in 45 patients (42%) as primary intensification or as a part of refractory KD therapy.

COMPARISION OF CLINICAL AND LABORATORY PARAMETERS BETWEEN SINGLE CORONARY ARTERY ANEURYSM AND MULTIPLE CORONARY ARTERY ANEURYSMS AT DIAGNOSIS OF KAWASAKI DISEASE : A SINGLE-CENTER RETROSPECTIVE OBSERVATIONAL STUDY FROM EASTERN INDIA

Introduction: Kawasaki Disease (KD), an acute systemic vasculitis of children is diagnosed clinically supported by laboratory criteria. Most dreaded complication of KD is coronary artery lesions (CAL) in the form of dilatation or aneurysm. Predicting the risk of developing coronary artery aneurysm (CAA) in KD remains challenging.

Objectives: To compare the clinical and laboratory parameters between patients with single CAA and patients with multiple CAA at diagnosis of KD.

Methods: A retrospective observational study on KD patients with CAL at diagnosis at Institute of Child Health, Kolkata from January 2020 to July 2024. Diagnosis was based on American Heart Association 2017 guidelines. Children with congenital heart disease and those who received treatment for KD prior to admission, were excluded from the study. Coronary artery lesions were diagnosed by echocardiography using Z score classification.

Results: Among 130 KD patients included in the study, 82 were male and 48 were female. CAL at diagnosis was detected in 51 (39.23%) patients, among them 49 (96.07%) patients had CAA at diagnosis. Hence overall occurrence of CAA was 37.69% (49/130) of all KD patients at diagnosis. Among the 49 patients who had CAA at diagnosis, 36 (73.46%) were male and 13 (26.53%) were female (p= 0.0001); 23 (46.93%) patients had single aneurysm and 26 (53.06%) patients had multiple aneurysms at diagnosis. Parameters noted in single CAA group: male (n=17), female (n=6), complete KD (n=11), incomplete KD (n=12). Parameters noted in multiple CAA group: male (n=19), female (n=7), complete KD (n=14), incomplete KD (n=12). Median values noted in single CAA group and multiple CAA group respectively were: age 14 months (range 8-30 months) in both, duration of fever before diagnosis 7 days (range 6-10.25 days) and 8 days (range 7-10 days), platelet 4,4,2000/mm3 and 5,20,000/mm3, hemoglobin 9.3 g/dL and 8.95 g/dL, TLC 18,200/mm3 and 19,000/mm3, serum albumin 3.3 g/dL in both group, CRP 84.9 mg/L (range 33.95-228) and 89.5 mg/L (range 19.02-117).

Conclusions : CAA was significantly more in males. No statistically significant diferrence in parameters were found between the single CAA and multiple CAA groups. There was no significant diferrence in ocurrence of single and multiple aneurysms in complete and incomplete KD.

Title: Kawasaki disease in infant, is it different?

Authors: Ritu Turaiha1, Sandesh Guleria2, Avinash Sharma3
Author Affiliation-
1Junior resident, department of pediatrics, Dr. RPGMC Tanda at Kangra, HP.
2Assistant Professor, Cell of pediatric immunology and Rheumatology, Dr. RPGMC Tanda, at Kangra, HP.
3Associate Professor, Cell of pediatric immunology and Rheumatology, Dr. RPGMC Tanda, at Kangra, HP.

Background: Kawasaki disease commonly involving children age group 1 to 5 year, can present in a child from birth to 18. It is common cause of acquired heart disorder in young children.

Methods: A retrospective comparative study of Kawasaki disease was performed in a tertiary care center in sub-Himalayan region of India, between infants and young children. All kids presenting to the center diagnosed with Kawasaki disease were included in the study, 43 cases were enrolled from April 2017 to October 2024, Of which 4(9.3%) cases were infants. Children less than 1 year age were categorized as group1 while those above 1, group 2.

Result: All infants were found to have prolong fever, mean duration of fever is 9.5 days. Irritability observed in all (100%), pleomorphic rash and desquamation of skin observed in 50% children of group 1 (92.30% in group 2). Puffiness of hands and feet was seen in 2(50%) cases (84.61% group 2). Strawberry tongue was spotted in 75% group1 cases (87.17% in group 2). All group 1 children found to have thrombocytosis with platelet count >4lac (38.46% in group 2). Cervical lymphadenopathy observed in 25% children in group 1 (74.35% in group 2), non-purulent conjunctivitis was seen in all infants (84.61% cases in young). BCG scar rea-activation was examined in 50% infants, while none of group 2child showed scar re-activation. Coronary artery abnormality evaluated in 75% group1 children, while it is observed in 33.33% cases in group 2. 75% infants in group1 were diagnosed with incomplete Kawasaki disease and 25% with typical KD. All received IVIG at diagnosis and aspirin for 6 weeks.

Discussion: Kawasaki disease in infantile period have more incomplete presentation compared to young. Infants with Kawasaki disease found to have more coronary artery abnormalities than other children, which is comparable to a study in a tertiary care center from Bengaluru, India. Incomplete presentation does not necessitates reduced severity of disease, Kawasaki disease in infants is associated with prolong duration of illness at presentation and more severe course. There is higher incidence of coronary artery abnormalities compared to other children.

Conclusion: There is a significant difference in clinical presentation and disease sequalae in infants compared to young children in our study.

Title: Retrospective Analysis of Outcomes of Giant aneurysms and risk factors for the development in Kawasaki Disease: A Tertiary Centre Experience in Kerala of 15 cases of giant aneurysms.

Kumar Abhinav1, Suma Balan2, Manyam Prudhvi Krishna1, Manu Pradeep3 , Mahesh Kappanayil 4, Shine Kumar4, Balu Vaidyanathan4, Navaneetha Sasikumar5, Krishna Kumar 4
1. Senior resident, Department of clinical Immunology and Rheumatology, AIMS, KOCHI
2. Prof and HOD, Department of clinical Immunology and Rheumatology, AIMS, KOCHI
3. Research fellow, Department of clinical Immunology and Rheumatology, AIMS, KOCHI
4. Prof, Department of Pediatric Cardiology AIMS, KOCHI
5. Associate Professor, Department of Paediatric Cardiology, AIMS, KOCHI

Abstract
Background: Kawasaki disease (KD) is an acute vasculitis primarily affecting children under the age of five, with coronary artery complications being a significant concern. Approximately 25% of untreated KD cases develop coronary artery aneurysms (CAA), with giant aneurysms (>8 mm diameter) leading to a higher risk of thrombosis, myocardial infarction, and ischemia. Risk stratification for these patients is essential to guide treatment and predict long-term outcomes.
Objectives: This study aims to describe the clinical profile, risk factors, interventions, and outcomes among patients with KD in a tertiary care centre who were referred and diagnosed here as giant coronary artery aneurysms.

Methods: This retrospective case series, conducted at AIMS between 2019 and 2024, included KD patients with giant coronary aneurysms. Clinical data, treatment modalities, and outcomes were obtained from electronic health records and analysed. Paediatric cardiologist diagnosed giant aneurysms via echocardiography based on a z-score >10 or an inner coronary diameter >8 mm. Statistical analysis included the use of SPSS 22 and Epi-info 7.2.1, with continuous data represented as mean, standard deviation, and median.

Results: Between 2019 and 2024, 54 children were diagnosed with Kawasaki disease (KD), with 28 patients referred after initial treatment elsewhere and 26 presenting here with the first manifestation. During their clinical course, 15 out of 54 patients (27.8%) were diagnosed with giant aneurysms, 93.3% of whom had giant aneurysms at the time of admission. Among these, 40% were referred due to sequelae, and 60% had ongoing active vasculitis. Most of the patients (53.3%) were under one year old, and 73.3% were male. The most common symptoms were fever (100%), rashes (93.3%), and conjunctival congestion (80.0%), along with persistently elevated inflammatory markers and proBNP levels. Incomplete KD was diagnosed in 29% of cases, and atypical KD in 15%. The diagnosis of KD was made at a median of 7 days (range 4-14 days); however, treatment intensification was delayed in this subset, with a median delay of 21.5 days (range 20.25-26.75 days). All patients were resistant to initial IVIG treatment, and despite intensified therapy with steroids, infliximab, cyclosporine, and anakinra after referral,

only 13.3% of aneurysms fully resolved. Aneurysms remained stable in 59.6% of cases, while complications included myocardial infarction in 13.4%, coronary artery bypass grafting in 6.7%, and death in 6.7%. 50% of patients remain on long-term anticoagulation.

Conclusion: Giant aneurysms in KD are associated with significant morbidity, particularly in infants. Despite aggressive treatment, complete resolution remains challenging, underscoring the need for prevention through early diagnosis, awareness of risk factors for IVIG resistance, and the necessity for primary treatment intensification in high-risk cases

Keywords: Kawasaki disease, giant coronary artery aneurysm, intravenous immunoglobulin, thrombosis, paediatric vasculitis, coronary artery bypass

USE OF INFLIXIMAB IN INFANTS WITH KAWASAKI DISEASE

BACKGROUND:
About 10-20% of KD patients who do not respond to IVIG, have higher risk of developing CAA and will require further management. Infliximab (IFX) has been recommended for treatment intensification in patients with IVIG resistance as well as those presenting with aneurysms.

OBJECTIVES:
1.Evaluate the response to IFX by fever defervescence ,normalization of CRP in IVIG resistant KD.
2. Evaluate the response to IFX by regression in size of aneurysms, for those with CAA at diagnosis.

METHODOLOGY:
Observational study at Institute of Child Health, Kolkata, India from January 2016 to December 2023. Infants with KD, treated with IFX for IVIG resistance, presence of coronary artery aneurysms at diagnosis or increasing size of CAA following IVIG therapy were enrolled. IFX was administered and clinical characteristics and outcomes were analyzed.

RESULTS:
260 children were diagnosed with KD, 51 were infants,32 received IFX. Median duration of illness at presentation was 7.5days. The median duration for IFX administration was 13 days from the onset of fever.

14/ 32 in this cohort received IFX for IVIG resistance, 2/14 also had CAA at presentation. Following IFX administration 12 became afebrile within 24 hours , 2 within 48hours.

All small and 9 medium aneurysms completely regressed. Among the 9 giant aneurysms,3 completely regressed, 4 reduced to small aneurysms ,1 continues to have persistent giant aneurysm. One infant with giant aneurysm and 1 with medium aneurysm have been lost to follow up. Median time for regression was 3 months.

Pulmonary presentation of Kawasaki disease complicated with Macrophage activation syndrome: diagnostic and treatment challenges in a 10-year-old boy

Juri Kaman1 , Prabal Barman1
1Department of Pediatrics, Gauhati Medical College, Guwahati

INTRODUCTION

Kawasaki disease (KD) is a common vasculitis in children with a predilection for coronary arteries. Atypical manifestations such as lung involvement are distinctly unusual. Macrophage activation syndrome (MAS) has been reported in < 2% of patients with KD. Herein, we report a child with KD who had lung symptoms, and whose clinical course was complicated by MAS.

OBJECTIVE

To report a case of Atypical Kawasaki disease with Pulmonary presentation in a 10-year-old boy, complicated by MAS.

METHODS AND RESULTS

A 10-year-old boy presented with fever for 10 days, difficulty in breathing for 3 days and rash for 1 day. On examination, he had tachypnea and bilateral crepitations on auscultation. Investigations revealed neutrophilic leucocytosis, raised ESR and C-reactive protein (CRP). Chest X-ray showed patchy consolidation of right lower lobe. He was commenced on intravenous antibiotics; however, within next 48 hours he developed worsening sensorium, hypotension and low pulse volume. He also started developing periungual peeling of fingers and toes. Repeat investigations showed thrombocytopenia, raised CRP, increased triglyceride and ferritin, and low fibrinogen level. Two-dimensional echocardiography was suggestive of dilated left anterior descending artery with ejection fraction: 20%. Considering the constellation of clincal symptoms and investigations, a diagnosis of Atypical Kawasaki disease with MAS was considered. He was initiated on intravenous immunoglobulin(2g/kg) and methylprednisolone (30 mg/kg/day for 3 days, followed by oral taper), and aspirin (3 mg/kg/day). He responded briskly and is doing well on follow-up.

CONCLUSIONS

Pulmonary manifestations and MAS in KD are often underrecognized in clinical practice leading to delays in diagnosis and treatment initiation. Clinicians should be cognizant of these subtle clinical clues to promptly identify these patients and initiate therapy to offset morbidity and mortality.

Title: The CGD and Kawasaki Crossover

Authors: Dixit N, Maheshwari A, Mahto D
Division of Immunology & Rheumatology, Department of Paediatrics, Lady Hardinge Medical College & Hospital

Introduction: Chronic granulomatous disease (CGD) is an inherited immune disorder caused by mutations in genes encoding components of the NADPH oxidase complex, leading to a deficiency in reactive oxygen species (ROS) production. This deficiency impairs phagocyte function and results in recurrent infections and autoimmune-like hyper-inflammatory responses. Kawasaki disease (KD), a form of acute vasculitis primarily affecting children, has been observed in association with primary immunodeficiencies, including CGD.

Case Description: A 3-year-old girl, born to consanguineous parents, presented with a history of recurrent respiratory issues. She first developed pneumonia at 3 months, treated successfully with oral antibiotics. At 11 months of age, she was hospitalised with prolonged fever and cough, treated with ceftriaxone and azithromycin. Persistent symptoms led to extensive investigations, revealing multifocal lung consolidation with multiple nodules in bilateral lung fields and lymph node enlargement on CECT Chest, though tuberculosis was ruled out. She was later diagnosed with chronic granulomatous disease (CGD) after showing reduced neutrophil oxidative index. Following another severe pneumonia episode at 14 months, she received prolonged IV antibiotics.

At age three, she returned with fever and respiratory symptoms. Bloodwork showed severe anemia and elevated inflammatory markers. Despite broad-spectrum antibiotics, fever persisted, and 2D Echo suggested Kawasaki disease (KD)-like illness. Echocardiography revealed coronary artery dilation, treated with intravenous immunoglobulin (IVIG) and prednisolone. After clinical improvement, she was discharged on aspirin and prophylactic antibiotics.

Discussion and Conclusion: This case describes a 3-year-old girl diagnosed with chronic granulomatous disease (CGD) at age one, confirmed by a positive neutrophil oxidase test and genetic analysis showing a homozygous NCF1 mutation, indicating autosomal recessive inheritance. The child had recurrent pneumonia, managed with antibiotic prophylaxis. At age three, she presented with persistent fever and respiratory symptoms. Despite broad-spectrum antibiotics, child had persistent fever and further work up showed Kawasaki disease (KD)-like features, including coronary artery dilation and pericardial effusion. Treatment with IVIG and steroids improved her symptoms. Among similar cases, KD-like illness has been previously reported in 4 such cases of CGD, predominantly in X-linked cases. However, recurrent pneumonia more common in autosomal recessive CGD.

Title: CLINICO-EPIDEMIOLOGICAL PROFILE OF KAWASAKI DISEASE FROM A TERTIARY CARE CENTER IN SUB-HIMALAYAN REGION OF INDIA.

Authors: Ritu Turaiha1, Sandesh Guleria2, Avinash Sharma3
Author Affiliation- 1Junior resident, department of pediatrics, Dr. RPGMC Tanda at Kangra, HP. 2Assistant professor, cell of pediatric immunology and rheumatology, Dr. RPGMC Tanda at Kangra, HP. 3Associate professor, cell of pediatric immunology and rheumatology, Dr. RPGMC Tanda at Kangra, HP.

Background: Kawasaki disease is a medium vessel vasculitis affecting pediatric age group, having variable presentation among population of different geographical region.
Methods: An observational cross-sectional study was performed in our hospital from April 2017 to October 2024, to know clinic-epidemiological profile of disease.

Results: Total 43 cases were seen during this period. Of these, 30 (69.8%) were males. 30 patients (69.76%) were between 1 and 5 year, 9 (20.9%) more than 5 year and 4 (9.3%) were less than one year of age. Fever was present in all, desquamation (perianal and periungual) in 40(93.02%), rash in 38 (88.37%), puffiness of hands and feet observed in 35 (81.39%)children. Unilateral cervical lymphadenopathy recognized in 21 (48.83%), bilateral cervical lymphadenopathy in 6 (13.95%), generalized lymphadenopathy in 1 (2.3%) case. Strawberry tongue spotted in 36 (83.72%), irritability in 26 (60.4%) and conjunctivitis in 38 (86.0%) cases. BCG scar reactivation examined in 2 children. ESR more than 20 in 30 (69.76%) and positive CRP >6 marked in 22 (51.16%)kids. All enrolled children were anemic, mild - 23 (53.48%), moderate- 19 (44.18%), severe -1 (2.3%). Thrombocytosis identified in 19 (44.18%), while low platelet counts in 1(2.3%) child. Mean platelet count is 455 x 103/mm3. 2D-ECHO for coronary artery abnormality, was performed in all, showed hyperechoic wall in 1(2.3%), coronary dilatation in 4(9.3%), mild aneurysm in 10(23.25%), moderate aneurysm in 1(2.3%) and no CAA in 26(60.04%). Follow-up ECHO in all children were normal. Coronary CT-Angiography was performed in 7 children, all were normal. There were 21 (48.83%) typical, 20 (46.51%) incomplete and 3 (6.97%) atypical Kawasaki disease. One child (2.3%) diagnosed with co-existing tuberculosis, one(2.3%) had pre-existing malignancy and one (2.3%) found with UTI. All cases of Kawasaki diseases received IVIG at diagnosis and aspirin for 6 weeks. One child received infliximab.

Discussion: In our study maximum clinical finding other than fever was desquamation (peri-anal and periungual) of skin, followed by rash, non-purulent conjunctivitis and mucosal changes, which is comparable to a study in a tertiary care center in Bengaluru in India. Anemia was found in all children, maximum coronary abnormalities were found in infants which is also comparable to the above mentioned study.

Conclusion: Incidence of Kawasaki disease is more in males, mean age at diagnosis was 3.82 year, Mean duration of fever 8.7 days. The disease may co-exist with illness such as TB, UTI, malignancy, seizure disorder and meningitis as observed in our study.

Low dose vs medium dose aspirin in acute phase of Kawasaki disease- an experience from a tertiary care hospital of West Bengal, India

Sarbik Sarkar1, Deblina Patra1, Bodhisatya Das1, Sayani Pan2, Kamirul Islam2, Taraknath Ghosh3, Mithun Chandra Konar4, Archan Sil5
1 Post graduate Trainee, Department of Pediatrics, Burdwan Medical College and Hospital, Bardhaman, West Bengal
2 Senior Resident, Department of Pediatrics, Burdwan Medical College and Hospital, Bardhaman, West Bengal
3Professor, Department of Pediatrics, Burdwan Medical College and Hospital, Bardhaman, West Bengal
4 Associate Professor, Department of Pediatrics, Burdwan Medical College and Hospital, Bardhaman, West Bengal
5Assistant Professor and In charge Pediatric Clinical Immunology and Rheumatology Clinic, Department of Pediatrics, Burdwan Medical College and Hospital, Bardhaman, West Bengal

Low dose vs medium dose aspirin in acute phase of Kawasaki disease- an experience from a tertiary care hospital of West Bengal, India

Background- Kawasaki disease (KD) is a common childhood vasculitis. There are controversies regarding the dosage of aspirin in the acute phase of KD, despite years of extensive research. The aim of this study was to evaluate the efficacy of intravenous immunoglobulin (IVIG) plus low-dose aspirin (3-5 mg/kg/d) in comparison to IVIG plus medium-dose aspirin (30-50 mg/kg/d) in the treatment of acute KD.

Methods- Data were collated by reviewing clinical records of patients registered between August 2022 and August 2024 in the Pediatric Rheumatology clinic of a tertiary care centre of eastern India. AHA (American Heart Association) 2017 criteria were used for diagnosis of KD. All patients initially received a single dose of IVIG (2g/kg) in the acute phase. Patients were classified into two groups. Group 1 included 26 cases treated with IVIG plus 3-5 mg/kg/day aspirin (low-dose group), and group 2 included 30 cases treated with IVIG plus 30-50 mg/kg/day aspirin (medium-dose group). Clinical (fever) and laboratory response (inflammatory parameters) were compared between the two groups. The primary outcome measure was the occurrence of coronary artery lesions (CALs) at the subacute or convalescent stage. $P < 0.05$ was considered as significant.

Results- Fifty-six patients who fulfilled the criteria of KD were included in this study. While 30 patients with KD were found to be treated by low-dose aspirin, 26 of them received medium-dose aspirin in the acute phase. There was no significant difference among the two groups (low dose vs. medium dose) in terms of the anti-inflammatory effect revealed by white blood cell count, percentage of neutrophils in white blood cells, platelet count, and C-reactive protein level. CALs were found in 11% (3/26) of the low dose group and 13% (4/30)

of the medium dose group ($\chi 2$ = 0.04, P= 0.84). Those who had CALs were subsequently treated as per the institutional protocol. Incidence of adverse effects, particularly gastritis was
lesser in children who received low-dose aspirin (3.8% vs 23.3%, P=0.04).

Conclusion- We found no significant benefit of using medium dose aspirin in acute phase of KD but there were higher incidences of complications. Further prospective multicentric studies incorporating a large number of patients are needed to confirm this finding.

Title: Epidemiological shifts in Kawasaki disease incidence during and after the covid-19 pandemic: observations from Chandigarh, north India, during the period 2020-2023

Authors: Yamini Sharma, Suprit Basu, Rakesh Kumar Pilania, Saniya Sharma, Manpreet Dhaliwal, Ankur Jindal, Pandiarajan Vignesh, Deepti Suri, Amit Rawat, Surjit Singh
Affiliation: Pediatric Allergy Immunology Unit, Department of Pediatrics, Advanced Pediatric Centre, Postgraduate Institute of medical education and Research, Chandigarh

Background: Global studies have reported that the incidence of Kawasaki disease (KD) declined during the COVID-19 pandemic. These studies suggest that the global pandemic and its accompanying mitigation measures may have been responsible for this reduction. The present study aims to estimate the incidence of KD during the pandemic period 2020-2023 at Chandigarh, North India.

Methodology: All children suffering from KD residing within the Union Territory [UT] of Chandigarh and diagnosed between January 2020 and December 2023 were enrolled. Annual incidence rates were calculated using decadal growth rates based on National Census Data, 2011. The methodology was similar to previously published studies from our centre pertaining to the periods 1994-2008 and 2009-2014. We computed the incidence of KD in children aged <5 as well as in children aged <15 years.

Results: During the period 2020-20223, 22 patients (17 boys, 5 girls) were identified to have KD in UT Chandigarh. Annual incidence rates during these 4 years were 2.65, 2.61, 6.1 and 6/100,000 in children below 5 and 0.7, 1.24, 1.74, and 1.65/100,000 in children below 15. There was a drastic reduction in incidence of disease from the pre-pandemic period (9.72/100,000 children below 5 in 2019). Median age at diagnosis was 36 months (range: 4-108 months). Maximum cases was seen in March (n=8) followed by May (n=3), whereas there was nadir seen in July. Coronary artery abnormalities (CAAs) during the acute phase of illness were noted in 4 (18.2%) patients.

Conclusions: This study showed that the number and incidence of KD was dramatically reduced during the COVID-19 pandemic in Chandigarh, India like other parts of the world. This change may be due to decreased incidence of other infectious agents other than COVID-19, supporting the hypothesis of infection-triggered pathogenesis in KD.

Title: Jejunal Perforation in Convalescent Phase of Kawasaki Disease – An unusual presentation

Authors: Yamini Sharma, Gayathri CV, Rakesh Kumar Pilania, Surjit Singh
Affiliation: Pediatric Allergy Immunology Unit, Department of Pediatrics, Advanced Pediatric Centre, Postgraduate Institute of medical education and Research, Chandigarh

Background: Acute abdomen-like presentation is seen in 2-5% of patients with Kawasaki disease (KD) during acute phase. Cholecystitis, intestinal obstruction, acute appendicitis, acute pancreatitis, acute peritonitis, intestinal volvulus, duodenal, ileal perforation, and intussusception have been reported so far in the acute phase. We report a child with an acute abdomen in the convalescent phase.

Case report: A 4-year-old girl was brought with vomiting, loose stools, abdominal pain, and high-grade fever for ten days. She also had redness of oral mucosa, conjunctival injection, perianal erythema, and purpuric rashes over trunk extremities. Diagnosis of complete KD was considered. 2D-echocardigraphy showed left main coronary artery (LMCA) dilatation (2.9mm; +2.8 Z) and left anterior descending coronary artery (LAD) small aneurysm (2.8mm; +3.5 Z), respectively. Laboratory investigations revealed anemia (75 g/L), thrombocytopenia (75x103/L), hyponatremia (131 mEq/L), hypoalbuminemia (2.4 g/L), and elevated C-reactive protein (189mg/L). She was managed with intravenous immunoglobulin (IVIG) 2g/kg, oral prednisolone (2mg/kg), and aspirin at an antiplatelet dose (3 mg/kg/day). She became afebrile, her rashes disappeared, and her abdominal symptoms improved. Oral prednisolone was tapered and stopped over 6 weeks. During follow-up 5 months later, she presented with acute abdominal distension and bilious vomiting. Abdominal X-ray showed gas under the right diaphragm suggestive of intestinal perforation. Laparotomy showed jejunal perforation 200 cm distal to duodeno-jejunal flexure. She underwent resection of an unhealthy small bowel and end-to-end anastomosis. Physical examination revealed the presence of Beau's lines over all toes. CT coronary angiography showed dilated proximal LMCA 3.2mm (+2.2Z) with no other aneurysms, and abdominal angiography was normal. She was then discharged on aspirin. The child is doing well in the follow-up.

Discussion: Intestinal perforation in KD is rare; Spontaneous duodenal perforation and ileal perforation have been reported in the acute phase of KD. To the best of our knowledge, this is the first case of spontaneous intestinal perforation in the convalescent phase of KD. It might suggest ongoing endothelial dysfunction /vessel wall involvement even in the convalescent phase. This report highlights that those who present with abdominal symptoms in the acute phase require close monitoring and long-term follow-up as life-threatening complications can occur even in the convalescent phase.

A Real-Time PCR Based Gene Expression profiling study of Kawasaki disease in North Indian children: Implication for targeted therapy

Authors: Abarna Thangaraj1, Rakesh Kumar Pilania1, Vibhu Joshi1, Kanika Arora1, Amit Kumar1, Taranpreet Kaur1, Priyanka Srivastava1, Amit Rawat1, Surjit Singh1
1 Pediatric Allergy and Immunology Unit, Advanced Pediatrics Centre, PGIMER, Chandigarh
Submitting author: Dr Abarna Thangaraj
Email: dr.abarna.t@gmail.com
Phone number: +91-9443880637
Presenting author: Dr Abarna Thangaraj
Email: dr.abarna.t@gmail.com
Phone number: +91-9443880637

ABSTRACT
Background: Kawasaki disease is a common childhood vasculitis and the pathogenesis of KD is still an enigma.

Objectives:
To study the expression of 15 genes (IL1B, S100A12, MMP9, ITGAM, TLR2, PRF1, CD44, TLR8, TREM1, UBB, IL7R, FCER1G, CXCL8, SPI1, and FCGR1A) in North Indian patients with KD by real-time- polymerase chain reaction (RT-PCR) method. To compare gene expressions in children with KD versus healthy controls (HC) and disease (SLE) control (DC). To elucidate the impact of treatment on gene expression in children with KD and the correlation with the presence of coronary artery abnormalities (CAAs) in children with KD.

Design/Methods:
Study was conducted from July 2022 - December 2023. We enrolled 34 patients with KD (samples were taken before and after treatment with IVIG), 9 healthy controls, and 4 disease controls (systemic lupus erythematosus). We used the TaqMann assay for RT-PCR in the Step-one plus system. Relative gene expression was calculated as 2-ΔΔCT (Livak method).
Results

Twelve genes were up regulated (IL1B, ITGAM, TLR2, CXCL8, SPI1, S100A12, MMP9, PRF1, TLR8, CD44, FCER1G, and FCGR1A), while 3 genes (UBB, IL7R, and TREM1) were down regulated. IL1B and MMP9 showed 6-fold increase in expression, and S100A12 showed 4-fold increase in expression (Figure 1, Table 1). IL1B showed statistically significant upregulation compared to controls (P=0.049). SPI-1 (p=0.0425) and TREM1 (p=0.0180) showed a significant downregulation compared to healthy controls. On comparing the pre- vs post-IVIG group in KD, genes that were downregulated during the pre-IVIg phase have shown increased expression post-IVIg [PRF1 (p=0.0313), UBB (p=< 0.0001), IL7R (p=0.0022) and TREM1 (p=0.0187)]. CD44 (p=0.0400) and TLR2 (p=0.0184) expression was significantly low in KD patients compared with the disease controls. IL1B expression is increased in the CAA group compared

to controls (p=0.025) The relative gene expression of CD44 (p=0.023), ITGAM (p=0.026), TLR2 (p=0.008) and FCGR1A (p=0.017) were found to be significantly downregulated in CAAs group compared to KD without CAAs.

Conclusions:
This is the first gene expression study on children with KD in India. IL1B and MMP9 showed a 6-fold increase in expression, with IL1B significantly reduced after IVIG treatment, indicating the role of the inflammasome pathway in the pathogenesis of KD. Reduced UBB and PRF1 in the pre-IVIG group may suggest a role of interferon pathway and NK cell dysregulation in the pathogenesis of KD. In KD patients with CAA, IL1B was upregulated, while FCGR1A, TLR2, ITGAM, and CD44 were expressed lower compared to those without CAAs.

Funding: Indian Rheumatology Association (IRA research grant).

Keywords: Kawasaki disease, RT-PCR, IL1B, Coronary artery abnormality, Intravenous immunoglobulin

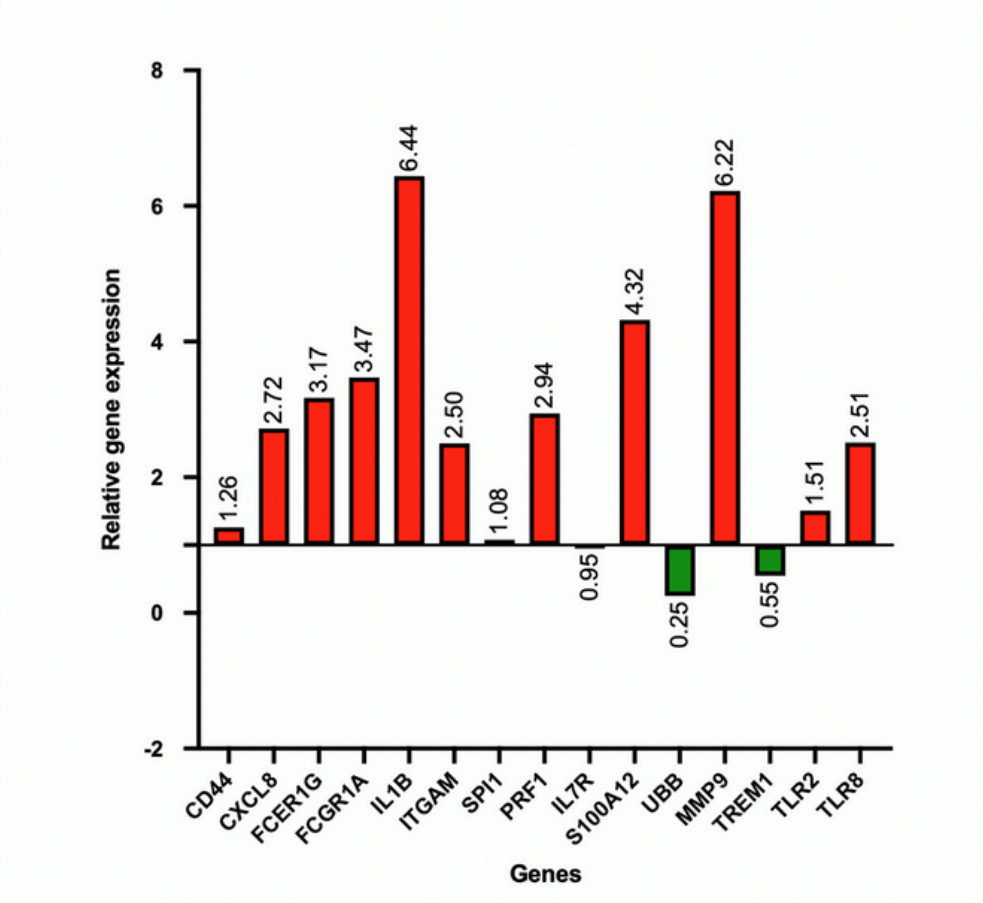
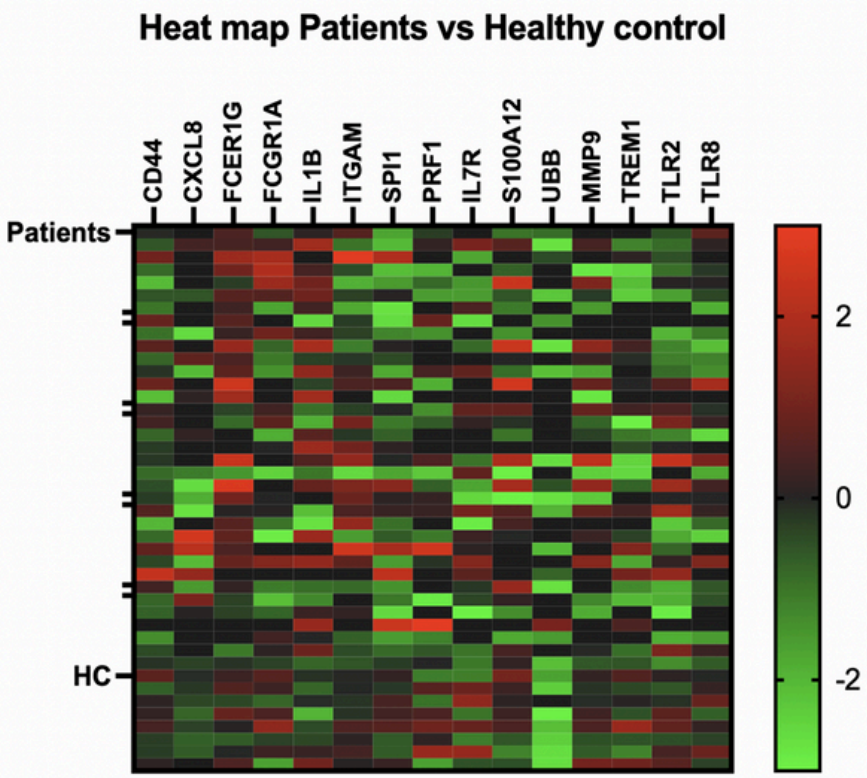


Figure: Relative Gene Expression of 15 Genes in patients with KD in the study (red bars indicate upregulation and green bars indicate downregulation)



Heatmap of relative expression of 15 genes in patients with KD and healthy controls (Red squares indicate upregulation and green squares indicate downregulation)

Table: Relative gene expression between patients with KD (pre and post-treatment) and healthy control

Genes	Healthy control (n=9) Median fold change	KD patients (Pre IVIg) (n=34) Median fold change	KD patients (Post IV Ig) (n=34) Median fold change	P value KD patients Vs Healthy control	P value KD patients (Pre IVIg) VsKD patients (Post IVIg)
CD44	1.049	0.7568	1.126	0.2430	0.0874
CXCL8	0.7695	0.2409	0.9826	0.2310	0.094
FCER1G	1.008	1.466	0.6996	0.1679	0.1965
FCGR1A	1.199	1.030	0.4283	0.8679	0.1024
IL1B	0.7566	1.506	0.6867	0.049	0.0976
ITGAM	0.9890	0.8723	1.3993	0.8488	0.7613
SPI1	1.112	0.5389	0.8332	0.0425	0.3011
PRF1	1.085	0.4622	1.6943	0.0754	0.0313
IL7R	0.6954	0.4282	2.1822	0.0613	0.0022
S100A12	1.107	1.053	0.6672	0.9183	0.5542
UBB	0.1895	0.1124	1.9238747	0.3532	<0.0001
MMP9	0.9429	1.059	1.0894	0.999	0.8793
TREM1	0.7626	0.2712	1.2651	0.0108	0.0187
TLR2	1.046	0.4886	0.6149	0.0852	0.4798
TLR8	0.9844	0.8117	0.5138	0.5051	0.6980

Parallel Chromonychia with Prominent Beau's Lines: A Novel Nail Change in the Subacute Phase of Kawasaki Disease

Authors: Rashmi Gupta1, Gayathri CV1, Rakesh Kumar Pilania1

1. Pediatric Allergy Immunology Unit, Department of Pediatrics, Advanced Pediatric Centre, Postgraduate Institute of medical education and Research, Chandigarh

Background:

Extremity changes in Kawasaki disease (KD) typically include dorsal edema during the acute phase, followed by periungual desquamation in the recovery phase. Several nail alterations have been documented, such as onychomadesis, Beau's lines, pincer nails, leukonychia, and transverse orange-brown chromonychia, particularly in the late acute to subacute phase.
Objective: We report a unique nail finding in the subacute phase of KD in a child.

Case Report:

An 8-year-old boy presented with a 10-day history of fever and rash. He had red, dry, cracked lips and strawberry tongue. On day 10 of illness, when the child presented to us, periungual peeling had begun. A 2D echocardiogram revealed normal coronary arteries and cardiac function. Laboratory investigations showed neutrophilic leukocytosis (total leukocyte count: 20.6 × 10^9/L, 87% neutrophils) and elevated inflammatory markers (C-reactive protein: 167 mg/L, erythrocyte sedimentation rate: 62 mm/hr). A diagnosis of incomplete KD was made, and the patient was treated with intravenous immunoglobulin (IVIG) at 2 g/kg and started on aspirin. The child showed clinical improvement. At the 4-week follow-up, two transversely parallel orange-brown chromonychia with prominent Beau's lines were observed. As the coronary arteries remained normal throughout, aspirin was discontinued at six weeks.

Discussion:

Beau's lines, orange-brown chromonychia, leukonychia, onychomadesis, splinter hemorrhages, and pincer nail changes have all been reported as nail changes associated with KD. Orange-brown chromonychia and Beau's lines are known to occur in the late acute and subacute phases of the disease. To the best of our knowledge, the concurrent appearance of parallel chromonychia with prominent Beau's lines has not been previously reported in a KD patient.

Conclusion:

In addition to Beau's lines, onychomadesis, and splinter hemorrhages, parallel chromonychia can be considered a novel nail change occurring in the subacute phase of KD.

CORONARY THROMBOSIS IN KAWASAKI DISEASE: COMPARATIVE DATA PRE & POST COVID PERIOD

BACKGROUND

Coronary thrombosis is a well described complication in Kawasaki disease (KD), usually in giant coronary aneurysms (CAA) after 2 weeks. Following the Covid pandemic, there has been a shift in clinical presentations of different diseases. This case series of 6 patients over 17 years with multiple CAA & thrombus aims to compare the pattern of coronary thrombosis before and following the pandemic.

PRE COVID PERIOD (2007- 2019) :

CASE 1: 10 months girl presented on day 18 of fever. Diagnosed as complete KD, echo showed giant aneurysm in LMCA, distal RCA, LAD with clot. Treated with IVIG, oral steroid, heparin , alteplase. However during alteplase infusion baby succumbed to death due to myocardial infarction.

CASE 2: 6 weeks old male with KD shock .On day 21 had proximal LAD giant aneurysm(+14.9Z), LMCA and RCA ectasias. IVIG, infliximab. Echo 7 days later showed a thrombus in LAD. LMWH 2mg/kg was initiated. Follow up showed dissolution of clot and regression of aneurysms by 2 years .

POST COVID PERIOD (2020- present day) :

CASE 3: A 3 month old boy, with fever for 26 days, giant aneurysm in LAD with thrombus, small aneurysm in LMCA & RCA, diagnosed as complete KD. Treated with Infliximab, IVIG ,oral steroids, aspirin , LMWH, oral cyclosporine ,clopidogrel. 4 weeks echo showed persistent CAA, with resolution of thrombus.

CASE 4: A 2 month old boy with high grade fever for 10 days with multiple giant aneurysms involving all major coronaries with thrombus in LAD. IVIG, aspirin, LMWH, infliximab, streptokinase .At 5 years follow-up, persistent giant aneurysm of LAD with complete dissolution ofclot.

CASE 5: A 9month old boy presented with 8 days of fever, irritability, thrombocytosis, diagnosed as incomplete KD with RCA & LCX, giant CAA in LAD,LMCA with thrombus. Treated with IVIG, infliximab, cyclosporine, LMWH, steroids. Follow up echo after 5 days show persistent thrombus in LMCA with new developing LAD thrombus.

CASE 6: A 6year old girl, presented with KD shock on day 6 of fever, initial echo showing LMCA,LAD small CAA. Treated with IVIG, steroids, aspirin. Day9 echo revealed giant CAA in LAD, received Infliximab, pulse methyl prednisolone, LMWH. Repeat echo on day 11 showed persistent giant CAA in LAD with thrombus. LMWH dose was increased, 2nd dose of Infliximab, cyclosporine was given. Echo after a month shows diminution of thrombus with multiple CAA.

Observation:

Following the pandemic we have observed an increased incidence of thrombosis, earlier onset by 2 weeks which sometimes progressed in spite of therapy.

"Leptospira-Induced Kawasaki Disease: Unmasking the Hidden Trigger"

Shilpa Kalowar1, Dipangkar Hazarika1, Prabal Barman1
Department of Paediatrics and Neonatology
Gauhati Medical College and Hospital, Guwahati

Introduction: The overlap between kawasaki disease and leptospirosis presents a diagnostic challenge due to their shared symptoms and inflammatory nature. The constellation of clinical similarities between leptospira infection and Kawasaki disease is leading to an increasing consideration of the former as a potential trigger or mimic for the latter, as both conditions can induce widespread vascular inflammation.

Aims and Objectives: To explore the potential association between kawasaki disease and leptospirosis by examining a rare case of leptospira-associated kawasaki disease and the diagnostic challenges posed by their overlapping clinical features.

Materials and Methods: An 8-year-old girl who presented with history of persistent fever for 10 days, pain abdomen ,vomiting with history of conjunctival congestion on day 5 of illness and appearance of rash on day 10.

On examination, she was noted to have unilateral cervical lymphadenopathy with peri-ungual desquamation noted on day 15 during the hospital stay.

Investigations revealed normocytic anaemia with thrombocytosis, leucocytosis, elevated erythrocyte sedimentation rate, raised liver enzyme levels, hypoalbuminemia, elevated c-reactive protein. Relevant investigations for endemic infections were done out of which Leptospira IgM ELISA tested positive. Two-Dimensional Echocardiography was normal.

Results: Patient was treated with intravenous doxycycline followed by intravenous immunoglobulin and patient responded well to treatment and eventually discharged.

Conclusions: This case highlights the possible association of leptospirosis and subsequent kawasaki disease, which has been rarely described.

CLINICAL PROFILE OF CORONARY ARTERY LESIONS AT DIAGNOSIS OF KAWASAKI DISEASE : A RETROSPECTIVE OBSERVATIONAL STUDY FROM A TERTIARY CARE CENTER IN THE EASTERN INDIA

Introduction: Kawasaki disease (KD), a leading cause of acquired heart disease in children is increasingly being reported from India. The coronary artery lesion (CAL) of KD varies from dilatation to aneurysm; hence early diagnosis and treatment are crucial to reduce morbidity and mortality.

Objectives: To note the number of coronary artery lesions at diagnosis of KD, number of coronary artery dilatation (CAD) and coronary artery aneurysm (CAA), frequency of aneurysms in Left Anterior Descending (LAD), Right Coronary Artery (RCA), Left Main Coronary Artery (LMCA), number of small, medium and giant aneurysm in different coronary arteries.

Methods: A retrospective observational study of patients diagnosed with KD from January 2020 to July 2024, at Institute of Child Health Kolkata. Diagnosis was based on American Heart Association 2017 criteria and classified as either complete KD (cKD) or incomplete KD (iKD). Children with congenital heart disease and those who received treatment prior to admission were excluded. CAL were diagnosed by echocardiography using Z score classification.

Results: Among 130 (82 male, 48 female) KD patients included in the study, 51 (39.23%) had CAL at diagnosis. Among these 51 patients, 2 (3.9%) had only CAD while 49 (96.07%) had both CAD and CAA; thus patients with CAA at diagnosis were 37.69% (49/130) of all KD. Complete KD 25/49 (51.02%), incomplete KD 24/49 (49.97%); Patient with single aneurysm 23/49 (46.93%), multiple aneurysm 26/49 (53.06%). Median interval from onset of symptoms to detection of aneurysm was 9 days. A total number of aneurysms noted among those 49 patients with CAA at diagnosis were 101 aneurysms. Number of aneurysms in LAD, LMCA and RCA were 44 (43.56%), 29 (28.71%) and 28 (27.72%) respectively. Number of small (z-score 2.5 to <5) medium (z-score 5 to <10) and giant (z-score >= 10) aneurysms were 76 (75.24%), 23 (22.77%) and 2 (1.9%) respectively. Among all small aneurysm (n=76), LAD had 32 (46.10%), LMCA 24 (31.57%), RCA 18 (23.68%). Of all medium aneurysms (n= 23), 10 (43.47%) in LAD, 5 (21.73%) LMCA, 8 (34.78%) RCA. Giant aneurysms (n=2) involved LAD only.

Conclusion: High number of children had CAA at diagnosis of KD. More than half of CAA patients had multiple aneurysms at diagnosis. Among CAA patients, number of cKD and iKD was comperable. Aneurysms were more common in LAD, followed by LMCA and RCA. Giant aneurysms were detected only in LAD.

.

RESPIRATORY PRESENTATION OF KAWASAKI DISEASE : A DIAGNOSTIC DILEMMA

Introduction : Kawasaki disease (KD) is an acute inflammatory vasculitis affecting young children. Atypical features like respiratory presentations are uncommon, diverse and often unrecognized.

Objective: To present 2 cases who had respiratory presentation but did not respond to antimicrobials and noted to develop features of KD on subsequent monitoring.

CASE 1 : One year old girl with 5 days of fever, cough, a tender erythematous swelling on the anterior part of neck. Suspecting cellulitis of neck, intravenous antibiotic was started. Initially neutrophilic leucocytosis, normal platelet, high C-reactive protein (CRP) (109.2mg/L), sterile blood culture. On day 7 of fever, developed stridor, settled with single dose of dexamethasone injection 0.6mg/kg; became afebrile. MRI neck revealed anterior mediastinitis; antibiotic was upgraded. Repeat total leucocyte count (TLC) and CRP decreased but platelet increased. Fever recurred after 3 days with increasing TLC, CRP, platelet; echocardiogram showed small aneurysm (+2.93z) of left main coronary artery (LMCA); BCG scar reactivation was noted. With a diagnosis of atypical KD, treatment initiated; became afebrile after IVIg infusion. Repeat echo revealed medium sized aneurysm (+6.91z) of LMCA; intensified with infliximab 10mg/kg; low molecular weight heparin (LMWH) added. Oral prednisolone 2mg/kg was added for persistently high CRP (67.3mg/L); tapered over next 2 weeks. 2 weeks echocardiogram showed small aneurysm (+2.1z) of LMCA; LMWH was stopped. Echocardiogram at 4 weeks was normal.

Case 2 : Two months old girl with 6 days of fever, cough, redness of eyes, loose greenish stool, 1 day of respiratory distress; had tachypnea, intercostal retraction; started on antibiotics, nebulizations, oxygen. Investigations revealed anaemia; normal TLC, platelet; increased CRP (58.3 mg/L). Chest X ray showed right upper lobe opacities. On day 7 of fever, developed redness of lips, edema of feet, rashes over body, erythema at BCG site with increasing TLC (28,400/cmm,N63%) CRP (192.3mg/L), platelet (10,18,000/cmm). With a diagnosis of complete KD, IVIg, aspirin were started; antibiotics omitted, became afebrile the next day. In view of infantile onset with of atypical feature (pneumonitis), oral prednisolone 2mg/kg was added. Repeat X ray after 48 hours of IVIg showed improvement. Echocardiography at onset through 2 weeks were normal.

Conclusion : Looking beyond the obvious through a proper clinical examination is the key to a successful diagnosis. Careful monitoring is essential because some features of KD may evolve over time. Timely initiation of therapy is crucial as atypical KD carries a high risk of coronary aneurysm.

OUTCOME OF KAWASAKI DISEASE PATIENTS TREATED WITH INTRAVENOUS IMMUNOGLOBULIN + CORTICOSTEROIDS

INTRODUCTION:

The RAISE (Randomized Controlled Trial to Assess Immunoglobulin plus Steroid Efficacy for Kawasaki Disease) based on the Kobayashi score to predict IVIG resistance revived interest worldwide in the use of steroids.

AIM OF THE STUDY:

To evaluate the response in patients treated with steroids in addition to Intravenous Immunoglobulin (IVIG)+ low dose aspirin (5mg/kg/day).

MATERIALS AND METHODS:

Ø A Retrospective observational study January 2020 to October 2024 in Institute of Child Health, Kolkata

ØInclusion criteria: Children diagnosed with Kawasaki Disease (KD) as per AHA 2017 guidelines were treated with IVIG (2gm/kg), low dose aspirin (5mg/kg/day).

Patients with any of the following features received additional oral prednisolone / intravenous methylprednisolone and were enrolled in this study:

1.Kobayashi score >5
2.IVIG resistance
3.Persistently high inflammatory marker (CRP) 36 hrs following IVIG
4.Presence of myocarditis
5.Dilated coronary artery (Z score 2 to 2.4)
6.Small Coronary artery aneurysm (CAA) (Z score 2.5 to 4.9)

Data was collected from hospital records, as well as follow up data of KD patients attending OPD, and recorded in excel sheets. Clinical characteristics and echocardiogram findings were analyzed in these infants.

Exclusion criteria:

1. KD patients with CAA Z score >/= 5.
2. Treated with Infliximab (IFX) prior to corticosteroids.

RESULTS:

145 (106+39) children were diagnosed with KD during the study period, of these 51 received corticosteroids. Median age of recipients 17 months. There were 31 males and 20 females. Median time for IVIG administration was 8 days. The median time for corticosteroids initiation was 10 days from the onset of fever.

29/51 received steroids for small aneurysm (Z 2.5 to 4.9), 9/51 for dilated coronaries, 2/51 for presence of myocarditis.

7/51 patients received steroids +IVIG for Kobayashi score >5, of which 4 continued to have fever and later received IFX. 6/51 received steroids for persistently high CRP following IVIG along with clinical signs of ongoing inflammation (such as oral mucositis).

Of the 29 patients who received corticosteroids for small CAA, 7 progressively increased to medium CAA. 22 small CAA and all dilated coronaries have regressed after a median duration of 21 days.

Limitation: Single arm retrospective study with small sample size.

CONCLUSION:

Corticosteroids can be used for treatment intensification in KD patients with dilated coronaries (< 2.5 Z). However, 57% patients who received steroid based on Kobayashi scoring needed therapy augmentation and 28% patients with small CAA progressed to medium size.

Low dose vs medium dose aspirin in acute phase of Kawasaki disease- an experience from a tertiary care hospital of West Bengal, India

Sarbik Sarkar1, Deblina Patra1, Bodhisatya Das1, Sayani Pan2, Kamirul Islam2, Taraknath Ghosh3, Mithun Chandra Konar4, Archan Sil5
1 Post graduate Trainee, Department of Pediatrics, Burdwan Medical College and Hospital, Bardhaman, West Bengal
2 Senior Resident, Department of Pediatrics, Burdwan Medical College and Hospital, Bardhaman, West Bengal
3Professor, Department of Pediatrics, Burdwan Medical College and Hospital, Bardhaman, West Bengal
4 Associate Professor, Department of Pediatrics, Burdwan Medical College and Hospital, Bardhaman, West Bengal
5Assistant Professor and In charge Pediatric Clinical Immunology and Rheumatology Clinic, Department of Pediatrics, Burdwan Medical College and Hospital, Bardhaman, West Bengal
Low dose vs medium dose aspirin in acute phase of Kawasaki disease- an experience from a tertiary care hospital of West Bengal, India

Background- Kawasaki disease (KD) is a common childhood vasculitis. There are controversies regarding the dosage of aspirin in the acute phase of KD, despite years of extensive research. The aim of this study was to evaluate the efficacy of intravenous immunoglobulin (IVIG) plus low-dose aspirin (3-5 mg/kg/d) in comparison to IVIG plus medium-dose aspirin (30-50 mg/kg/d) in the treatment of acute KD.

Methods- Data were collated by reviewing clinical records of patients registered between August 2022 and August 2024 in the Pediatric Rheumatology clinic of a tertiary care centre of eastern India. AHA (American Heart Association) 2017 criteria were used for diagnosis of KD. All patients initially received a single dose of IVIG (2g/kg) in the acute phase. Patients were classified into two groups. Group 1 included 26 cases treated with IVIG plus 3-5 mg/kg/day aspirin (low-dose group), and group 2 included 30 cases treated with IVIG plus 30-50 mg/kg/day aspirin (medium-dose group). Clinical (fever) and laboratory response (inflammatory parameters) were compared between the two groups. The primary outcome measure was the occurrence of coronary artery lesions (CALs) at the subacute or convalescent stage. $P < 0.05$ was considered as significant.

Results- Fifty-six patients who fulfilled the criteria of KD were included in this study. While 30 patients with KD were found to be treated by low-dose aspirin, 26 of them received medium-dose aspirin in the acute phase. There was no significant difference among the two groups (low dose vs. medium dose) in terms of the anti-inflammatory effect revealed by white blood cell count, percentage of neutrophils in white blood cells, platelet count, and C-reactive protein level. CALs were found in 11% (3/26) of the low dose group and 13% (4/30)

of the medium dose group ($\chi 2$ = 0.04, P= 0.84). Those who had CALs were subsequently treated as per the institutional protocol. Incidence of adverse effects, particularly gastritis was lesser in children who received low-dose aspirin (3.8% vs 23.3%, P=0.04).

Conclusion- We found no significant benefit of using medium dose aspirin in acute phase of KD but there were higher incidences of complications. Further prospective multicentric studies incorporating a large number of patients are needed to confirm this finding.

CHANGING EPIDEMIOLOGY OF KAWASAKI DISEASE IN EASTERN INDIA

Jigna N Bathia1, Nazneen Ahmed2, Subhajit Dey Sarker2, C Ravali Pratima Goud3, Anindita Nandi 3Priyankar Pal4
1 Assistant Professor, 2 Senior Resident, 3 Fellow, 4Prof and HOD,
Department of Paediatric Rheumatology, Institute of Child Health, Kolkata

INTRODUCTION: Incidence of Kawasaki disease (KD) is rising, however there is limited epidemiological data available from Eastern India.

AIM: To assess the epidemiological profile of patients diagnosed as Kawasaki Disease from 2009 to December 2023.

METHODOLOGY: Retrospective, observational study of patients diagnosed as KD at a tertiary referral centre in Eastern India.

RESULTS: 2009 to Feb 2022: A total of 229 patients were diagnosed as Kawasaki disease this period. 20% were incomplete KD. 63 (27%) were less than one year of age, 13 were more than 5 years of age (2%), 21 (9%) had intravenous immunoglobulin (IVIG) resistant of whom 14 received infliximab (IFX) after IVIG. Over the years the incidence of coronary artery aneurysms (CAA) increased from 15% till 2017 to 17% in 2018 and 32% in 2019 to February 2020. 10 patients had giant coronary artery aneurysms. Two of whom with late presentation and intraluminal thrombus of whom one succumbed.

March 2020 to March 2022: This period was majorly affected by COVID 19. A total of 54 patients were diagnosed as Kawasaki disease during this period. 16 (30%) were less than one year of age, 3 (6%) were more than 5 years of age, 27 were males, 10 were incomplete KD (19%) , 4 (7%) had intravenous immunoglobulin (IVIG) resistant who received infliximab (IFX) after IVIG. 15 (28%) had CAA none giant.
April 2022 to December 2023: 80 patients were diagnosed as KD , 22 (28%) were less than one year of age, 1 (2%) was more than 5 years of age, 71 (89%) were males, 24 were incomplete KD (30%) , 6 (8%) had intravenous immunoglobulin (IVIG) resistant who received infliximab (IFX) after IVIG. 26 (32.5%) had CAA, none giant. One had intraluminal thrombus formation.
The epidemiological trends of the last few years have been enlisted in Table 1.

CONCLUSION : There is an annual rise in the number of patients diagnosed as KD with increasing number of patients with incomplete presentations of KD , rising number of infants, rising number of patients with CAA at diagnosis.

Table 1: Trends from 2019 to 2023

	2019	2020	2021	2022	2023
Number of patients	32	33	22	35	36
Median age (mo)	29	17	17.5	16	18
No of infants	27%	58%	32%	40%	33%
No of males	75%	88%	63%	74%	63%
Incomplete Kd	37.5%	62%	63%	42%	36%
IVIG resistance	15%	8%	22%	14%	8%
CAA	25%	29%	45%	31%	36%
Day of illness admission (median)	8	7	6	6	7
Day of illness CAA being diagnosed (median)	Data unavailable	12	9.5	9	9.5

Title:Role of CT Coronary Angiography for Coronary Artery Assessment in Kawasaki Disease During the Acute Phase – Experience from Chandigarh, North India

Authors: Dev Desai1, Rakesh Kumar Pilania1, Suprit Basu1, Ankur Kumar Jindal1, Pandiarajan Vignesh1, Deepti Suri1, Manphool Singhal2, Surjit Singh1

Affiliations
1Pediatric Allergy Immunology Unit, Department of Pediatrics, Advanced Pediatric Centre, PGIMER
2Department of Radiodiagnosis and Imaging, PGIMER

Introduction: Kawasaki disease (KD) is a common medium vessel vasculitis of childhood with a predilection of coronary artery involvement which can have long-term sequelae. Traditionally, 2D echocardiography (2DE) has been the primary modality for assessing coronary arteries in KD. However, CT coronary angiography (CTCA) is emerging as a superior tool for coronary evaluation. This study compares CTCA with 2DE in assessing coronary arteries during the acute phase of KD.

Methods:
We have reviewed the records of patients with KD who underwent CTCA during the acute phase of the disease. Since 2013, we have been carrying out additional imaging in children with CAAs on TTE using dual-source CT coronary angiography (CTCA) on a 128-slice scanner (Siemens SOMATOM Definition Flash, Erlangen, Germany). After February 2022, these scans were performed on a 192-detector dual-source CT scanner (Siemens SOMATOM Force, Erlangen, Germany). Findings on CTCA were compared with 2DE.

Results:
Out of 56 patients, indications for CTCA were confirmation of coronary artery aneurysms (CAAs) detected on TTE in 49 patients, non-visualization of coronaries, non-tapering of coronaries, and anterior chest wall ulcer in 4, 2, and 1 patient respectively.
In the 49 patients who had CAA on TTE, CTCA confirmed CAAs in 36 cases while the others had normal coronaries on CTCA. One patient with non-tapering of coronaries on TTE had abnormal findings on CTCA, while all 4 patients with non-visualized coronaries and the lone patient with anterior chest wall ulcer had normal coronaries on CTCA.

Additional advantages observed with CTCA were the detailing of aneurysms in left circumflex coronary (n=8) and distal segments of coronaries (n=4), the ability to determine the detailed morphology and dimensions of aneurysms, and the detection of thrombi.

Conclusions:
CTCA demonstrates significant advantages over 2DE in detecting coronary lesions during the acute phase of KD, offering enhanced visualization of aneurysm morphology and distal coronary segments.

Title: Recurrent Skin Peeling in a child with Kawasaki Disease: An anamnestic respone

Authors: Mrinmoy Das, Sneh Kumar, Pratap Kumar Patra, Swarnim Swarim, Chandra Mohan Kumar
All India Institute of Medical Sciences, Patna.

Background:
Kawasaki Disease (KD) is an acute, self-limiting vasculitis predominantly affecting children under the age of 5, with potential complications involving coronary artery aneurysms if untreated. We report a patient with recurrent skin peeling on day 2 and day 8 of illness which is extremly uncommon.

Case Presentation:
A 7-year-old male was admitted with complaints of high-grade fever persisting for eight days, painful swallowing, redness of the palms with desquamation on day two, and a non-pruritic erythematous rash spreading across the body. The fever was sudden in onset, without diurnal variation, and transiently responsive to antipyretics. He was treated elsewhere without any improvement. The palmar peeling subsided on day four of illness.

He was referred to us on day eight of illness, and on examination, he was noted to have palmar peeling,"strawberry tongue", cervical lymphadenopathy, and bilateral conjunctival redness without discharge. Systemic examination was normal. Vital signs were stable, and laboratory findings showed elevated inflammatory parameters. 2D echocardiography showed normal coronary artery dimensions.

The diagnosis of Kawasaki Disease was established based on clinical features. Immediate treatment was initiated with IVIg at a dose of 2 g/kg over 12 hours and aspirin at an anti-inflammatory dose of 50 mg/kg/day in 4 divided doses. The child demonstrated significant clinical improvement, with fever resolution within 48 hours of treatment. Laboratory markers, including CRP, showed a marked decrease. A follow-up echocardiogram performed before discharge confirmed normal coronary artery Z-scores. Desquamtion resolved after day 15 of illness.

Conclusion:
Periungual skin peeling is a characteristic features of KD and is usually seen in second week of illness. However, we noted two episode of skin peeling in the index case and it has been not reported earlier.

CLINICAL AND LABORATORY PROFILE OF CHILDREN WITH KAWASAKI DISEASE AT A SINGLE TERTIARY CARE HOSPITAL IN NORTH-EAST INDIA

Rupam Baruah1, Prabal Barman1, Dhrubajyoti Sharma2, Anupama Deka1
1Department of Pediatrics, Gauhati Medical College, Guwahati
2Department of Pediatrics, Fakhruddin Ali Ahmed Medical College, Barpeta

ABSTRACT:

INTRODUCTION:

Kawasaki disease (KD), also known as mucocutaneous lymph node syndrome is a common cause of acquired heart disease in children less than 5 years of age. There is paucity of data on the clinical phenotype and outcome of patients with KD from North-East India.

OBJECTIVE:

To study the clinical profile and outcome of patients admitted with KD at a single tertiary care center in North-East India.

METHODS: A retrospective study of patients with KD admitted in Pediatric ward, Gauhati Medical College, Guwahati from April 2017 to April 2024 was undertaken. The clinical phenotype, laboratory investigations and 2-dimensional echocardiographic findings were analyzed in detail.

RESULTS: Of the total 38,290 admissions, 101 patients (0.26%) were admitted as KD. Male: female ratio was 65:36. The most common clinical findings were fever (100%), oral mucosal changes (86.1%), extremity changes (79.2%), conjunctival injection (74.2%), polymorphous rash (68.3%), and cervical lymphadenopathy (61.3%). Nineteen patients had coronary artery abnormalities (CAA) and 26 (25.7%) patients had incomplete presentation. Atypical presentation included acute gastroenteritis (n=6), arthritis (n=3), acute hepatitis (n=3), acute intestinal obstruction (n=1), seizure (n=1), pulmonary manifestation (n=1) and macrophage activation syndrome (n=1). All patients received intravenous immunoglobulin (IVIG) (2g/kg) and aspirin (3 mg/kg/day). Eleven patients were IVIG resistant. Of these 9/11 received a single-dose infliximab (5-7 mg/kg) and 2/11 received intravenous methylprednisolone (30 mg/kg/day for 3 days followed by oral taper). Fourteen patients had regression of CAA at median follow-up of 20 months.

CONCLUSION: Patients with KD in North-East India had similar presentation as compared to other population cohorts in India. Low proportion of incomplete presentation may suggest the still lack of awareness and missed diagnosis of KD in this part of the country.

Title: Progressive Enlargement of a Giant Coronary Aneurysm in a Child with Kawasaki Disease: Long-Term Follow-Up

Authors: Gaurav A. Kachole, Reva Tyagi1, Rakesh Kumar Pilania1, Manphool Singhal2, Surjit Singh1
1 Allergy and Immunology Unit, Advanced Pediatrics Centre, Postgraduate Institute of Medical Education and Research, Chandigarh, India
2 Department of Radiodiagnosis, Postgraduate Institute of Medical Education and Research, Chandigarh, India

Background: Kawasaki disease (KD) is the most common vasculitis in childhood. It is the leading cause of heart disease in the pediatric age group in the West. We present a 5-year-old boy who showed progression in the size of an aneurysm during long-term follow-up.

Case details: A 5-year-old boy presented to us with acute febrile illness along with redness of eyes, dry, cracked lips, and strawberry tongue. Physical examination revealed non-exudative conjunctival injection along with changes in oral mucosa. Blood investigations revealed leucocytosis, elevated acute phase reactants (C-reactive protein- 78mg/L, erythrocyte sedimentation rate- 74 mm/hour), and elevated pro-brain natriuretic peptide (798pg/ml). The possibility of KD was considered. 2D-echocardiogram of the coronary arteries showed loss of tapering of left anterior descending artery [LMCA: 3mm (-0.08 z), LAD: 2.0mm (+1.13z), RCA: 2.3mm (-0.62z)]. Intravenous (IV) immunoglobulin was given @2g/kg and oral aspirin at anti-inflammatory dose with which the fever subsided. On follow up after 2 weeks he was found to have ectasia of left coronary artery [LMCA: 3.7mm (+3.42z), LAD: 2.5mm (+1.89z), RCA: 2.3mm (+1.28z), LCx: 2.7mm (+2.32z)] and aneurysm in the distal end of right coronary artery [5.8mm (+9.08z)] which was confirmed on computed tomography (CT) coronary angiogram (CTCA) on 128-slice dual source platform. He received IV Infliximab (5 mg/kg). He was discharged on subcutaneous low molecular weight heparin (LMWH) which was later changed to oral warfarin 4 months later. He also received aspirin and atorvastatin. A repeat CTCA 5 years later (LMCA: 3.6mm, LAD: 2.5mm, RCA:2.7mm, LCx: 2.7mm) showed progression in the size of the aneurysm in the distal right coronary artery [7.2 mm (+9.99z] but there was no calcification. He remained well since then. Factor Xa levels (0.48 IU/ml) and international normalized ratio (INR- ranging1.6 to 3.16) were monitored during follow-up and were well within the expected range on anticoagulation.

Conclusion: This case highlights the role of CTCA in the detailed visualization of coronary artery abnormalities in children with KD. Though enlargement of a coronary aneurysm after the acute phase in KD is an extremely rare phenomenon, physicians should be aware of coronary sequelae, including dilation of prior aneurysms. Growth of the coronary artery with somatic growth, abnormalities of the coronary arterial wall, and hemodynamic factors are among the many causes cited in the causation of aneurysm in the late period after KD.

Title: ASSESSMENT OF MYOCARDIAL STRUCTURE AND FUNCTION IN MULTISYSTEM INFLAMMATORY SYNDROME IN CHILDREN USING CARDIAC MRI AND SERUM GALECTIN-3: FOLLOW-UP STUDY AFTER A MEAN DURATION OF 23.58 MONTHS

Authors: Akanksha Dhiman1, Rakesh Kumar Pilania1, Sathish Kumar1, Manphool Singhal2, Ankur Kumar Jindal1, Sanjeev Naganur3, Surjit Singh1

Affiliation:
1Pediatric Allergy Immunology Unit, Department of Pediatrics, Advanced Pediatric Centre, Postgraduate Institute of medical education and Research, Chandigarh
2Department of Radiodiagnosis and Imaging, Postgraduate Institute of medical education and Research, Chandigarh
3Department of Cardiology, Advanced Cardiac Centre, Postgraduate Institute of medical education and Research, Chandigarh

Background: MIS-C is a novel disease in children and adolescents that emerged in April 2020 in the context of SARS-CoV-2 pandemic. It is associated with significant myocardial dysfunction and the development of coronary artery abnormities (CAAs) during acute phase. Although ECHO has been the preferred imaging modality for follow-up of patients with MIS-C, it has several limitations, as it is not a sensitive technique for assessment of myocardial dysfunction. Cardiac Magnetic Resonance Imaging (CMRI) is now being increasingly recognized as a useful imaging modality for myocardial assessment. There is paucity of follow-up data on this subject.

Methods: This prospective observational study was conducted between July 2022 - December 2023 in the Allergy Immunology Unit, Advanced Pediatrics Centre, PGIMER, Chandigarh. Diagnosis of MIS-C was based on WHO guidelines. Twelve patients with MIS-C underwent CMRI on 3 Tesla - Siemens Ingenia platform. Serum Galectin-3 levels were also analyzed on the same day.

Results: The mean age of the study cohort was 11.33 years (range 9-13 years). Of the 12 patients, 7(58%) had low ejection fraction (EF < 55%), and 3(25%) presented with CAAs. ECHO and CMRI were carried out at a mean follow-up of 23.58 months. While ECHO showed normal EF in all patients, CMRI revealed low EF (<55 %) in 4 (33.3%) patients. Further, on CMRI 5(42%) had end-diastolic volume <-2 Z score, 1(8.3%) had end-systolic volume <-2 Z score, and 7(58%) had stroke volume <-2 Z score. EF on CMRI was lower (mean-58.34±7.75) in comparison to ECHO (mean-62.33±5.28). Correlation coefficient for EF on CMRI and on ECHO was 0.2. There were no regional wall motion abnormalities or late Gadolinium enhancement in any of the patients. However, 2 (16.6%) patients had abnormal native myocardial T1 values (>1250ms) suggestive of myocardial fibrosis. None had any CAAs detected on CMRI or ECHO at follow-up. Mean serum Galectin-3 in patients with

MIS-C at 23.58 months of follow-up was 13.49±7.63 ng/ml. This was much higher when compared to age-matched healthy controls (mean-7.48±3.13) (p=0.063).

Conclusions: Our study highlights the efficacy of CMRI as a useful imaging modality for long-term assessment in patients with MIS-C. At a mean follow-up of 23.58 months, no patient showed low EF on ECHO, but CMRI detected impairment in 33.3% of cases. Persistent elevation of Galectin-3 suggests ongoing myocardial inflammation. Patients with MIS-C may continue to have functional abnormalities. CMRI appears to be a better imaging modality for the diagnosis of myocarditis in this situation, as compared to ECHO.

Title: Cardiac and Coronary Abnormalities in Incomplete Kawasaki Disease: A Single-Center Study

Authors: Mahapatra A1, Patro D2, Rath D3

Background: Kawasaki disease (KD) is a systemic vasculitis predominantly affecting children. While incomplete KD is recognized as a diagnostic challenge, the prevalence and spectrum of cardiac complications in these patients remain less well understood.
Objective: To investigate the incidence and nature of cardiac and coronary abnormalities in incomplete Kawasaki disease.

Methods: A retrospective study was conducted on 21 patients diagnosed with Kawasaki disease at our center. Patients were classified as complete or incomplete KD based on clinical criteria. Cardiac evaluation was performed using echocardiography, focusing on myocarditis and coronary artery abnormalities.

Results: Of the 21 patients, 11 (52.4%) were diagnosed with incomplete Kawasaki disease. Cardiac involvement was identified in 8 of these 11 patients (72.7%). Notably, 4 patients had myocarditis, while the remaining 4 exhibited coronary artery dilatation. These findings represent a significantly higher prevalence of cardiac complications compared to the current literature on incomplete KD. In contrast, 4 of the 10 patients (40%) with complete KD had cardiac abnormalities, primarily coronary artery involvement.

Conclusion: Our study highlights a strikingly high prevalence of cardiac complications, including myocarditis and coronary artery dilatation, in incomplete Kawasaki disease. This underscores the critical need for thorough cardiac assessment in these patients, even in the absence of full diagnostic criteria for KD. Larger, multicenter studies are needed to corroborate these findings and guide future management strategies.

1.Prof Dr. Anuspandana Mahapatra, Fellow Pediatric Cardiology, Professor
2.Dr Debasis Patro, Pediatric Rheumatologist, Assistant Professor
3.Prof Dr Debasmita Rath, Pediatric Intensivist, Professor
Department of Pediatrics, IMS & SUM Hospital, Bhubaneswar, Odisha

Title: Autoimmune Shenanigans: The SLE and Kawasaki Crossover

Authors: Ghosh R, Maheshwari A, Mahto D
Division of Immunology & Rheumatology, Department of Paediatrics, Lady Hardinge Medical College & Hospital

Introduction: Systemic lupus erythematosus (SLE) is a systemic autoimmune disease with a multifactorial pathogenesis, including immune dysregulation, genetics, environmental factors, and viral influences. Its incidence ranges from 0.36 to 2.2 per 100,000, with variable presentations that can delay diagnosis, particularly in children who exhibit a more aggressive disease course than adults. In contrast, Kawasaki disease (KD) is a self-limiting febrile illness characterized by medium vessel inflammation, predominantly affecting coronary arteries and leading to acquired heart disease in children. This case highlights the association of vasculitis with coronary artery dilation, reinforcing the diagnosis of KD.

Case Description: A 13-year-old girl presented with a one-week history of fever that subsided with antibiotics, followed by 10 days of blackish discoloration of fingers and toes, accompanied by burning sensations. She had a significant past medical history, including self-amputation of a digit and toe due to similar symptoms. Examination revealed gangrenous changes in both upper and lower limbs, with variable peripheral pulses. Blood pressure was 140/110 mmHg with no significant differences in four limb BP, and serological tests confirmed systemic lupus erythematosus (SLE) with positive ANA and elevated dsDNA.
ECHO findings: moderate pericardial effusion. CT angiography showed thrombosis in the right distal radial artery and cephalic vein, coronary artery dilation, and evidence of pulmonary artery hypertension. A working diagnosis of SLE overlapping with Kawasaki disease was made, and the patient was treated with IVIG, aspirin, and hydroxychloroquine, resulting in clinical improvement.

Discussion and Conclusion: Coronary artery vasculitis and gangrene can manifest as complications of Kawasaki disease (KD, especially in the context of systemic lupus erythematosus (SLE). The overlapping triggers for autoimmune diseases suggest that KD can occur against the backdrop of SLE. It is possible that the child had an undiagnosed case of KD a year prior, leading to coronary artery dilation and subsequent gangrene. The documented decrease in coronary artery diameter on 2D echocardiogram following IVIG treatment supports the KD diagnosis. Given the concurrent autoimmune processes in both SLE and KD, symptoms may present more aggressively, increasing the likelihood of misdiagnosis. Since IVIG is not a standard treatment for SLE, the progression of KD could go unrecognized in such patients.

Defervescence in Kawasaki disease when spontaneous

Debate in treatment decisions becomes conspicuous

Authors: Gayathri C V1, Rakesh Kumar Pilania1, Prabal Barman1, Yamini Sharma1, Dev Desai1, Ankur Kumar Jindal1, Pandiarajan Vignesh1, Deepti Suri1, Manphool Singhal2, Surjit Singh1

1- Pediatric Allergy Immunology Unit, Department of Pediatrics, Advanced Pediatric Centre, Postgraduate Institute of medical education and Research, Chandigarh
2- Department of Radiodiagnosis, Postgraduate Institute of medical education and Research, Chandigarh

Abstract:
Objectives: Intravenous immunoglobulin (IVIg) is the standard of care in Kawasaki disease (KD). However, a subset of patients defervesces spontaneously, and guidelines are equivocal about the need for IVIg in such cases. We aimed to evaluate the clinical profile of children with KD who defervesced spontaneously and did not receive IVIg.

Methods: A review of records of patients with KD who defervesced spontaneously were analysed in detail. Patients who were afebrile 48 hours with normal inflammatory [k1] parameters (C-reactive protein, ESR) and no coronary artery abnormalities (CAA) at presentation were taken. [k2] We divided this cohort into 2 groups - Early defervescence (edKD): children with KD who defervesced <10 days of onset, and Late defervescence (ldKD): children with KD who defervesced 10 days of onset. Their clinical characteristics at diagnosis and at follow up were analysed in detail.

Results: Of the 1350 patients with KD, 109 patients [k3] (83 boys) defervesced spontaneously. Median age at onset of disease was 6 years (range: 0.8-15 years). Median duration of fever and median time for arrival at hospital was 5 days (range: 1-21 days) and 15 days (range: 4-40 days) respectively[k4] . Sub[k5] -group analysis showed that rash, oro-mucosal changes, changes in extremities and cervical lymphadenopathy were significantly increased in ed KD in comparison to ld KD. None of the patients in either group had developed any CAA either at baseline or over a cumulative follow-up of 862 patient-years.

Conclusions: Patients with KD who defervesce spontaneously and have normal inflammatory parameters with no CAA at presentation have good clinical and coronary outcomes, and may not require treatment with IVIg.

Keywords: Kawasaki disease, IVIg, spontaneous defervescence

COMPARISON OF CLINICAL, LABORATORY PARAMETERS AND OUTCOMES IN CHILDREN DIAGNOSED WITH KAWASAKI DISEASE (KD) AND MULTISYSTEM INFLAMATORY RESPONSE SYNDROME IN CHILDREN (MIS-C) DURING COVID-19 PANDEMIC AT A TERITARY CARE HOSPITAL

Shivani Singh a, Annu Maheshwaria, Department of Paediatrics, Lady Hardinge Medical College and Kalawati Saran Children's Hospital, New Delhi, India.

Introduction: Kawasaki Disease (KD) and Multisystem Inflammatory Syndrome in Children (MIS-C) are both innate immune hyper inflammatory states with multisystem involvement with varying severity. Both of the diseases having diagnostic criteria well established till date (1,2). Various similarities and dissimilarities among the two have been recognised over the past couple of years(3,4).

Objectives: Keeping in mind the various differences in the diagnostic criteria and the management plans while dealing with KD as well as MISC, we ought to compare the clinical as well as laboratory parameters and outcomes in children diagnosed with MISC and KD during the advent of SARS COVID pandemic.

Methods: This was a descriptive comparative study done during the advent of COVID-19 pandemic. Univariate analysis was done using various statistical tests including Student t- test, Fischer exact tests for the numerical data and Chi-square test for the categorical data. Multivariate logistic regression was done for the significant findings and the Odd's ratio were calculated. We had anticipated the differences in the age groups, prevalence of GI symptoms, sepsis markers and manifestation of MODS in our comparison of the two groups.(5)

Results: We found significant difference in terms of Clinical presentation including history of fever and its duration, being higher in cases of KD with mean duration being 9.214±7.195 S.D. days (p=0.004) and predominance of GI symptoms in cases of MISC (p=0.002) along with various laboratory markers including Procalcitonin (PCT (p=0.001), rise in AST (p=0.029) and titres of proBNP all three being higher in cases of MISC (p=0.001). Mean age group of 0- 60 months was found to be having maximum no. of children of both KD as well as MISC. Significant difference was also found in terms of therapy regarding use of anticoagulants predominantly in cases of KD (p=0.001). No significant difference was found among mean age, nutritional status, other laboratory markers like Haemogram, Coagulation profile and markers of inflammatory response.

Conclusion: Various similarities and differences were revealed on analysis of our study which might help in the better understanding of the clinical profile of both the diseases and guiding through adequate management.

References:

1. Zhu F, Ang JY. 2021 Update on the Clinical Management and Diagnosis of Kawasaki Disease. Curr Infect Dis Rep. 2021 Mar;23

2. CDC. Multisystem Inflammatory Syndrome (MIS). 2024. Case Definitions and Reporting. Available from: https://www.cdc.gov/mis/hcp/case-definition-reporting/

3. Tong T, Yao X, Lin Z, Tao Y, Xu J, Xu X, et al. Similarities and differences between MIS-C and KD: a systematic review and meta-analysis. Pediatr Rheumatol. 2022 Dec 5;20

4. Darby JB, Jackson JM. Kawasaki Disease and Multisystem Inflammatory Syndrome in Children: An Overview and Comparison. 2021;104

5. Yavuz L, AlHamdani S, Alasrawi S, Wafadari D, Al-Fraihat A, Bebars MA, et al. Kawasaki disease (KD) and multisystem inflammatory syndrome in children (MIS-C) in a Middle Eastern patient cohort. Pediatr Rheumatol. 2023 Jun 29;21