

Untreated Kawasaki Disease with Giant Coronary Aneurysm with Thrombosis Presenting in STEMI

Anindita Nandi¹, Payel Kundu², Rhea Singh², Ravali P Goud³, Jigna N Bathia⁴, Debasree Gangopadhyay⁵, Lopamudra Mishra⁶ Suprit Basu⁷, Priyankar Pal⁸

1. Senior Resident in Pediatric Rheumatology Unit, Institute of Child Health, Kolkata, India

email- aninditarheumat24@gmail.com

phone- 9836030578

2. Fellow in Pediatric Rheumatology Unit, Institute of Child Health, Kolkata, India

3. Ex-Fellow in Pediatric Rheumatology Unit, Institute of Child Health, Kolkata, India

4. Ex-Consultant in Pediatric Rheumatology Unit, Institute of Child Health, Kolkata, India

5. Consultant Pediatric Cardiology, Narayana Health, Kolkata, India

6. Assistant Professor, IPGMER and SSKM Hospital, Kolkata, India

7. Consultant in Pediatric Rheumatology, IPGMER and SSKM Hospital, Kolkata, India

8. Professor and Head, Department of Pediatrics and Pediatric Rheumatology Unit, Institute of Child Health, Kolkata, India

Introduction:

Giant coronary artery aneurysm (CAA) with thrombosis is the most severe complication of Kawasaki disease (KD). Timely diagnosis and treatment is crucial to prevent such complications.

Case:

A 4-year-7-month-old boy had 10-day high-grade fever with right cervical adenopathy treated with oral antibiotics. Day 4: of fever, itchy maculopapular rash; started oral prednisolone 1mg/kg/day. Day 5: rash increased with bilateral non-purulent conjunctivitis, redness of lips and hand-foot edema; started on oral antiallergics. Fever subsided by day 9-10. On day10, KD was suspected. Echo: CAA LAD Z+4.9; ESR 30 mm/hr, CRP 3.28 mg/L. Aspirin 5mg/kg/day was started but IVIG was withheld as he was afebrile and was discharged for echo follow-up.

Four days later, he presented with recurrent non-bilious vomiting, chest and abdominal pain, initially treated as acute gastroenteritis with antiemetics. He became lethargic

next day. Repeat echo: giant CAA in LAD with near-occlusive thrombus and medium CAA in LMCA. ECG: ST-elevation myocardial infarction (STEMI).

Thrombolysis was done with Alteplase (3 doses) with recanalization of LAD. Intensified immunomodulation was given with infliximab and cyclosporine along with dual antiplatelet, continuous IV heparin infusion, atorvastatin, beta-blocker, diuretics and ACE inhibitor. At 4 weeks echo still showed giant CAA in LAD, RCA, LMCA with residual thrombus. By 5 weeks thrombus resolved. Cyclosporin was stopped after 8 weeks. Transition to warfarin in interim led to recurrence of LAD thrombus requiring escalation back to LMWH, highlighting need for potent sustained anticoagulation. At 1 year, echo and CTCA showed giant CAA in LAD, RCA with moderate LV dysfunction. Presently stable on beta-blocker, statin, ACE inhibitor, dapagliflozin, aspirin and rivaroxaban.

Conclusion:

This case highlights that afebrile status after steroid therapy does not mean inactive KD and should not preclude IVIG/immunomodulation in late presenting KD with CAA. Giant CAA thrombosis can masquerade as acute gastroenteritis before manifesting as STEMI. Intensification with infliximab-based polytherapy plus aggressive thrombolysis and prolonged LMWH/DOAC-based anticoagulation with dual antiplatelets was associated with favourable coronary recanalization despite persistent giant CAA. Lifelong surveillance is mandatory.

Title -KAWASAKI DISEASE -THE GREAT IMITATOR OF PEDIATRIC INFECTIONS

Authors & Affiliations with Email:

Presenter - Payel Kundu¹, Rhea Singh², Anindita Nandi³, Jigna N Bathia⁴, Priyankar Pal⁵

- 1- Fellow in Pediatric Rheumatology, Pediatric Rheumatology Unit, Institute of Child Health, Kolkata, India, email-dr.payelkundu@gmail.com, phone-8240211780**
- 2- Fellow in Pediatric Rheumatology, Pediatric Rheumatology Unit, Institute of Child Health, Kolkata, India**
- 3- Senior Resident in Pediatric Rheumatology, Pediatric Rheumatology Unit, Institute of Child Health, Kolkata, India**
- 4- Consultant, Pediatric Rheumatology Unit, Institute of Child Healths, Kolkata, India**
- 5- Professor and Head, Department of Pediatrics & Pediatric Rheumatology, Institute of Child Health, Kolkata, India**

Introduction:

Kawasaki disease (KD) is an acute medium-vessel vasculitis and a leading cause of acquired heart disease in children. From a pediatric rheumatology perspective, KD represents a prototypical inflammatory vasculitis with significant overlap with infectious and inflammatory conditions. Its early clinical phenotype is frequently incomplete and dominated by nonspecific systemic inflammation, often mimicking common pediatric infections such as pneumonia, urinary tract infection, or intra-abdominal sepsis. This diagnostic ambiguity, coupled with the evolving nature of mucocutaneous features, poses a significant challenge and may delay timely immunomodulatory therapy, thereby increasing the risk of coronary artery abnormalities (CAA).

Case Series:

We present a series of young children with persistent fever and hyperinflammatory state, initially managed as infectious illnesses. Index presentations included lower respiratory tract infection with worsening radiographic findings, pyuria suggestive of urinary tract infection, mediastinal inflammation mimicking deep neck/mediastinal

sepsis, and acute abdomen with features of pseudo-obstruction. Laboratory evaluation consistently demonstrated markedly elevated acute phase reactants (CRP, ESR), evolving thrombocytosis, and anemia of inflammation. Notably, lack of clinical response to appropriate antimicrobial therapy and persistently elevated inflammatory markers prompted reconsideration of an underlying inflammatory vasculitis. Over time, patients demonstrated evolving mucocutaneous features including non-purulent conjunctivitis, oral mucositis, rash, and extremity changes, fulfilling criteria for complete or incomplete KD. Echocardiography revealed coronary involvement ranging from dilatation to giant aneurysms. Patients were managed with IVIG, aspirin, and escalation to second-line immunomodulation (steroids, infliximab, cyclosporine, anakinra) in refractory cases, with subsequent clinical and biochemical improvement.

Conclusion:

KD remains a “great imitator” within pediatric rheumatology, often presenting as infection-like inflammatory syndromes. Recognition of KD as a dynamic, evolving vasculitis is critical. In any child with persistent fever, elevated inflammatory markers, and poor response to antimicrobials, early rheumatology referral and cardiac evaluation are essential. Timely initiation and escalation of immunomodulatory therapy are pivotal in reducing the risk of coronary artery complications and improving outcomes.

RESURRECTION OF A FORGOTTEN DISEASE : THE MIS-C EPILOGUE- A Five-Case Series

Presenter - Payel Kundu¹, Rhea Singh², Anindita Nandi³, Mittranjan Khan⁴, Priyankar Pal⁵

- 1- Fellow in Pediatric Rheumatology, Pediatric Rheumatology Unit, Institute of Child Health, Kolkata, India, email-dr.payelkundu@gmail.com, phone- 8240211780
- 2- Fellow in Pediatric Rheumatology, Pediatric Rheumatology Unit, Institute of Child Health, Kolkata, India
- 3- Senior Resident in Pediatric Rheumatology, Pediatric Rheumatology Unit, Institute of Child Health, Kolkata, India
- 4- Junior Resident , Institute of Child Health, Kolkata, India
- 5- Professor and Head, Department of Pediatrics & Pediatric Rheumatology, Institute of Child Health, Kolkata, India

Background: Multisystem inflammatory syndrome in children (MIS-C) emerged as a delayed hyperinflammatory complication of SARS-CoV-2 2020-21 pandemic. With the pandemic receding, the MIS-C epitaph was written with hardly any cases reported beyond 2022.. Following a minor resurgence of Covid during June- July 2026 in Kolkata, that caused mild disease in adults, there has been a sudden spurt of MIS-C like illness over the last month. We describe five children presenting with MIS-C-spectrum illness in the recent period, highlighting their diverse clinical and cardiovascular phenotypes.

Methods: We conducted a retrospective descriptive case-series analysis of five children managed at a tertiary pediatric rheumatology centre in 2026. Demographic, clinical, inflammatory, hematological and cardiovascular parameters, initial diagnostic impressions, immunomodulatory treatment and short-term outcomes were reviewed. The fifth child was classified as suspected/probable MIS-C with a Kawasaki-like phenotype in view of incomplete etiological confirmation.

Results: Five children aged 1–7.5 years demonstrated marked clinical heterogeneity. Initial presentations mimicked septic/toxic shock syndrome, Mycoplasma sepsis/myocarditis, Kawasaki disease with shock and refractory Kawasaki-like inflammation. Cardiovascular involvement ranged from cardiogenic shock with left ventricular ejection fraction (LVEF) 39–47%, to marked NT-pro-BNP elevation despite preserved ventricular function, and early diffuse coronary dilatation with subsequent

regression. One infant had prominent neurological involvement with preserved ventricular function and normal coronaries. The Kawasaki-like phenotype demonstrated hypoalbuminemia, hyponatremia, elevated D-dimer and triglycerides, followed by reactive thrombocytosis. SARS-CoV-2 RT-PCR was positive in 1, Covid IgG positive in 2, IgG report awaited in other 2. IVIG with corticosteroid-based immunomodulation, including steroid escalation according to clinical severity, resulted in rapid clinical and biochemical improvement. Ventricular function recovered and coronary abnormalities regressed, with favorable short-term outcomes.

Conclusion: MIS-C continues to present beyond the pandemic and remains a diagnostic chameleon, closely mimicking tropical infections, sepsis and Kawasaki disease. Cardiovascular involvement may range from occult myocardial injury to shock and coronary vasculopathy, and may occur despite preserved LVEF. Persistent fever +/- rash, abdominal symptoms, encephalopathy, with disproportionate hyperinflammation, hematological abnormalities or cardiovascular manifestations should prompt consideration of MIS-C and timely immunotherapy.. Maintaining post-pandemic diagnostic vigilance is essential.

Title:

Proteasome-Associated Autoinflammatory Syndrome Presenting with a Refractory Hepatic Abscess and Incomplete Kawasaki Phenotype

Background:

Proteasome-associated autoinflammatory syndromes (PRAAS) are rare disorders of immune dysregulation characterized by systemic inflammation and heterogeneous clinical manifestations. Their presentation may mimic severe infection or other inflammatory disorders, resulting in diagnostic challenges. We report a child with a persistent hepatic abscess, incomplete Kawasaki disease (KD) phenotype, and a pathogenic PSMB8 variant identified on whole-exome sequencing.

Case Report:

A young boy presented with fever, abdominal pain, poor oral intake and a large right-lobe hepatic abscess. He was clinically very sick and developed generalized edema, ascites and a right-sided pleural effusion. Despite prolonged antimicrobial therapy, there was inadequate clinical and radiological response. During the hospital course, the child continued to have significant systemic inflammation with irritability and cracked lips, and laboratory evaluation revealed features supportive of incomplete KD. However, echocardiography showed no coronary artery abnormalities. The hepatic abscess showed an initial reduction with prolonged antimicrobial therapy, followed by subsequent re-enlargement despite continued treatment. The persistent and recurrent hepatic lesion, lack of satisfactory response to appropriate antimicrobial therapy and accompanying systemic inflammatory/Kawasaki-like phenotype raised suspicion of an underlying immune dysregulatory disorder. Whole-exome sequencing identified a heterozygous pathogenic frameshift variant in PSMB8, c.107_108delCT (p.Pro36Argfs*28), a gene associated with proteasome-associated autoinflammatory syndrome 1. A heterozygous likely pathogenic HBB variant was also identified, which could account for the child's anemia but did not explain the Kawasaki-like phenotype, hepatic abscess or pleural effusion. Given the strong clinical correlation with the PSMB8-associated disorder, further genomic evaluation for a possible second pathogenic allele was recommended.

Conclusion:

This case highlights an unusual presentation of suspected proteasome-associated autoinflammation with a Kawasaki-like phenotype and a refractory hepatic abscess. Persistent or recurrent inflammatory lesions despite prolonged antimicrobial therapy should prompt consideration of an underlying monogenic immune dysregulatory disorder. Recognition of atypical inflammatory presentations and early genomic evaluation may facilitate diagnosis of rare autoinflammatory syndromes and guide further management.

Keywords:

Proteasome-associated autoinflammatory syndrome; PSMB8; hepatic abscess; incomplete Kawasaki disease; autoinflammation; interferonopathy; whole-exome sequencing.

Kawasaki Disease Presenting as Retropharyngeal Phlegmon in Infancy: A Diagnostic Challenge

Background:

Kawasaki disease (KD) is an acute systemic vasculitis of childhood in which early recognition and treatment are essential to prevent coronary artery complications. Although cervical lymphadenopathy is a recognized manifestation, retropharyngeal inflammation mimicking a deep neck infection is an uncommon and diagnostically challenging presentation. Retropharyngeal edema/phlegmon in KD has been increasingly recognized as an abscess-like inflammatory lesion, often without a drainable collection. A retrospective series of 24 children with KD and retropharyngeal edema reported that none fulfilled diagnostic criteria at presentation, all were initially treated with antibiotics without clinical response, and the diagnosis became apparent only with evolution of KD features and supportive inflammatory findings.

Case Description:

We report two infants, aged 2 months and 6–8 months, who presented with approximately one week of fever associated with painful neck swelling and restricted neck movements. The first infant was initially diagnosed with a retropharyngeal abscess, while the second was diagnosed with mumps. Cervical lymphadenopathy was present in the older infant. Neck imaging demonstrated retropharyngeal inflammatory changes with associated airway compression, leading to an initial consideration of deep neck infection. Both infants received empirical antibiotic therapy; however, there was no clinical improvement despite appropriate treatment.

The diagnostic challenge was particularly evident in the 2-month-old infant, in whom classical mucocutaneous manifestations of KD were initially absent. In the older infant, characteristic KD features subsequently evolved, prompting reconsideration of the diagnosis. Both infants had markedly elevated inflammatory markers, while echocardiography showed no coronary artery abnormalities. In view of the persistent fever, inflammatory response, atypical retropharyngeal involvement and evolution of clinical features, KD presenting with

retropharyngeal phlegmon was considered. Both infants were treated with intravenous immunoglobulin and aspirin, with cyclosporine used as additional immunomodulatory therapy. A rapid and marked clinical response was observed following initiation of KD-directed therapy, with resolution of fever and improvement in the neck manifestations.

Conclusion:

Retropharyngeal phlegmon may represent an important but under-recognized manifestation of Kawasaki disease and can closely mimic a bacterial retropharyngeal abscess, particularly during the early phase when classical diagnostic criteria may be incomplete. The combination of persistent fever, painful neck swelling or restricted neck movement, retropharyngeal inflammatory changes and failure to respond to appropriate antibiotics should prompt consideration of KD. Importantly, absence of classical mucocutaneous features or a normal initial echocardiogram does not exclude the diagnosis. Early recognition and timely IVIG therapy may prevent diagnostic delay and unnecessary invasive intervention. Our cases highlight the need to maintain a high index of suspicion for KD even in very young infants presenting with an apparent deep neck infection.

When the Gut Misleads the Heart: Gastrointestinal Pseudo-obstruction as a High-Risk Presentation of Kawasaki Disease—A Three-Case Series

AUTHORS: Presenting author- Dr Rhea Singh¹, Dr Payel Kundu¹, Anindita Nandi¹, Ravali Pratima Goud¹, Jigna Bathia¹, Prof. Priyankar Pal¹

Affiliation- 1. Institute of child health, Kolkata

Background: Kawasaki disease (KD) is classically recognized by prolonged fever with mucocutaneous inflammation; however, prominent gastrointestinal (GI) involvement may dominate the early course, mimic sepsis or a surgical abdomen, and delay definitive therapy. We describe three children with KD presenting with severe GI manifestations, particularly intestinal pseudo-obstruction, highlighting the diagnostic and coronary implications of this phenotype.

Case Series:

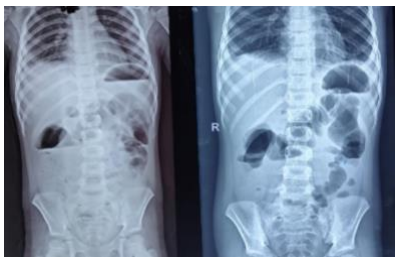
Three children aged 1–3 years 8 months presented with fever and marked GI involvement, including abdominal pain/distension, vomiting or diarrhoea, sluggish bowel sounds and radiological features of intestinal pseudo-obstruction.

Case 1, a 3-year-8-month-old girl, developed severe abdominal pain, diarrhoea, distension, hypotension and oedema and was initially managed as sepsis with paralytic ileus. Investigations showed marked inflammation (CRP 245.9), hypoalbuminaemia, sterile pyuria, gallbladder hydrops and pseudo-obstruction; CT later demonstrated ascites, bilateral pleural effusions and peritonitis-like changes. An early echocardiogram showed normal coronaries. With evolving rash, cervical lymphadenopathy, mucositis, conjunctivitis and periungual peeling, complete KD was recognized on **day 15**. Repeat echocardiography revealed a giant LMCA aneurysm (Z 12.73), with LAD (Z 3.0) and RCA (Z 2.66) involvement. IVIG and aspirin were followed by corticosteroids, infliximab, cyclosporine and enoxaparin; the giant LMCA aneurysm persisted at discharge.

Case 2, a 3-year-old boy presented with fever, extremity oedema, cervical lymphadenopathy, mucocutaneous features and abdominal distension, with marked leukocytosis, thrombocytosis, elevated CRP, hyponatraemia, hypoalbuminaemia and sterile pyuria. KD was **diagnosed on admission** and treated with IVIG, aspirin and methylprednisolone. At 36 hours post-IVIG, fever recrudesced with worsening abdominal distension, ascites and pseudo-obstruction, creating a diagnostic dilemma between intra-abdominal infection and IVIG-resistant KD. Antibiotics, bowel rest and NG decompression were instituted, with surgical evaluation favouring conservative management. Persistent inflammation without an identifiable surgical focus favoured IVIG-resistant KD. Infliximab (10 mg/kg) resulted in defervescence within 12 hours and marked abdominal improvement within 48 hours; follow-up echocardiography was normal.

Case 3, a 1-year-old boy, presented with fever, loose stools, vomiting, abdominal distension, tenderness, sluggish bowel sounds and cervical lymphadenopathy. CRP was 146 mg/L;

mucositis, bilateral non-purulent conjunctivitis and maculopapular rash appeared subsequently. Despite antibiotics and conservative management, fever and abdominal distension persisted. Rising platelets and evolving mucocutaneous features prompted rheumatology review and diagnosis of complete KD. IVIG, aspirin and corticosteroids produced rapid resolution of fever and GI manifestations; repeat echocardiography remained normal.

Feature	Case 1	Case 2	Case 3
Age/Sex	3 y 8 mo/F	3 y/M	1 y/M
Dominant GI phenotype	Severe pain, diarrhoea, distension; pseudo-obstruction; GB hydrops; ascites	Distension, ascites; pseudo-obstruction/acute-abdomen mimic	Vomiting, diarrhoea, distension; sluggish bowel sounds
Initial diagnostic challenge	Sepsis/paralytic ileus	KD → IVIG-resistant KD with acute-abdomen phenotype	GI illness; KD features evolved sequentially
Early echo	Normal coronaries	LAD Z 3.5*	Normal
KD-directed escalation	IVIG → steroids → infliximab + cyclosporine	IVIG + steroids → infliximab	IVIG + aspirin + steroids
Coronary outcome	Giant LMCA aneurysm, Z 12.73	Normal on follow-up	Normal
Key message	Delayed recognition → major coronary morbidity	Pseudo-obstruction may accompany IVIG resistance	Classical KD signs need not appear simultaneously
			

Conclusion: Persistent fever and hyperinflammation without an identifiable infectious or surgical focus should prompt reconsideration of the initial diagnosis, with KD remaining an important differential despite predominant gastrointestinal manifestations. A normal early echocardiogram should not provide false reassurance or delay treatment when the clinical picture supports KD. Careful history-taking is crucial, as characteristic mucocutaneous features may appear sequentially, precede presentation, or resolve before evaluation. Recognizing this evolving phenotype and initiating timely KD-directed therapy may prevent serious coronary complications.

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

AUTHORS: Presenting author- Dr Rhea Singh¹, Dr Payel Kundu¹, Anindita Nandi¹, Ravali Pratima Goud¹, Jigna Bathia¹, Prof. Priyankar Pal¹

Affiliation- 1. Institute of child health, Kolkata

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 10 patients over 17 years with multiple CAA & thrombus aims to compare the pattern of coronary thrombosis before and following the pandemic.

Total KD from 2009 to 2020- 266

(Pre Covid)

Thrombus: 3

Total KD from 2020 to 2025- 190

(Post Covid)

Thrombus: 7

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 14 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.

CASE 3: 2-year 3-month male with KD shock presented on day 5 of fever. Child treated with IVIG and Aspirin, and later Infliximab due to increasing size of LMCA and LAD aneurysm. But 20 days later child develop LAD thrombus. Child treated with LMWH and aspirin. F/U Echo after 2 months later showed resolution of thrombus and regression of aneurysms.

POST COVID PERIOD (2020- present day):

CASE 4: 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. 6 weeks echo showed persistent CAA, with resolution of thrombus.

CASE 5: 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 of clot. Child died of sudden cardiac arrest at age 7 years.

CASE 6: A 9month old boy presented with 11 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 7: 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. Day 9 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.

CASE 8: 9 months infant diagnosed as KD on D11 of fever with multiple CAA, giant aneurysms of LMCA and LAD, with a 4 x 5 mm large thrombus in LMCA and went on to develop a 6 x 5mm thrombus in LAD within 4 days of initiation of therapy with IVIG+

Infliximab. Baby was managed further with Cyclosporine, dual antiplatelets, sc LMWH 2mg/kg. Echo after 10 days showed resolution of both thrombi.

CASE 9: A 1-year infant, with Incomplete KD presented on day 4 of fever, initial echo showing small CAA in LAD with large thrombus (6mm x 2.4mm). Treated with IVIG, steroids, aspirin, Infliximab, LMWH, clopidogrel and cyclosporin. Repeat echo on day 10 showed persistent CAA in LAD but resolution of thrombus.

CASE 10: 3 Year 7 month child with complete KD presented on day 22, with giant CAA in LAD, LMCA and RCA with complete obstruction of LAD with Thrombus with MI. Child treated with dual antiplatelet, Infliximab and cyclosporin, Thrombolysis done, started on statins, diuretics, betablockers and LMWH. Thrombus resolved in 12 days but Giant CAA persists.

Observation:

- Following the pandemic, there has been an increased incidence of KD thrombosis. (1.1% pre-covid 3/266 to **3.7%** post covid- 7/190)
- Post pandemic 5/7 patients had **early onset of thrombus formation within 2 weeks** that sometimes progressed in spite of therapy.
- Post COVID there has been an increase in reports of hypercoagulable states and endothelial vasculopathy. Although this is a very small study, but can we speculate this evidently increased incidence of KD thrombosis to be part of the same spectrum?

Clinical Spectrum and Short-term outcome of Multisystem Inflammatory Syndrome in Children (MIS-C) in Tertiary Care Paediatric Teaching Hospital in Manipur

Presenting author: Dr Melwin Shaji

Contact details: Mobile number- 9495829475

Email ID: melwinshaji99@gmail.com

Authors: Dr. Melwin Shaji¹, Dr. Kh Ratankumar², Dr. Ng Sonamani³, Dr. M Bisheshwor⁴, Dr. Bidyarani Oinam⁵

- 1. Junior Resident, Mother's Care Children Hospital**
- 2. Consultant, Mother's Care Children Hospital**
- 3. Division of Pediatric Hematology Oncology ,Dept of Pediatrics , RIMS , Imphal.**
- 4. Senior Resident, Mother's Care Children Hospital**
- 5. SMO- Pediatrician, DH Bishnupur**

Abstract

Introduction: Multisystem inflammatory syndrome in children (MIS-C) is a severe post-SARS-CoV-2 inflammatory condition with variable clinical manifestations and multisystem involvement.

Objective: To assess the recognition and describe the clinical spectrum, epidemiological and laboratory characteristics, and short-term outcomes of MIS-C in children.

Methods: A retrospective observational study was conducted at Tertiary care paediatric teaching hospital, from June 2021 to December 2023. Children fulfilling the WHO 2020 case definition of MIS-C were included. Demographic, clinical, laboratory, echocardiographic, treatment, and outcome data were analysed.

Results: Fifty-two children were included in 2 ½ years duration, with a median age of 3 years and male predominance (57%). The highest number of cases occurred in 2023, with a summer–monsoon predominance. The median duration of fever at presentation was 4 days and hospital stay was 8 days. Kawasaki disease phenotype was observed in 48%, while shock occurred in 28%. Respiratory and gastrointestinal involvement occurred in 26% and 25%, respectively. Cardiac complications were identified in four children, including coronary artery dilatation in two and myocardial dysfunction in two. Median CRP was 150 mg/L with pro-BNP 1513 pg/mL, D-dimer 2292 ng/mL, and ferritin 268 nmol/L. IVIG was administered

to 90% of patients and steroid to the majority. Six (11%) children required mechanical ventilation, and mortality was 3% (2/52).

Conclusion: MIS-C in this Manipur cohort predominantly affected younger children and demonstrated frequent Kawasaki phenotype and respiratory involvement, with relatively low cardiac complications. Despite favourable overall outcomes, significant morbidity and mortality occurred, highlighting the need for early recognition, timely treatment, and preparedness for MIS-C in the region.

INTRODUCTION:

In May 2020 WHO¹ and CDC² gave international preliminary case definition and Early U.S. surveillance/case identification for Multisystem inflammatory syndrome in children (MIS-C) respectively, updated in 2023 by Council of State and Territorial Epidemiologists (CSTE)/Centres for Disease Control and Prevention (CDC) for surveillance case definition³.

Difference between CDC and WHO were noted as below-

	CDC 2020	WHO 2020
Age	<21 years	0–19 years
Fever	≥38°C for ≥24 h	≥3 days
Hospitalization	Required	Not required
Inflammation	≥1 elevated inflammatory marker	CRP/ESR/procalcitonin
Multisystem involvement	≥2 organ systems	≥2 specified clinical features
SARS-CoV-2	Infection/exposure within 4 weeks	Infection /likely exposure
Exclusion	No alternative diagnosis	No obvious microbial cause

According to IAP guidelines MIS-C is phenotypically classified in 4 groups¹⁵-

Phenotype	Definition
MIS-C without shock	WHO-defined MIS-C, stable, no shock .
MIS-C with shock/MODS	MIS-C with shock and/or ≥2 organ involvement .
MIS-C with Kawasaki phenotype	MIS-C fulfilling complete/incomplete AHA KD criteria .
MIS-C with refractory disease	No response after 48 h of IVIG + low-dose steroids.

Most of case were seen from Western and European countries, A total of 1733 patients with MIS-C were identified⁴ in US during March 2020 to January 2021, Toubiana et al in Paris documented 21 children in 15 days in (April -may) 2020⁵ and Ludwikowska et al in Poland (May 2020- Feb 2021) noted 399 children⁶. Flood et al in UK-216 cases of Paediatric

Multisystem Inflammatory Syndrome temporally associated with SARS-CoV-2 (PIMS-TS)⁷. In Turkey total of 614 patients were enrolled by Yilmaz Ciftoglu et al⁸ as MIS-C cases.

To our surprise Asian countries like China and Japan, places known for birth of COVID-19 and Kawasaki disease respectively report very minimal cases. Japan's nation-wide survey from November 2020 to March 2023, 129 cases were identified as MIS-C with overall incidence rate of 1.5 per 100,000⁹.

As of India significant cases were reported all over the country, in South India Sai et al¹⁰ Tiwari et al¹¹ identified 78 and 41 cases within one year duration respectively, in north – Sharma et al reported 31 in just 6 months¹², Eastern India -Nayak et al included 134 cases during one year study period¹³ and in North east India- Bezboruah et al included 25 cases in one year¹⁴.

In our institute MIS-C was first clinically documented in June 2021 thereafter we noted significant rise in the cases from June 2021 to end of 2023, this motivated our study.

METHODOLOGY:

• Study Design

This is a retrospective observational study in a Tertiary care Pediatric Teaching hospital – Mother's Care Children Hospital and Research Centre Imphal Manipur, using Records from June 2021 to December 2023 (2½ years). Epidemiological, clinical profile and laboratory profile were recorded, 2D-Echocardiography reports using Z-score for calculating coronary artery size as given by Dallaire and colleagues¹⁶ were collected in Microsoft Excel and analyzed.

Research Objectives and Questions

1. To know presence and awareness of the MIS-C in our population
2. To study clinical spectrum and short-term outcome in our cohort.

Inclusion criteria

Children and infants admitted in PICU and general wards eligible for WHO 2020 MIS-C Case Definition¹ were enrolled in our study

Age: 0–19 years + fever >3 days

AND ≥2 of:

- Rash/conjunctivitis/mucocutaneous signs
- Hypotension or shock
- Cardiac involvement
- Coagulopathy
- Acute GI symptoms

AND elevated inflammatory markers (CRP/ESR/procalcitonin)

AND no other microbial cause

AND evidence of SARS-CoV-2 infection or likely COVID-19 contact.

Exclusion criteria

Kawasaki disease was excluded from our study as it may mimic the disease pattern.

- **Ethical Considerations**

Ethical approval was obtained from Research Ethical Committee before collecting retrospective data and analysed.

RESULTS:

Epidemiology and Demography

We enrolled 52 patients in last 2 ½ years, median age of presentation was 3 years, ranged from 4 months to 10 years, maximum cases were in 1-5 year group, male dominance 30/52 (57%) was seen. Seasonal peak in summer- monsoon, Imphal East reported highest -48% among the district, maximum case was seen in year 2023[Fig.1]. Median days of fever on presentation and median days of stay were 4, 8 respectively. Forty-six (88%) required PICU admission.

KD phenotype was seen in 25(48%) however we couldn't further classify as there were overlapping of clinical features. Gastrointestinal symptoms were seen in 13(25%), Respiratory infection in 14 (26%), Meningitis in 4, Dengue in 2 and scrub in one. Shock was noted in 15 (28%), total 6 patients needed mechanical ventilation, HFNC in 3 and O2 support in three.

Cardiac complication was seen in 5, two out them had coronary dilatation and remaining three had myocardial dysfunction, all the children with coronary involvement recovered with a month, one out of three myocardial dysfunctions improved on follow up and we lost remaining two.

On Investigation Median Hb, CRP, TLC, Platelet, Pro BNP, D-dimer and Ferritin were 11mg/dl, 150 mg/L, 12000/cumm, 200000/cumm, 1513 pg/ml, 2292 ng/ml and 268 nmol/L were recorded respectively.

IVIg received in (90%), pulse- Methylprednisolone in 36(69%), oral prednisolone 34 (65%) and Aspirin in 22(42%), had mortality of 2/52(3%)- both patients had acute respiratory distress syndrome (ARDS), Refractory shock, Acute kidney injury (AKI)and myocardial dysfunction. [Table 1].



	(n=52)	Results
Median age (years)		3
Male: female		1.3:1
Age distribution		
Infants		14
1- 5 years		24
>5 years		14
Distribution		
Imphal West		19
Imphal East		11
Ccpur		4
Noney /Thoubal/ Bishnupur / Ukhrul		3 each
Kakching/ non-local		2
Senapati / Kangpokpi		1 each
Seasonal		
Summer- monsoon		21
Spring		14
Autum		11
Winter		6
Yearly distribution		
2021		12
2022		15
2023		25
Median days of fever(days)		4
Median days of stay (days)		8
Classification		
KD- phenotype		25(48%)
Cardiac abnormality		
Coronary dilatation		2
Left Ventricular dysfunction		2
Other systemic involvement		
Respiratory tract infection		14 (26%)
GI symptoms		13 (25%)
Meningitis		4 (7%)
Respiratory Supportive		
Mechanical ventilation		6
HFNC/ O2 support		3
Shock		15 (28%)
Investigation (median)		
Hb	44	11mg/dl
CRP	46	150 mg/L
TLC	44	12000/cumm
Platelet	44	200000/cumm
Pro BNP	43	1513 pg/ml
D-dimer and	46	2292 ng/ml
Ferritin	19	268 nmol/L
Treatment (n/%)		
IVIG		47(90%)
Pulse MP +IVIG		36(69%)
Pulse MP		14 (28%)
Prednisolone		34(65%)
Aspirin		22(52%)
Mortality		2(3%)

Table 1: Clinico- Demographic- laboratory Profile

KD- Kawasaki phenotype; TSS-Toxic shock syndrome; HFNC- High flow nasal cannula; Hb- haemoglobin; CRP-C reactive protein; TLC-Total leucocyte count; Pro-BNP-pro B type natriuretic peptide; IVIG- intravenous immunoglobulin; Pulse MP- methylprednisolone, GI - Gastrointestinal

DISCUSSION:

We Enrolled 52 cases of MIS-C in last 2 ½ years from a single centre tertiary care paediatric teaching hospital as we were aware of the disease since the first case was diagnosed in June 2021.

Our study had male dominance similar to studies from India and parts of the world as authored by Nayak et al¹³ -East India, Bhattad et al -South India¹⁷, Baley et al -US⁴, Ciftoglan et al- Turkey⁸, Flood et al -UK⁷ and Matsubara et al -Japan¹⁸. Median age of presentation was youngest (3years) in our cohort and oldest in US and Japan. Median days of illness/ fever on presentation was almost same in India and Turkey.

Respiratory involvement was most common systemic symptoms seen our cohort as compared to Gastro-intestinal in rest of the studies. Highest neurological and cardiac involvement was reported from UK⁷ and we had the lowest of all. Inflammatory and cardiac markers were significantly elevated in all the studies. We documented high percentage of IVIG used along with Turkish⁸ and highest Aspirin used in our cohort. Mortality was significant in eastern Indian study as they had higher portion with co morbidity, delayed referral, and delayed initiation of immunomodulators and were found to have shock, coma, leukopenia, thrombocytopenia, transaminitis and hypoalbuminemia [Table:2]. We lost 2 patients 3years/female and 4 years/male both from same locality in the month of April 2023, having ARDS, refractory shock, AKI and myocardial dysfunction along with highly elevated inflammatory, cardiac, coagulopathy markers and hemoglobinemia, one of them had multiple blood culture proven sepsis. Possible cause may be delayed referral and severe ARDS.

Limitations of the Study:

Data of a single Paediatric Hospital in Manipur

Studies	Index (n=52)	Nayak et al ¹³ East India 2020-21 (n=134)	Bhattad et al South India ¹⁷ 2020-21 (n=81)	Baley et al US ⁴ 2020-21 (n=1733)	Ciftdogan et al Turkey ⁸ 2020-21 (n=614)	Flood et al UK ⁷ 2020 (n=268)	Matsubara et al Japan ¹⁸ 2020-23 (n=129)
Median/mean (years)	3	7	6.8	9	7.4	8.2	Mean 8.8 ± (3-7)
Male dominance	60%	66.7%	70%	57.6%	57.7%	60%	68.8%
Median/ mean days of fever/illness	4	Mean 5.8	Mean=4.6	-	4	-	-
Clinical feature							
Respiratory	26%	39%	-	-	34.6%	17.9%	-
Gastrointestinal	25%	50%	53%	-	77%	99.3%	89.7%
Neurological	7%	21%	9.8%	-	-	32.5%	-
Shock	28%	28%	29.6%	50%	12%	42.5%	28.7%
Rash	48%	39%	54%	55.6%	54%	52.6%	[KD phenotype-72%]
Mucositis	48%	-	-	-	43%	33.2%	
Conjunctivitis	10%	-	54%	53%	49%	50%	
Cardiac findings							
CAA	3.8%	10.4%	27%	16.5%	9.9%	26.6%	15%
Myocardial dysfunction	3.8%	33.3%	9.8%	31%	-	38%	38.8%
COVID 19 PCR/ Serology							
PCR	100%	100%	75%	82%	94.2%	63.6%	Confirmed by either of the way
Contact			4.9%	51%	9.4%	14.8%	
Investigation							
CRP (mg/L)	Median 150	Median 66	Mean 151		Median 165	Median 223	
ESR (mm/h)	-	50	47		53	-	10.5
TLC (cumm)	12000	13700	14901		8300	-	51
Platelet(L/mm ³)	2	2.3	1.7		1.5	-	12770
Pro-BNP (pg/ml)	1513	-	6072	-	5656	-	-
D-dimer (ng/ml)	2292	-	5978		2787	3400	111
Trop I(ng/L)	-		41		13.3	-	4600
Ferritin (ng/ml)	268	346	-		553	543	10000
Treatment							
IVIG	50(90%)	9 (6.6%)	15.3%	80%	92%		51.1%
Steroid +IVIG	36(69%)	25 (18%)	-	-	-		43%
Steroid	2(3.8%)	88(65%)	11%	71%	58.4%		3.1%
Aspirin	22(52%)	(14.1%)	-	-	-		
Biologic	-	-	TCZ-1 Anakinra - 2	-	-		(2.3% received others)
Duration of Stay	8	8	6 < 5yrs 8 >5yrs	-	-	8	4
Mortality	2(3%)	15(11%)	No	1.4%	6.9%	1.1%	No

Table 2: comparisons of studies
TCZ- Tocilizumab; CAA- coronary artery abnormality;

Conclusion

- Even though most Asian country had lower incidence of MIS-C, India had reported very significant numbers of cases causing morbidity and mortality, acknowledgement and future preparedness of this disease have to be done.
- Our local cohort had enlightened few discrete epidemiological-clinical pattern- Younger median age, more respiratory system involvement, low cardiac complications,

Bibliography

1. Multisystem inflammatory syndrome in children and adolescents temporally related to COVID-19 [Internet]. [cited 2026 Aug 22]. Available from: <https://www.who.int/news-room/commentaries/detail/multisystem-inflammatory-syndrome-in-children-and-adolescents-with-covid-19>
2. CDC. Centers for Disease Control and Prevention [Internet]. 2020 [cited 2026 Aug 19]. Multisystem Inflammatory Syndrome (MIS). Available from: https://archive.cdc.gov/www_cdc_gov/mis/mis-c/hcp/index.html
3. Multisystem Inflammatory Syndrome in Children (MIS-C) Associated with SARS-CoV-2 Infection 2023 Case Definition | CDC [Internet]. 2024 [cited 2026 Aug 19]. Available from: <https://ndc.services.cdc.gov/case-definitions/multisystem-inflammatory-syndrome-in-children-mis-c-2023/>
4. Belay ED, Abrams J, Oster ME, Giovanni J, Pierce T, Meng L, et al. Trends in Geographic and Temporal Distribution of US Children With Multisystem Inflammatory Syndrome During the COVID-19 Pandemic. *JAMA Pediatr.* 2021;175:837–45. doi:10.1001/jamapediatrics.2021.0630
5. Toubiana J, Poirault C, Corsia A, Bajolle F, Fourgeaud J, Angoulvant F, et al. Kawasaki-like multisystem inflammatory syndrome in children during the covid-19 pandemic in Paris, France: prospective observational study. *The BMJ.* 2020;369:m2094. doi:10.1136/bmj.m2094 PubMed PMID: 32493739; PubMed Central PMCID: PMC7500538.
6. Ludwikowska KM, Okarska-Napierała M, Dudek N, Tracewski P, Kusa J, Piwoński KP, et al. Distinct characteristics of multisystem inflammatory syndrome in children in Poland. *Sci Rep.* 2021;11:23562. doi:10.1038/s41598-021-02669-2
7. Flood J, Shingleton J, Bennett E, Walker B, Amin-Chowdhury Z, Oligbu G, et al. Paediatric multisystem inflammatory syndrome temporally associated with SARS-CoV-2 (PIMS-TS): Prospective, national surveillance, United Kingdom and Ireland, 2020. *Lancet Reg Health – Eur.* 2021;3. doi:10.1016/j.lanepe.2021.100075 PubMed PMID: 34027512.
8. Yilmaz Ciftdogan D, Ekemen Keles Y, Cetin BS, Dalgic Karabulut N, Emiroglu M, Bagci Z, et al. COVID-19 associated multisystemic inflammatory syndrome in 614 children with and without overlap with Kawasaki disease-Turk MIS-C study group. *Eur J Pediatr.* 2022;181:2031–43. doi:10.1007/s00431-022-04390-2
9. Nationwide Survey of Multisystem Inflammatory Syndrome in Children Associated with Coronavirus Disease 2019 in Japan | *Journal of Clinical Immunology* | Springer Nature Link

[Internet]. [cited 2026 Aug 22]. Available from: <https://link.springer.com/article/10.1007/s10875-024-01845-z>

10. Sai BVK, Kumar H, Arun Babu T, Chaitra R, Satapathy D, Kalidoss VK. Clinical profile and outcome of multisystem inflammatory syndrome in children (MIS-C) associated with COVID-19 infection: a single-center observational study from South India. *Egypt Pediatr Assoc Gaz*. 2023;71:4. doi:10.1186/s43054-022-00156-5
11. Tiwari A, Balan S, Rauf A, Kappanayil M, Kesavan S, Raj M, et al. COVID-19 related multisystem inflammatory syndrome in children (MIS-C): a hospital-based prospective cohort study from Kerala, India. *BMJ Paediatr Open*. 2021;5. doi:10.1136/bmjpo-2021-001195 PubMed PMID: 10.1136/bmjpo-2021-001195.
12. Sharma R, Sood A, Kumar P. Management of Multisystem Inflammatory Syndrome in Children (MIS-C) Admitted in a Tertiary Care Hospital of North India. *Int J Sci Healthc Res*. 2022;7:99–103. doi:10.52403/ijshr.20220117
13. Nayak S, Panda PC, Biswal B, Agarwalla SK, Satapathy AK, Jena PK, et al. Eastern India Collaboration on Multisystem Inflammatory Syndrome in Children (EICOMISC): A Multicenter Observational Study of 134 Cases. *Front Pediatr*. 2022;10:834039. doi:10.3389/fped.2022.834039 PubMed PMID: 35377583; PubMed Central PMCID: PMC8963178.
14. Bezboruah G, Chakraborty A, Barman D. CLINICAL PROFILE OF CHILDREN WITH MIS-C IN NORTH EAST INDIA : A TERTIARY CARE EXPERIENCE. *Int J Sci Res*. 2023;38–40. doi:10.36106/ijshr/9501348
15. Ayesha. Multisystem Inflammatory Syndrome in Children: IAP Guidelines [Internet]. 2023 [cited 2026 Aug 24]. Available from: <https://medicaldialogues.in/pediatrics-neonatology/guidelines/multisystem-inflammatory-syndrome-in-children-iap-guidelines-109449>
16. Dallaire F, Dahdah N. New equations and a critical appraisal of coronary artery Z scores in healthy children. *J Am Soc Echocardiogr Off Publ Am Soc Echocardiogr*. 2011;24:60–74. doi:10.1016/j.echo.2010.10.004 PubMed PMID: 21074965.
17. Ovid [Internet]. [cited 2026 Aug 23]. Profile of multi-system inflammatory syndrome in... : Pediatric Companion. Available from: https://www.ovid.com/jnls/pedi/fulltext/10.4103/pedc.pedc_4_22~profile-of-multi-system-inflammatory-syndrome-in-children doi:10.4103/pedc.pedc_4_22
18. Bing [Internet]. [cited 2026 Aug 23]. nationwide mis-c survey in japan. Available from: <https://www.bing.com/search?>

Dynamic shifts in peripheral blood neutrophil subsets across clinical timepoints in Kawasaki disease

Authors: Vaishali Thakur¹, Manpreet Dhaliwal¹, Rakesh Kumar Pilania¹, Srishti Sharma¹, Surabhi Mangal¹, Tarun Bhatt¹, Ekta Gupta¹, Man Updesh Singh Sachdeva², Saniya Sharma¹, Jayakanthan Kabeerdoss³, Amit Rawat¹, Anju Gupta¹

Affiliation: ¹Pediatric Allergy and Immunology Unit, Advanced Pediatrics Centre, PGIMER; ²Department of Hematology, PGIMER; ³Pediatric Biochemistry Unit, Advanced Pediatrics Centre, PGIMER

Introduction: Kawasaki disease (KD) is an acute pediatric vasculitis and the leading cause of acquired heart disease in children. Neutrophils play a central role in the inflammatory response, with distinct subsets exhibiting dynamic changes in maturation, activation, and migratory phenotype in peripheral blood. However, these changes across the course of KD remain poorly characterized.

Aims: To characterize peripheral blood neutrophil subsets (mature, immature, aged, activated, degranulating) in KD patients across three clinical timepoints: acute (pre-IVIg), sub-acute (post-IVIg) and convalescent phase using multiparameter flow cytometry.

Methods: Peripheral whole blood was collected from 14 KD patients at three predefined time points: acute phase (before IVIG administration and within 10 days of fever onset), post-IVIg (36–48 hours after completion of IVIG) and convalescent phase (3–4 weeks after fever onset). Among the 14 enrolled patients, 12 presented with incomplete and 2 with complete KD; with 9 responding to IVIg and 5 showing IVIg resistance. The median age was 3 years (IQR, 0.25–8 years), with 8 males and 6 females. Peripheral blood was stained with a customized antibody panel, followed by RBC lysis, washing, and acquisition on a BD LSR III Fortessa, with sequential gating and analysis performed using FlowJo v10. Neutrophils (CD15⁺CD66b⁺) were characterised as immature (CD10⁻CD184⁻), mature (CD10⁺CD184⁻), aged (CD10⁺CD184⁺), activated (CD11b⁺CD62L⁻), degranulated (CD63⁺CD66b⁺) and whole blood (CD177⁺) subsets. UMAP (v4.0) was used for dimensionality reduction and visualization, followed by FlowSOM clustering using FlowJo PluginPlay Bundle v1.1. Group

differences were assessed using the Kruskal–Wallis test followed by pairwise Mann–Whitney U comparisons, with $p < 0.05$ considered statistically significant.

Results: Immunophenotyping of neutrophils across the longitudinal time points in KD patients revealed changes in subset distribution during transition from the acute to convalescent phase. Total neutrophils declined significantly from the acute to convalescent phase (56.1% to 29.1%, $p = 0.033$), with a marked increase in mature neutrophils (1.0% to 8.9%, $p = 0.008$; overall $p = 0.026$) and a subsequent decline in CD177⁺ neutrophils (89.5% to 63.5%, $p = 0.027$). This was accompanied by reduction in immature (27.8% to 4.0%) and degranulated (3.6% to 2.2%) subsets, alongside increases in aged (8.7% to 51.4%) and activated (2.8% to 45.5%) subsets. Unsupervised FlowSOM clustering revealed immature neutrophils decreasing from 16.9% (pre-IVIg) to nearly 0% at convalescence, consistent with resolution of the acute-phase. Mature and aged neutrophil subsets increased steadily from 29.5% to 46.7%.

Conclusion: Neutrophil heterogeneity demonstrated dynamic maturation shifts from acute phase through post-IVIg to convalescence, suggesting distinct subset-specific contributions to inflammatory pathogenesis and disease resolution. Hence, neutrophil maturation profiles may represent potential peripheral biomarkers of disease activity in KD.

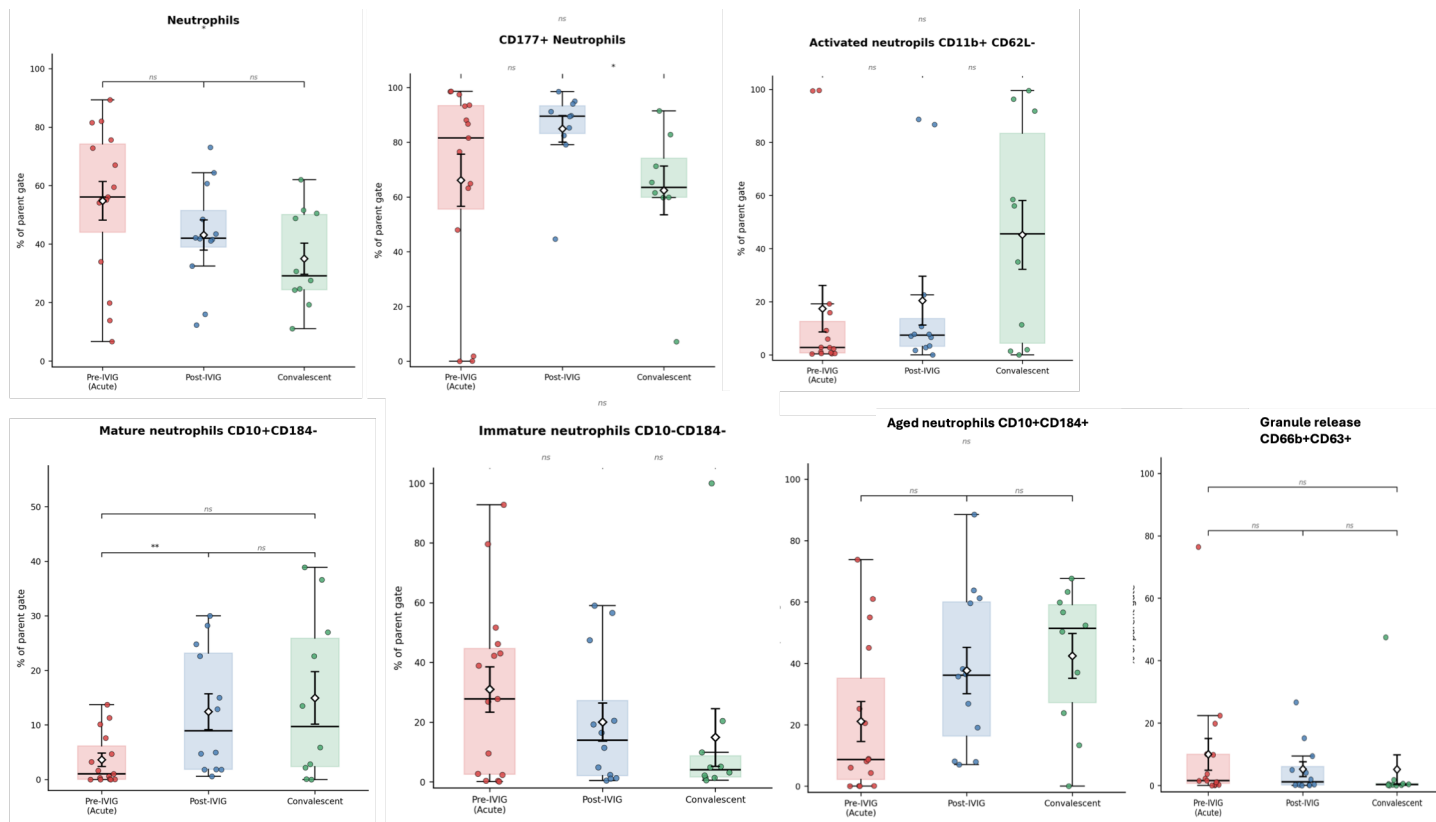


Figure 1: Neutrophil subset frequencies across the clinical course of Kawasaki disease. Frequencies are expressed as percentages of the respective parent population, with neutrophils (CD15⁺CD66b⁺) serving as the parent gate for neutrophil subset analyses. Data are shown for the pre-IVIG (acute), post-IVIG and convalescent phases.

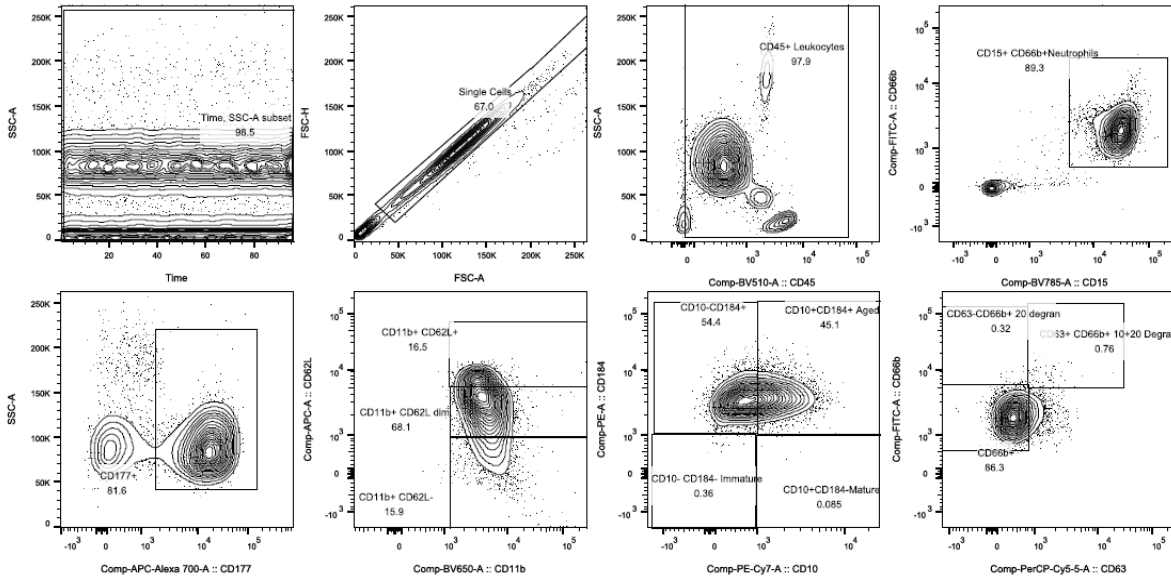


Figure 2: Gating hierarchy of a representative case showing neutrophils and subsets (maturation, activation, migratory state, granule release): Singlets were first identified using forward scatter area versus height (FSC-A vs FSC-H) parameters, followed by gating of CD45⁺ cells to define the leukocyte population. (Figure 2). Neutrophils were gated as CD66b⁺CD15⁺ cells to define immature (CD10⁻CD184⁻), mature (CD10⁺CD184⁻), aged (CD10⁺CD184⁺), activated neutrophils (CD11b⁺CD62L⁻), and granule release (CD66b⁺CD63⁺).

T- and NK-Cell Remodelling in Pediatric Kawasaki Disease

Srishti Sharma¹, Rakesh Kumar Pilonia¹, Vaishali Thakur¹, Surabhi Mangal¹, Tarun Bhatt¹, Manpreet Dhaliwal¹, Jayakanthan Kabeerdoss², Biman Saikia³, Saniya Sharma¹, Anju Gupa¹, Amit Rawat¹

Affiliation

¹Allergy Immunology Unit, Advanced Pediatrics Centre, PGIMER, Chandigarh

²Pediatric Biochemistry Unit, Advanced Pediatrics Centre, PGIMER, Chandigarh

³Department of Immunopathology, PGIMER, Chandigarh

Introduction: Kawasaki disease (KD) is an acute vasculitis that mainly affects children. Previous studies have highlighted the role of T and NK cells in immune response against KD. However, longitudinal data characterizing T and NK subsets in acute and sub-acute stages remains sparse.

Aim: To characterize the changes in peripheral T- and NK-cell subsets at acute (treatment naïve) and sub-acute (36 to 48 hours post-IVIg) stages in a pediatric KD cohort.

Methods: Whole blood samples were collected from KD patients (n=10) at acute and sub-acute phase. Samples were immunophenotyped across two flow cytometry panels capturing T-helper-CD4⁺ and T-cytotoxic-CD8⁺ cells and their differentiation subsets (naïve, central memory [TCM], effector memory [TEM], TEMRA), helper subsets (Th1, Th2, Th17) and activation/senescence markers (CD38⁺, PD-1/CD279⁺). Additionally, regulatory T cells (CD25^{high}, CD127^{low}), NK cells (CD3⁻, CD16⁺, CD56⁺) and their maturation and differentiation subsets based on CD56^{bright} CD16^{dim}, CD16^{bright} CD56^{dim}, CD57 and CD159c/NKG2C expression were also characterized. Pre- versus post-IVIg comparisons were performed using the Wilcoxon signed-rank test, with significance set at $p < 0.05$.

Results: Total T lymphocytes increased from acute to sub-acute phase (median 39.1% to 46.4%), accompanied by a significant decline in the naïve CD4⁺ (70.1% to 58.0%, $p < 0.05$) and naïve CD8⁺ (48.2% to 32.3%) subsets. A decline in CD4⁺CD38⁺ activated T cells (83.4% to 72.6%, $p < 0.05$) along with an increase in CD8⁺PD-1(CD279)⁺ expression (77.9% to 93.3%, $p < 0.05$) was also observed. Regulatory T cells showed an increase from 7.8% to 8.8% post treatment. The T-helper subsets, Th1 and Th2 slightly declined post-IVIg, while surface marker-defined (CCR6⁺CXCR3⁻CCR4⁺) Th17 frequency showed no significant change.

NK cell subset analysis showed an increase in total NK-cell frequency (median 4.81% to 6.69%), accompanied by increase in mature $CD16^{\text{bright}} CD56^{\text{dim}}$ (91.7% to 93.7%), $CD56^{\text{bright}} CD16^{\text{dim}}$ (3.53% to 5.85%), and terminally differentiated NK-cell populations (20.5% to 30.25%). Sub-group analysis further showed that IVIg-resistant KD patients had lower total and mature $CD16^{\text{bright}} CD56^{\text{dim}}$ NK-cell frequencies at baseline than responders. Also, both populations increased post-IVIg in the resistant group.

Conclusion: IVIg treatment induced remodelling of T- and NK-cell compartments in KD patients, marked by reduced naïve T cells, altered T-cell activation, and increased mature NK-cell populations.

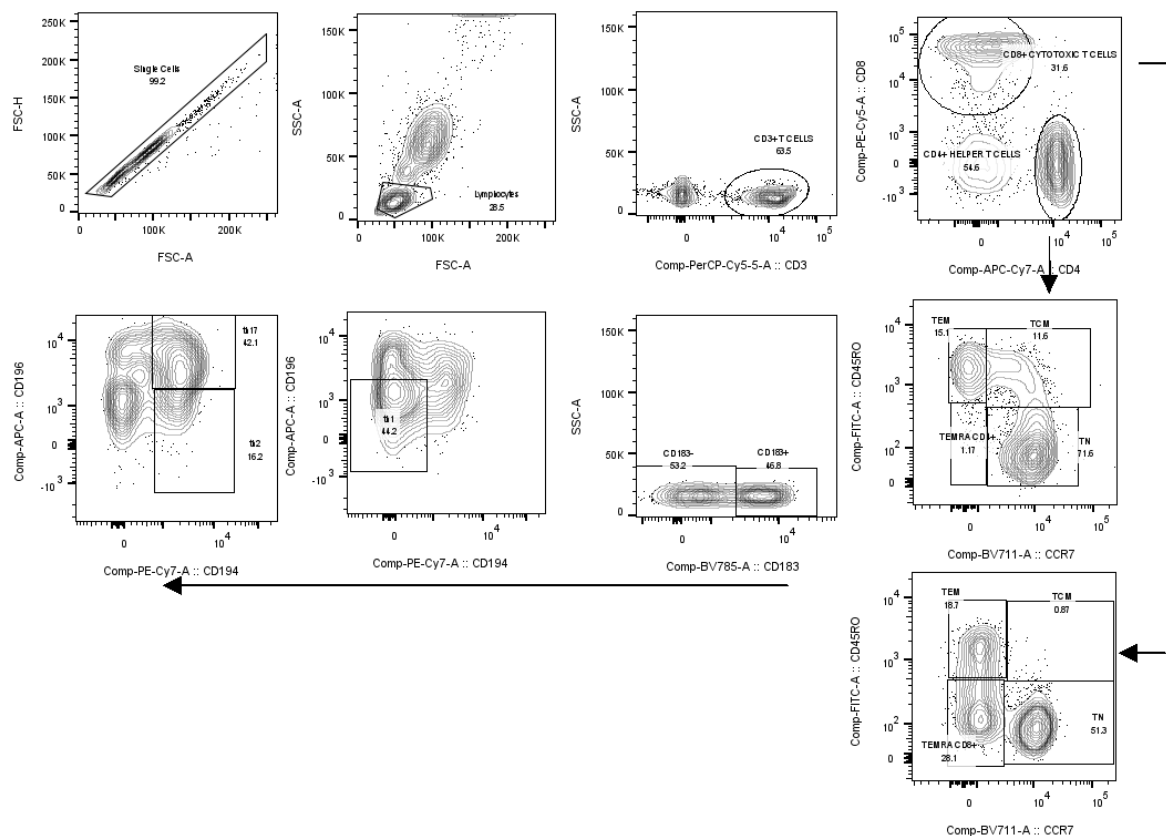


Figure 1. Gating strategy of a representative case showing T-cell differentiation and T-helper cell subsets. Following singlet and lymphocyte gating, $CD3^+$ T cells were identified and separated into $CD4^+$ helper and $CD8^+$ cytotoxic T-cell populations. T-cell differentiation was assessed using $CD45RA$ and $CD62L$ expression to delineate naïve (TN), central memory (TCM), effector memory (TEM), and terminally differentiated effector memory (TEMRA) populations within $CD4^+$ and $CD8^+$ T cells. T-helper subsets were subsequently characterized

using CCR6 and CXCR3/CCR4 expression to identify Th1, Th2, and surface marker-defined Th17 populations.

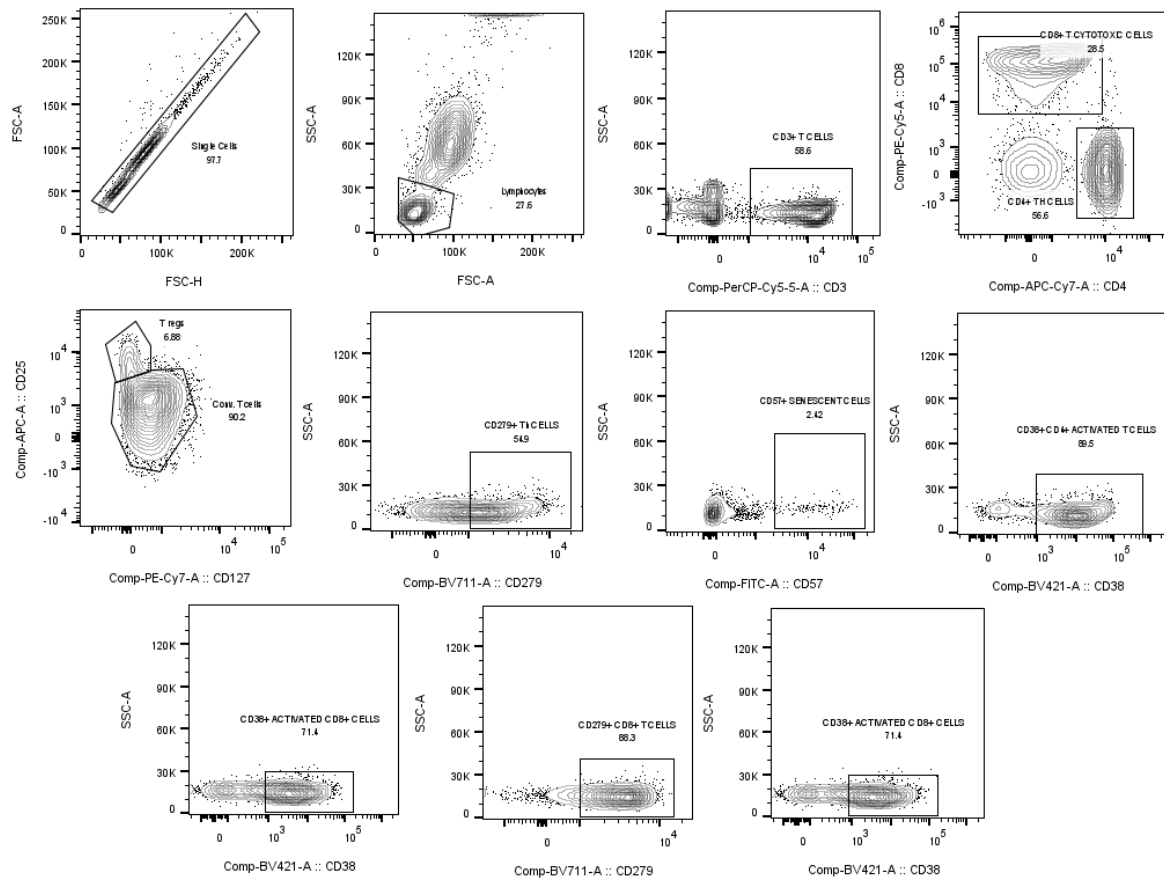


Figure 2. Gating strategy of a representative case showing T-cell subsets, activation, senescence, and regulatory phenotypes. Singlets were first identified using FSC-A versus FSC-H, followed by lymphocyte gating based on FSC-A versus SSC-A plot. CD3⁺ cells were subsequently identified and separated into CD4⁺ helper and CD8⁺ cytotoxic T-cell populations. Within the CD4⁺ compartment, regulatory T cells were identified based on CD25 and CD127 expression. T-cell activation and differentiation states were further characterized using CD38, CD279/PD-1, and CD57 expression on CD4⁺ and CD8⁺ T-cell populations.

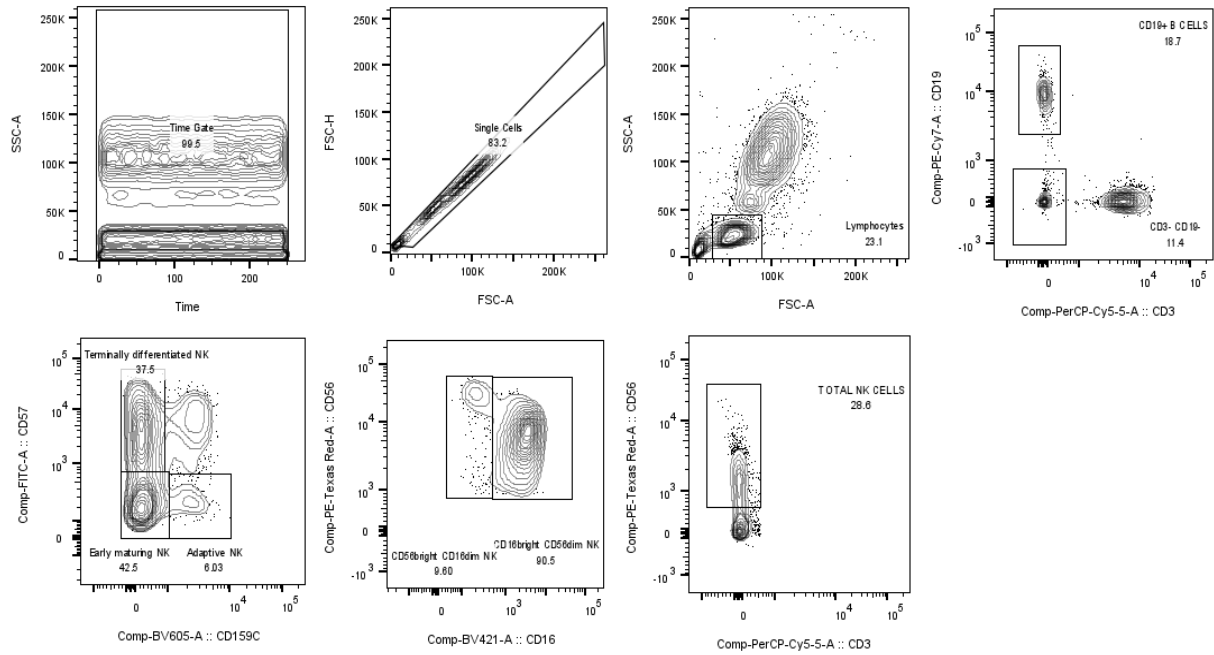


Figure 3. Gating strategy of a representative case showing NK-cell populations and differentiation states. Singlets were first identified using FSC-A vs FSC-H, followed by lymphocyte gating based on FSC-A versus SSC-A plot. Within the lymphocyte population, B cells were identified as CD19⁺, while CD3⁻CD19⁻ cells were used to define the NK-cell compartment. Total NK cells were further characterized using CD16 and CD56 expression to identify CD16^{bright} CD56^{dim} and CD56^{bright} CD16^{dim} subsets. NK-cell differentiation was assessed using CD57 and CD159c/NKG2C expression to define early-maturing, adaptive, and terminally differentiated NK-cell populations.

Kawasaki Disease in Infancy: The Continuing Enigma—A Case Series

Ashish Kumar, MBBS; Ajay Sharma, MD; Sandesh Guleria, DM, Avinash Sharma, DM
Dr Rajendra Prasad Government Medical College Kangra at Tanda, HP

Introduction:

Kawasaki disease (KD) is an acute childhood medium vessel vasculitis with a predilection for young children (less than 5 yr). Delayed recognition may increase the risk of coronary artery abnormalities (CAAs)^{1,2}. KD is now the leading cause of acquired heart disease in most developed countries and India as well.^{1,2}

Case 1: 8-month-old boy with 7 days of fever, irritability and diarrhea was initially treated for UTI/meningitis. Persistent fever, irritability, rash over bilateral hands and recalled conjunctival injection raised suspicion of incomplete Kawasaki disease (KD). Laboratory findings (Table 1) showed anaemia, neutrophilic leukocytosis and elevated inflammatory markers. ECHO revealed a medium LMCA aneurysm (4 mm; z-score 5.3). He received IVIG, aspirin and infliximab. LMCA at day 4 progressed to 5 mm (z-score 8.01), but regressed to 3 mm (z-score 3.65) at 4 weeks.

Case 2: 4-month-old girl presented with 6 days of high-grade fever and irritability, initially treated for sepsis. Rash, conjunctival injection, cracked lips, strawberry tongue, extremity edema and BCG scar reactivation suggested KD. Laboratory findings (Table 1) showed anaemia, neutrophilic leukocytosis, thrombocytosis and elevated ESR/CRP. ECHO revealed LMCA and LAD aneurysms (4 mm each; z-scores 6.41 and 12.81). She received IVIG, aspirin and infliximab, followed by anticoagulation. At 1 week, LMCA regressed to 2 mm, while LAD measured 5mm (z-score 17.67), necessitating continued antithrombotic therapy.

Case 3: 13-month-old girl presented with 2 weeks of high-grade fever, rash, extreme irritability, strawberry tongue and periungual desquamation, with elevated inflammatory markers (Table 1). Infection workup was negative and she was treated as a case of incomplete KD, with IVIG and aspirin; ECHO showed no coronary abnormality and fever subsided. Two months later, she presented with high-grade fever for 16 days, associated with lymphadenopathy and hepatosplenomegaly. Severe anaemia, thrombocytosis and markedly elevated ESR/CRP and ferritin were noted. Bone marrow and lymph node evaluation excluded malignancy. Development of knee arthritis led to consideration of systemic JIA, with improvement on naproxen, prednisolone and methotrexate.

Conclusion:

KD in infancy frequently occurs in incomplete/atypical form. Presentations like seizures, nephritis, meningitis and shock may be seen in infancy^{3,4}. Clinical clues including BCG scar reactivation, and early ECHO assessment are essential in infants with prolonged unexplained fever. Reactivation of BCG scar though does not find a place in diagnostic criteria, is nearly pathognomonic of KD⁵. Despite initiation of treatment in first 10 days of disease onset, infants with KD are more likely to develop CAAs. Study by Singh et al. on infants <6 months of age showed CAAs in 35% of the patients on ECHO³. Early recognition and timely treatment remain crucial to reduce coronary complications^{1,2}. Refractoriness to treatment was also significantly more in infants⁶. Thus, to reduce CAAs adjunctive therapies should target this age group.

KD and sJIA share overlapping features. Go et al. reported KD preceding sJIA in 7%. Arthritis most commonly observed in knee joint, followed by the ankle and wrist with equal frequency⁷. In both disease entities, fever is a major criteria. KD fever is usually self-limiting, whereas sJIA fever may be chronic or recurrent⁸. KD-associated arthritis is usually self-limiting within 2 weeks of IVIG⁹.

References:

1. Newburger JW, Takahashi M, Burns JC. Kawasaki Disease. J Am Coll Cardiol. 2016;67:1738–49.
2. Singh S, Kawasaki T. Kawasaki Disease in India, Lessons Learnt Over the Last 20 Years. Indian Pediatr. 2016;53:119–24.
3. Singh S, Agarwal S, Bhattad S, Gupta A, Suri D, Rawat A, et al. Kawasaki disease in infants below 6 months: a clinical conundrum? Int J Rheum Dis. 2016;19:924–8.

4. Burns JC, Wiggins JW, Toews WH, Newburger JW, Leung DY, Wilson H, et al. Clinical spectrum of Kawasaki disease in infants younger than 6 months of age. *J Pediatr*. 1986;109:759–63.
5. Rezai MS, Shahmohammadi S. Erythema at BCG Inoculation Site in Kawasaki Disease Patients. *Mater Socio-Medica*. 2014;26:256–60.
6. Bhattad S, Gupta S, Israni N, Mohanty S. Profile of Kawasaki disease at a tertiary care center in India. *Ann Pediatr Cardiol*. 2021;14:187–91.
7. Go E, van Veenendaal M, Manlhiot C, Schneider R, McCrindle BW, Yeung RSM. Kawasaki Disease and Systemic Juvenile Idiopathic Arthritis - Two Ends of the Same Spectrum. *Front Pediatr*. 2021;9:665815.
8. Behrens EM, Beukelman T, Gallo L, Spangler J, Rosenkranz M, Arkachaisri T, et al. Evaluation of the presentation of systemic onset juvenile rheumatoid arthritis: data from the Pennsylvania Systemic Onset Juvenile Arthritis Registry (PASOJAR). *J Rheumatol*. 2008;35:343–8.
9. Guleria S, Pilania RK, Jindal AK, Singh A, Vignesh P, Suri D, et al. Clinico-laboratory profile of Kawasaki disease with arthritis in children. *Eur J Pediatr*. 2020;179:875–9.

Table 1: Haematological investigations for 3 cases

Investigations	Case I	Case II	Case III
Hb (gm/L)	91	85	90
TLC (x10 ⁹ /L)	37.9	22.2	24.1
DLC	P ₄₉ L ₃₃ M ₁₀ E ₈	P ₆₀ L ₃₆ M ₂ E ₂	P ₅₄ L ₃₄ M ₈ E ₄
Platelets (x10 ⁹ /L)	394	467	700
ESR (mm in 1 st hour)	36	110	80
CRP (mg/L)	155	85	182

Figure 1: BCG scar reactivation



Clinical Profile and Outcome of 36 children with Kawasaki Disease treated with Infliximab - Experience from a Tertiary Care Center in Southern India

Jayanth B Nair¹, Uma Tejaswi¹, Bharath A P², Chetan Ginigeri³, Harish Kumar³, Syed Mohammed Naushad Ali³, Karthik Arigela³, Sagar Bhattad¹

Affiliation:

1Division of Pediatric Immunology, Rheumatology and BMT, Manipal hospital Yelahanka, Bangalore, India.

2Department of Pediatric Cardiology, Manipal hospital Yelahanka, Bangalore, India.

3Department of Pediatrics, Manipal hospital Yelahanka, Bangalore, India.

Introduction:

Infliximab, a monoclonal anti-TNF α , has emerged as a potential therapeutic option in Kawasaki disease (KD) particularly in cases resistant to conventional intravenous immunoglobulin therapy.

Objective:

To evaluate the clinical profile and treatment outcomes of children with KD who received Infliximab at our centre.

Methods:

A retrospective review of the clinical records of patients diagnosed with KD diagnosed between February 2017 and July 2026 was done. A total of 150 children were diagnosed with KD during the study period, of whom 36 received Infliximab IV 10 mg/kg. The clinical profile, treatment indications, response to therapy, and outcomes of these 36 patients were reviewed and analysed in detail.

Results

Table 1. Characteristics of patients with Kawasaki disease treated with Infliximab

Total number of Kawasaki disease cases	150
Treated with Infliximab (N)	36(24%)
Mean age	28 months (3-132)
Infants	16 (44.4%)
Male	21 (58.3%)
Female	15(41.6%)
Clinical features	
Oral mucosal changes	33 (91.7%)
Rash	28 (79.2%)
Conjunctival injection	24 (66.7%)
Peripheral edema	21 (58.3%)
Cervical lymphadenopathy	16 (45.8%)
Peri-ungual desquamation	11 (30 %)
Complete KD	19(52.7%)
Incomplete KD	17(47.2%)

Laboratory features	
Anaemia	14 (58.3%)
Leukocytosis	21 (87.5 %)
Thrombocytosis	20 (83.3%)
Echocardiographic changes	33 (91.6%)
Coronary Artery Aneurysm(CAA)	33 (91.6%)
Small CAA	10 (27.7%)
Medium CAA	16 (44.4%)
Giant CAA	7 (19.4%)
Severe Aortic Regurgitation	2 (5.5 %)
LAD involvement	28 (77.7%)
LMCA involvement	25 (69.4%)
RCA involvement	17 (47.2%)
LCx involvement	11 (30.5%)
Treatment	
Dual antiplatelets	23 (63.8%)
LMWH/Apixaban	7 (19.4 %)
Cyclosporine	6 (16.6%)
Anakinra	2 (5.5%)
Resolution of CAA (N=33)	
At end of 6 weeks	13 (39.3%)
6 weeks to 1 year	14 (42%)
Persisting CAA in last follow up (1 month to 6 year)	6 (18.1%)

All 36 children included in the study received standard therapy with intravenous immunoglobulin (IVIG) and corticosteroids. Infliximab was administered in view of refractory disease in 19 (52.7%) of patients, while 17 (47.2%) received it as part of primary treatment intensification. The mean time to achieve an afebrile state following Infliximab therapy was 5 days (Range 1 - 10 days). Two patients continued to have fever despite Infliximab and required additional immunomodulatory agents including Cyclosporine and Anakinra. The mean duration of hospital stay in the cohort was 6 days. Among those with coronary involvement, the average time for normalization was 10 weeks. Two out of seven having giant CAA showed complete normalisation of coronaries at 1 year follow-up.

Conclusion: This study highlights the clinical utility of Infliximab in managing refractory and high-risk cases of KD, particularly in achieving defervescence and coronary stabilization. However, the findings are limited by the small sample size and absence of a comparative control group.

Use of Anakinra in Severe and Refractory Kawasaki disease: Our experience from a tertiary care centre in South India

Authors:

Jayanth B Nair¹, Uma Tejaswi¹, Bharath A P², Chetan Ginigeri³, Harish Kumar³, Syed Mohammed Naushad Ali³, Karthik Arigela³, Sagar Bhattad¹

Affiliations:

¹ Division of Pediatric Immunology, Rheumatology and Bone Marrow Transplant, Manipal Hospital Yelahanka, Bangalore, India

² Department of Pediatric Cardiology, Manipal Hospital Yelahanka, Bangalore, Bangalore, India

³ Department of Pediatrics, Manipal Hospital Yelahanka, Bangalore, Bangalore, India

Introduction:

Kawasaki disease (KD) is a systemic vasculitis of childhood with coronary artery involvement being its major complication. Despite intravenous immunoglobulin (IVIG) and corticosteroids, a subset of children develop refractory disease, persistent inflammation, cardiovascular dysfunction or coronary artery aneurysms. Interleukin-1-mediated inflammation has been implicated in KD pathogenesis, and Anakinra, an IL-1 receptor antagonist, has emerged as a potential rescue therapy. However, evidence from clinical practice remains limited.

Aim

To evaluate the clinical profile and treatment outcomes of children with Kawasaki disease who received Anakinra at our center.

Methods

A retrospective review of the clinical records of patients diagnosed with Kawasaki disease between February 2017 and July 2026 was performed. During the study period, 150 children were diagnosed with Kawasaki disease, of whom three received Anakinra. The clinical profile, treatment indications, response to therapy, and outcomes of these three patients were reviewed and analysed in detail.

Results:

Three of 150 children with KD (2.0%) received Anakinra. M:F ratio was 1:2 and the median age at diagnosis was 65 months (range 8–84 months). The mean duration of fever at presentation was 10 days (range 6–13 days). All patients had received IVIG and methylprednisolone prior to Anakinra; two had additionally received Infliximab and Cyclosporine. Anakinra was administered for KD shock syndrome (n=1) and refractory disease (n=2).

Anakinra was administered intravenously at a dose of 5 mg/kg/day for 7 days in P1, 5 mg/kg/day for 2 days in P2 and 7 mg/kg/day intravenously for 5 days followed by subcutaneous therapy for 5 weeks in P3. The mean time to defervescence following the initiation of Anakinra was 1 day. Mean CRP decreased from 136.7 mg/L before Anakinra to 29.0 mg/L after treatment, representing a mean reduction of 78.8%. No adverse effects attributable to Anakinra were observed.

Cardiac involvement was present in all three patients. P1 had myocardial dysfunction without coronary artery abnormalities. P2 and P3 had coronary artery abnormalities: P2 had moderate aneurysm of the LAD and LMCA, while P3 had a giant LMCA aneurysm with small LAD and LCx aneurysms. At one year follow up, P1 and P3 had normal cardiac findings, while P2 had persistent fusiform LMCA dilatation at one year follow-up.

Conclusion:

In this single-center experience, Anakinra was used in 2% of children with KD, predominantly as rescue therapy for severe or highly refractory disease following multiple immunomodulatory treatments. A marked reduction in CRP and subsequent favourable cardiac evolution was observed, with normalisation of echocardiographic findings in two of three patients. No Anakinra related adverse effects were observed. These findings are consistent with published small cohorts suggesting potential benefits and favorable short-term safety of IL-1 blockade in refractory KD. The major limitations are the small sample size and absence of a comparison group. Larger prospective studies are required to establish the optimal timing, dose, and duration of Anakinra in KD.

A cross-cohort machine-learning blood signature discriminates Kawasaki disease from other febrile illnesses

Prabavathi Devarajalu¹, Vaishali Thakur², Anmol Gurgela¹, Manpreet Dhaliwal², Rakesh Kumar Pilania², Jayakanthan Kabeerdoss^{1*}

1 Biochemistry Unit, Department of Pediatrics, Postgraduate Institute of Medical Education and Research (PGIMER), Chandigarh

2 Allergy and Immunology Unit, Department of Pediatrics, Postgraduate Institute of Medical Education and Research (PGIMER), Chandigarh

Background. Distinguishing Kawasaki disease (KD) from other febrile illnesses in children remains a major diagnostic challenge because there is no disease-specific diagnostic test. Although transcriptomic classifiers have shown promise, models developed in individual datasets often have limited reproducibility across independent cohorts. We therefore used a cross-cohort machine-learning approach designed to identify transcriptomic features that remain stable across cohorts and platforms.

Methods. We analysed three whole-blood transcriptomic cohorts (GSE68004 and GSE73461, microarray; GSE178491, RNA-seq), comprising 275 children with KD and 353 febrile, infectious, or healthy controls, with 18,486 genes shared across datasets. Data were harmonised to a common gene-symbol space and standardised within each cohort. Candidate genes were selected based on consistent direction of effect across training cohorts and ranked according to their weakest cross-cohort effect size. An L2-regularised logistic regression model was evaluated using leave-one-dataset-out (LODO) validation. The final locked panel was then tested in two independent cohorts.

Results. A 15-gene concordance-stable panel achieved ROC-AUC values of 0.86–0.89 in LODO validation (mean, 0.88). A reduced six-gene panel comprising PDGFC, DIRC2, IER3, KLHL2, SLC12A9, and ZNF185 showed similar performance (mean AUC, 0.88) and replicated in GSE73464, distinguishing KD from febrile controls (AUC, 0.80). In GSE63881, the panel also distinguished acute from convalescent KD (AUC, 0.91), suggesting that it captures disease-related transcriptional changes beyond simple case-control discrimination. The six-gene panel outperformed canonical KD-associated neutrophil markers (AUC, 0.76), a single-cohort-derived panel (AUC, 0.80), and a random-panel null model (AUC, 0.67). Performance transferred across microarray and RNA-seq platforms, although assay-specific calibration was required for threshold-based classification. None of the six genes had a curated KD association in Open Targets, supporting the potential novelty of the signature.

Conclusions. A robustness-focused, cross-cohort machine-learning strategy identified a compact six-gene blood signature that maintained diagnostic performance

across independent cohorts, disease comparators, and transcriptomic platforms. The findings support prospective validation using targeted assays such as qPCR and further evaluation of the signature for clinical diagnostic and disease-monitoring applications.

(Category: Original research)

Title: Safety and efficacy of Direct Oral Anticoagulants in patients with Kawasaki Disease Associated Giant Coronary Aneurysms: A Prospective Pediatric Study

Authors: *Dr Renaisa Banik (2nd Year PGT, IPGMER and SSKM hospital, Kolkata)*

Dr Suprit Basu (Asst professor, IPGMER and SSKM hospital, Kolkata)

Introduction:

Kawasaki Disease (KD) is a multisystem vasculitis that predominantly affects medium-sized vessels, particularly the coronary arteries. Described initially in Japan, KD has become the leading cause of acquired heart disease among children, surpassing rheumatic fever. It is most common in children under 5 years of age and is characterized by high fever, mucocutaneous inflammation, and lymphadenopathy.

The most feared complication of KD is the development of coronary artery aneurysms (CAAs), which occur in approximately 15–25% of untreated cases. With intravenous immunoglobulin (IVIG) treatment, the incidence drops to 3–5%. Among CAAs, giant coronary aneurysms (GCAAs) represent the most severe form, defined by a diameter ≥ 8 mm or a z-score ≥ 10 . These are associated with a substantially higher risk of thrombosis, myocardial infarction, ischemia, and sudden cardiac death. As such, the management of GCAAs involves aggressive antiplatelet and anticoagulation therapy.

Aims:

To evaluate the efficacy and safety of DOACs in preventing thrombotic complications in children with giant coronary aneurysms due to Kawasaki disease.

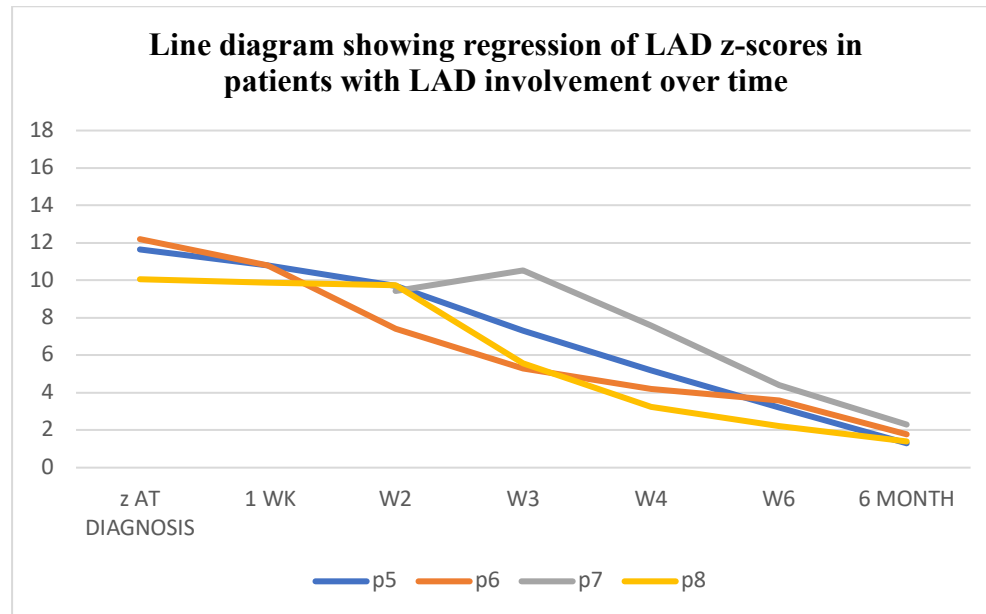
Methods:

This prospective observational study enrolled children aged <18 years with Kawasaki disease and giant coronary artery aneurysms (diameter ≥ 8 mm or Z-score ≥ 10), after obtaining requisite institutional ethical clearance. They were initiated on direct oral anticoagulant (Rivaroxaban). Patients were followed for 6 months with serial echocardiography to assess aneurysm size, thrombus formation, bleeding events, and thrombotic outcomes.

Results:

Nine children with Kawasaki disease associated giant coronary artery aneurysms were enrolled. Male: Female ratio 8:1. The age at presentation ranged from **4 months to 11 years, median age being 25 months**. **5/9(55%)** children presented with Incomplete KD, while **4/9(44%)** presented with classical KD. One child (**11.1%**) presented with acute cardiac manifestations. Coronary artery involvement included the left anterior descending artery in **5/9 (55.5%)**, left main coronary artery in **4/9 (44.4%)**, right coronary artery in **4/9 (44.4%)**, and multivessel involvement in **4/9 (44.4%)**. Median delay in diagnosis was **9 days**. Nine children received rivaroxaban in addition to standard antiplatelet therapy and underwent serial echocardiographic follow-up. Serial assessment demonstrated progressive regression in coronary artery aneurysm dimensions, with the greatest reduction observed during the first six weeks and sustained improvement at six months. Median duration of follow up was **12.86** months. The median maximum coronary artery Z-scores for respective vessels showed a marked decline from baseline to six-month follow-up, with most aneurysms regressing from the giant to the medium or small aneurysm category. No patient demonstrated progression of coronary dilatation during follow-up. One patient had coronary thrombosis at baseline, which

demonstrated complete/partial resolution on serial echocardiography during rivaroxaban therapy. No new thrombotic events occurred during follow-up. Following initiation of DOAC therapy, there were **no thrombotic events, myocardial infarction, ischemic stroke, major bleeding, clinically relevant non-major bleeding, or anticoagulant discontinuation due to adverse effects**, yielding an event-free rate of **100%** for both thrombotic and major bleeding outcomes.



Conclusion: Direct oral anticoagulants were well tolerated in this prospective paediatric cohort and were associated with favourable short-term safety and clinical outcomes, including no new thrombotic events after treatment initiation and regression of coronary artery aneurysms on serial echocardiography. However, these findings are limited by the small sample size, single-centre design, and lack of a comparator group, warranting validation in larger multicentre studies.

Title: Beyond injections: Apixaban in Kawasaki disease with Giant aneurysm.

Authors: Dr Dinesh Narayanan¹, Dr Nameirakpam Johnson²

¹Post Graduate Trainee, Department of Paediatrics, JNIMS, Imphal

²Consultant Paediatric Rheumatologist, Department of Paediatrics, JNIMS, Imphal

Objective:

To describe the tolerance and effect of apixaban in treatment and outcome of children with Kawasaki disease associated with giant coronary artery aneurysm (GCAA).

Results:

A total of 4 children with Kawasaki disease (KD) associated with GCAA were included in this case series. The median age of diagnosis was 1.7 (IQR- 0.75-2.75) years, with a male to female ratio of 3:1. 3 had incomplete KD and 1 had complete KD. All the children were started with IVIG initially followed by oral prednisolone, infliximab and cyclosporine. All 4 received enoxaparin initially for an average duration of 3.5 (IQR= 2.5-5) months and later switched to apixaban due to poor compliance and financial reasons. All children were started and continued on aspirin and atorvastatin, except 1 children in whom statin was stopped due to recurrent diarrhea. Follow-up duration ranged from 6 month to 2 years with serial echocardiography. 3 children showed regression from GCAA to medium sized aneurysm, while one continued to have gaint aneurysm despite therapy. No major bleeding complications were reported during follow-up.

Conclusion:

This 4 patient case series highlights the role of apixaban as alternative oral anticoagulant in KD with GCAA. Apixaban is feasible and well tolerated with no major bleeding or thrombotic complications during follow-up.

Correspondence: Dr. Nameirakpam Johnson, Consultant Paediatric Rheumatologist, Department of Paediatrics, JNIMS, Imphal