



KAWASAKI DISEASE

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DISCLOSURE STATEMENT

I have no financial disclosures or conflicts of interest.

OBJECTIVES

- Definition
- Signs & Symptoms
- Epidemiology
- Etiology & Risk Factors
- Atypical Presentation
- Diagnostic Testing
- Management
- Complications
- KD vs. MISC

”What is the age range of patients in whom you would consider a diagnosis of KD?”

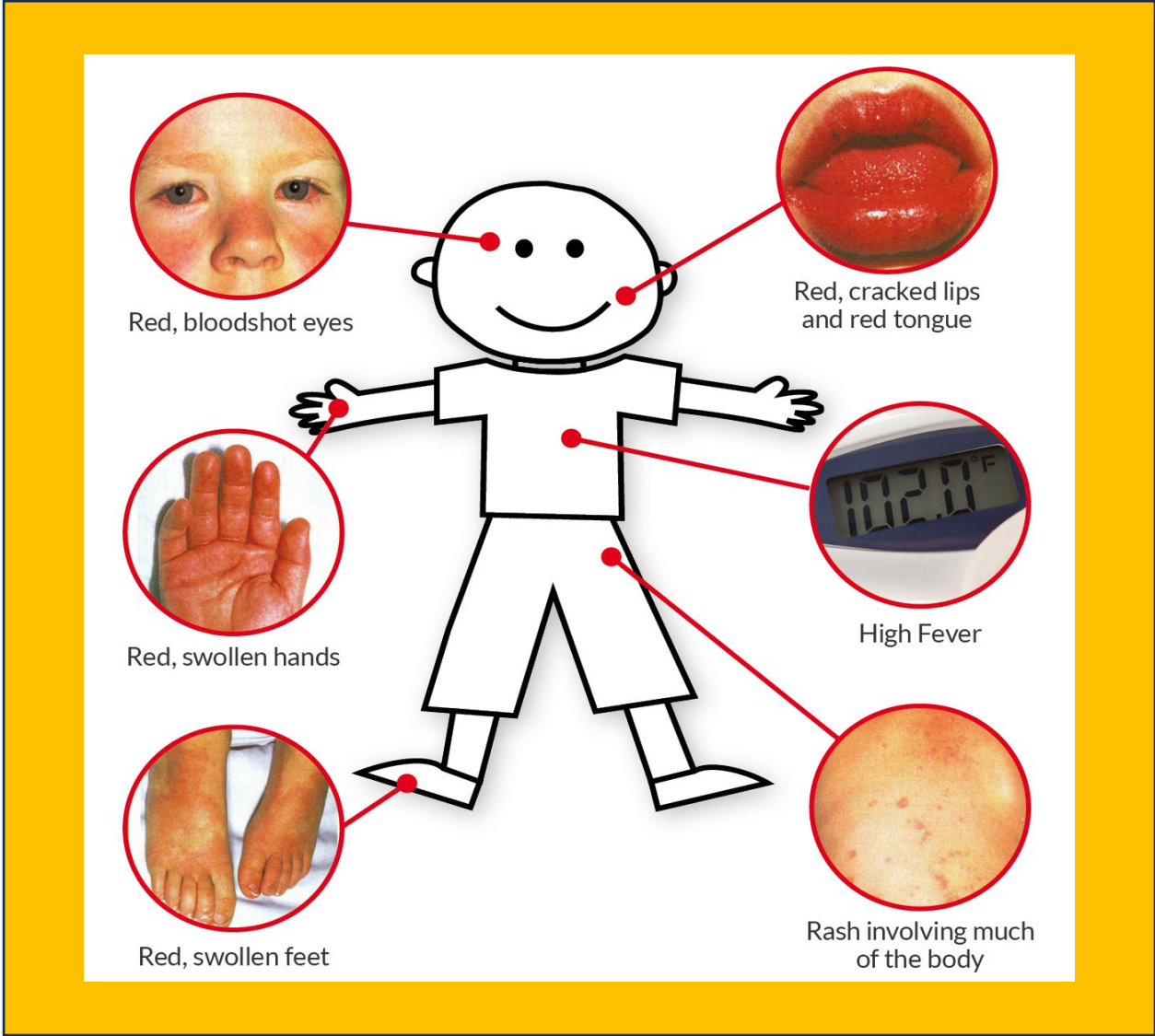
"GOK" DISEASE



Dr. Tomisaku Kawasaki

INTRODUCTION

- Age < 5
- Autoimmune disease
- Medium vessel vasculitis
- Acute; self-limited febrile disease
- Multiorgan involvement
- **Most common cause of cardiac disease in childhood**
- Low mortality rate worldwide (0-6%)³



DEFINITION

Kawasaki Disease [KD]

- Fever > 5 days + 4/5 clinical findings
- Fever > 4 days if > 4 principal clinical findings are present.

Incomplete KD

- Prolonged unexplained fever in an infant or child with < 4 of the principal/clinical features of KD

AND

- Compatible lab markers
 - ESR, CRP, transaminases, sterile pyuria

OR

- ECHO
 - Coronary artery dilation

FEVER

- First symptom
- $> 102^{\circ}\text{F}$
- ≥ 5 days to 2 weeks
- Not always responsive to antipyretics



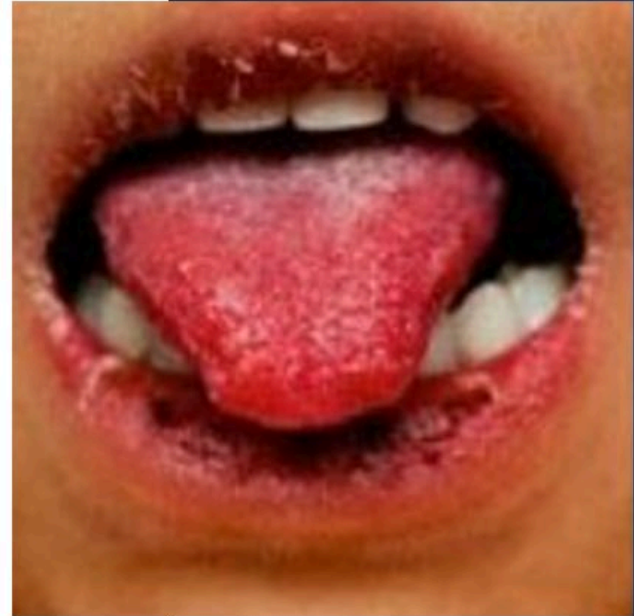


CONJUNCTIVITIS

- Bilateral
- Nonsuppurative
- Limbic sparing
- 2nd most common symptom

ORAL MANIFESTATIONS

- Erythema & cracking of the lips
- Erythema of the oropharyngeal mucosa
- "Strawberry" Tongue
- Pathophysiology:
 - Necrotizing microvasculitis with fibrinoid necrosis

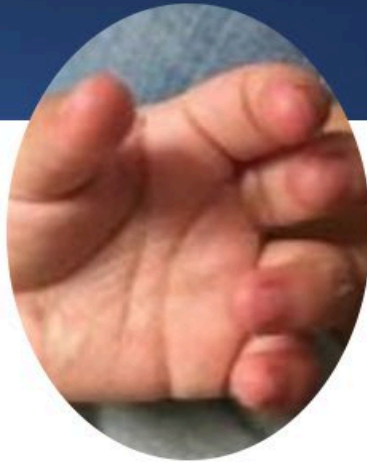
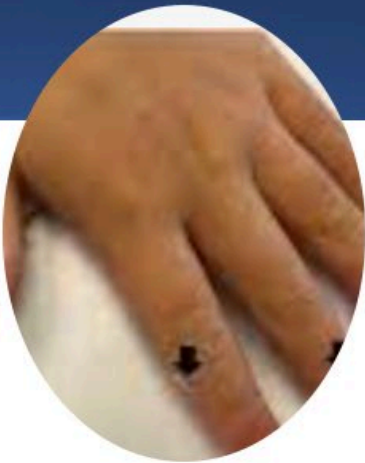


CERVICAL LYMPHADENOPATHY

- 50-75% of patients
- at least 1 LN > 1.5cm in diameter
- Painless, not fluctuant, nonsuppurative
- Not responsive to antibiotics



EXTREMITY INVOLVEMENT



- Erythema & edema of hands/feet
- Periungual desquamation
- Beau's lines

RASH



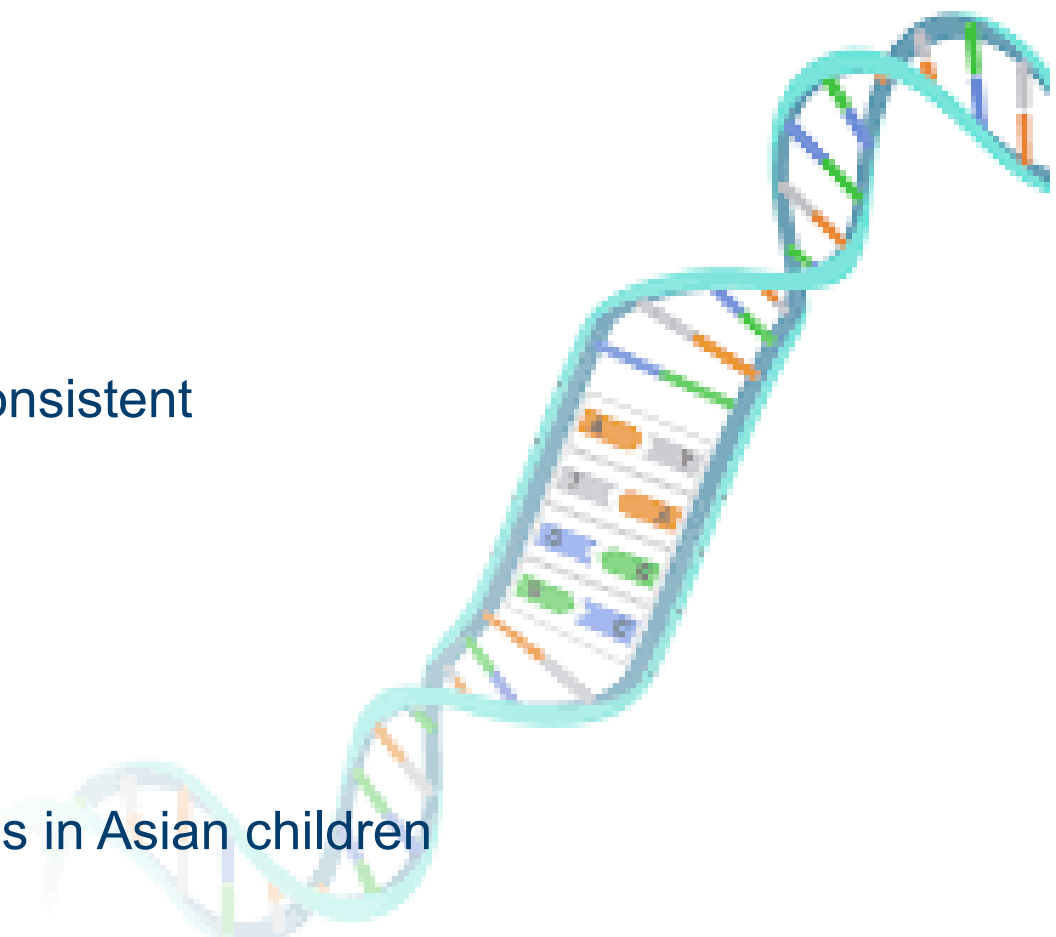
- Nonspecific maculopapular rash
- Diffuse erythroderma
- Location varies
- Not itchy

OTHER

Cardiovascular	<p>Myocarditis, pericarditis, valvular regurgitation, shock</p> <p>Coronary artery abnormalities</p> <p>Aneurysms of medium-sized non-coronary arteries</p> <p>Peripheral gangrene</p> <p>Aortic root enlargement</p>
Respiratory	<p>Peribronchial and interstitial infiltrates on chest X-ray</p> <p>Pulmonary nodules</p>
Musculoskeletal	<p>Arthritis, arthralgia (pleocytosis of synovial fluid)</p>
Gastrointestinal	<p>Diarrhea, vomiting, abdominal pain</p> <p>Hepatitis, jaundice</p> <p>Gallbladder hydrops</p> <p>Pancreatitis</p>
Nervous system	<p>Extreme irritability</p> <p>Aseptic meningitis (pleocytosis of cerebrospinal fluid)</p> <p>Facial nerve palsy</p> <p>Sensorineural hearing loss</p>
Genitourinary	<p>Urethritis/meatitis, hydrocele</p>
Other	<p>Desquamating rash in groin</p> <p>Retropharyngeal phlegmon</p> <p>Anterior uveitis by slit lamp examination</p> <p>Erythema and induration at bacillus Calmette-Guerin inoculation site</p>

EPIDEMIOLOGY

- Cause: unknown.
 - Autoimmune?
- Predisposing factors inconsistent
 - Genetics?
- M:F \approx 1.5:1
- The highest relative risk is in Asian children



EPIDEMIOLOGY

North America

- ≈25 cases/100000 children <5 years of age / year
- Winter/early spring in North America
- Hawaii: 34.3% of KD patients age < 1 year
- Connecticut: 18.7% of KD patients age < 1 year

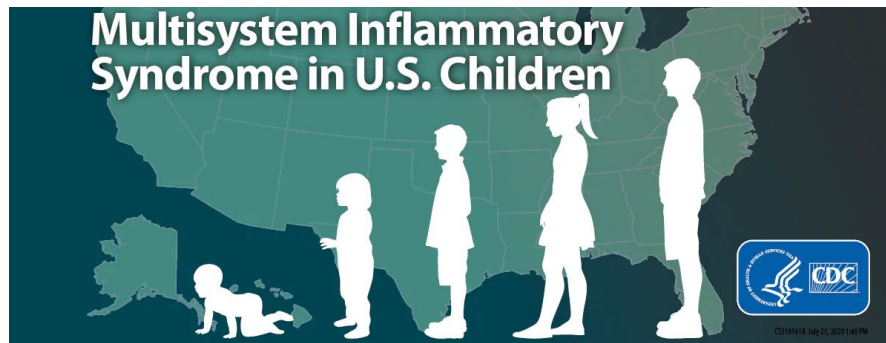
Japan

- 11.2% of KD patients age < 6 months
- 28.8% age < 1 year
- Recurrence rate: ≈3% in Japan
- RR in siblings is 10-fold higher in Japan
- Case fatality rate is <0.1% in Japan

**“Would it surprise you to hear that patients
ages < 6 months and > 8 years are at
highest risk of complications?”**

BEYOND THE SPECTRUM

- Incidence of coronary artery abnormalities is higher in young infants & adolescents
 - Delay in diagnosis?
 - Atypical evolution of disease?
- Or... have you considered the “Kawasaki look-a-like” – MIS-C



ATYPICAL PRESENTATIONS

- Diagnosis is challenging
- Conte, et al. narrative review (2023)
 - Case reports & cohorts of children with “atypical KD”
 - Canonical vs. non-canonical manifestations

CANONICAL MANIFESTATIONS

- Males > Females 1.85:1
- Age 2mo-14y
 - 1/3 studies age < 6 months
 - 1/2 studies age < 1 year
- 80% had fever > 7 days
- Most patients did NOT have:
 - Rash
 - Oral changes
 - Bulbar signs
 - Cervical adenitis
 - Extremity changes

Table 1

List of the general features and classical manifestations (according to the American Heart Association criteria) in case reports presenting 'atypical' pictures of Kawasaki disease.

	<i>Reference</i>	Age (Months)	Gender	Fever (>7 Days)	Rash	Oral Changes	Bulbar Signs	Cervical Adenitis	Extremity Changes
1	Ramamoorthy et al. [45]	19	M	YES	NO	NO	NO	NO	NO
2	Prokic et al. [30]	72	M	YES	YES	NO	NO	NO	NO
3	Rosario et al. [63]	2	M	YES	NO	NO	NO	YES	NO
4	Cason et al. [36]	168	M	YES	NO	NO	NO	YES	NO
5	Chaudhuri et al. [64]	5	M	YES	NO	NO	YES	NO	NO
6	Dyer et al. [65]	72	M	NO	NO	NO	YES	NO	NO
7	de Magalhães et al. [66]	3	F	YES	YES	NO	NO	NO	NO
8	Singh et al. [67]	102	M	NO	NO	NO	NO	NO	NO
9	D'Auria et al. [68]	48	M	YES	NO	NO	NO	YES	NO
10	Micallef et al. [69]	9	M	NO	NO	NO	NO	NO	NO
11	Tiao et al. [32]	30	M	YES	NO	NO	NO	NO	YES
12	Thapa et al. [70]	7	M	YES	NO	NO	NO	NO	NO
13	Peduzzi et al. [25]	3	F	NO	YES	NO	NO	NO	NO
14	Kim et al. [37]	108	F	NO	YES	YES	NO	YES	YES
15	Papadodima et al. [41]	132	M	YES	NO	NO	NO	NO	YES
16	Ren et al. [71]	12	M	YES	YES	NO	NO	NO	NO
17	Catalano-Pons et al. [49]	3	M	YES	NO	NO	YES	NO	NO
18	Sahoo et al. [72]	31	M	YES	NO	NO	NO	NO	YES
19	Sahoo et al. [72]	56	F	YES	YES	NO	NO	NO	NO
20	Sahoo et al. [72]	60	M	YES	NO	NO	NO	NO	YES
21	Behjati-Ardakani et al. [73]	120	F	YES	NO	NO	NO	NO	NO
22	Usta Guc et al. [74]	9	F	YES	NO	NO	YES	NO	NO
23	Godart et al. [33]	7	F	YES	NO	NO	YES	NO	NO
24	O'Byrne et al. [75]	8	M	YES	YES	NO	NO	NO	NO
25	Kritsaneepaiboon et al. [38]	10	F	YES	NO	NO	NO	NO	YES
26	Torres et al. [76]	31	M	YES	NO	NO	YES	NO	NO
27	Uziel et al. [46]	30	F	YES	NO	NO	NO	NO	YES

NON-CANONICAL MANIFESTATIONS

- Pulmonary
 - Respiratory distress
 - PNA
- GI
 - Nausea/vomiting/diarrhea
 - Hepatomegaly, cholestatic hepatitis, pancreatitis, and hydrops of the GB
- Renal
 - 80% sterile pyuria
 - Other rare findings: interstitial nephritis, HUS, immune-complex mediated nephropathy, acute nephritic syndrome
- Other
 - Severe irritability, meningitis, arthritis, retropharyngeal abscess

Table 2

List of the non-canonical findings in case reports presenting ‘atypical’ pictures of Kawasaki disease.

	<i>Reference</i>	Pulmonary Signs	Gastrointestinal Symptoms	Kidney Involvement	Other Involved Organs
1	Ramamoorthy et al. [45]	Pneumonia, respiratory distress	NO	NO	NO
2	Prokic et al. [30]	NO	Abdominal pain, hydrops of the gallbladder, pancreatitis	NO	NO
3	Rosario et al. [63]	NO	NO	NO	Pericardial effusion with shock
4	Cason et al. [36]	NO	NO	NO	Retropharyngeal abscess
5	Chaudhuri et al. [64]	NO	NO	NO	NO
6	Dyer et al. [65]	NO	NO	NO	Torticollis
7	de Magalhães et al. [66]	Pulmonary thrombosis	NO	NO	BCG scar erythema, systemic and pulmonary thrombosis, dilatation of iliac arteries
8	Singh et al. [67]	Respiratory distress, pleural effusion	Vomiting, nausea, abdominal pain	NO	Dilatation of systemic arteries with shock
9	D’Auria et al. [68]	NO	Vomiting, diarrhoea	NO	Exudative pericarditis, anasarca
10	Micallef et al. [69]	Tachypnoea	Vomiting Small bowel obstruction,	NO	Urticaria
11	Tiao et al. [32]	NO	vomiting with coffee-ground features	NO	NO
12	Thapa et al. [70]	NO	NO	NO	NO
13	Peduzzi et al. [25]	Tachypnoea	NO	NO	Pericardial effusion, myocardial infarction with shock
14	Kim et al. [37]	NO	NO	NO	Retropharyngeal abscess
15	Papadodima et al. [41]	NO	Melena	Hematuria	Pericardial effusion, coronary artery thrombosis
16	Ren et al. [71]	NO	NO	NO	NO
17	Catalano-Pons et al. [49]	NO	NO	NO	Aseptic meningitis
18	Sahoo et al. [72]	Respiratory distress	Vomiting, abdominal pain	NO	Congestive heart failure
19	Sahoo et al. [72]	Respiratory distress	NO	NO	Congestive heart failure
20	Sahoo et al. [72]	Respiratory distress	NO	NO	Congestive heart failure

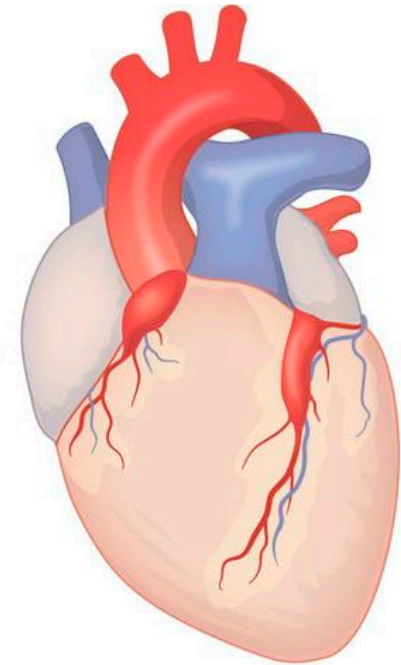
21	Behjati-Ardakani et al. [73]	NO	NO	NO	Wrist arthritis, arthralgia of hips knees and ankles, neck pain
22	Usta Guc et al. [74]	NO	NO	NO	Severe irritability
23	Godart et al. [33]	NO	Abdominal obstruction, necrosis of the gut, short-bowel syndrome	NO	NO
24	O'Byrne et al. [75]	Respiratory distress	Melena	NO	Pericardial effusion, coronary artery thrombosis
25	Kritsaneepaiboon et al. [38]	NO	NO	NO	Retropharyngeal abscess, brightening, cuffing or wall irregularities of coronary arteries
26	Torres et al. [76]	NO	NO	NO	NO
27	Uziel Y et al. [46]	Pulmonary consolidation	NO	NO	Bullous meningitis
28	Uziel et al. [46]	Pulmonary consolidation	Vomiting	NO	Anasarca
29	Yang et al. [77]	NO	Abdominal pain, hydrops of the gallbladder, pancreatitis	NO	Cardiac arrest
30	Doğan et al. [78]	NO	NO	NO	Myocardial infarction
31	Guile et al. [79]	NO	Gallbladder	NO	Pericardial effusion
32	Pinches et al. [80]	NO	NO	NO	Heart murmur
33	Uchida et al. [81]	NO	NO	NO	Coronary artery aneurysm
34	Choi et al. [82]	NO	NO	NO	Heart murmur

ATYPICAL KD LABS

- CRP \geq 3 mg/dL
- ESR \geq 40 mm/hr
- Anemia for age
- Neutrophil leukocytosis
- Hypoalbuminemia
- Transaminitis
- ***Elevated NT-proBNP**
 - Best cutoff values?
- **If you are considering incomplete KD on your list of diagnoses, order: CRP, ESR, CBC, CMP, UA, AND an ECHO!**

FAILURE TO DIAGNOSE

- 50% of general pediatricians and 25% of infectious disease specialists did not consider diagnosis of KD before 6 months or after 8 years of age.
- Delayed diagnosis → development of coronary artery aneurysms



Giant aneurysms of the right coronary artery and left anterior descending coronary artery

METHODS

- Location: San Diego county
- 132/227 (58.1%) general pediatricians from AAP
- 345/651 (53.0%) pediatric infectious disease specialists from Pediatric Infectious Disease Society
- Survey:
 - Year of graduation from Medical School
 - Participation in the care of KD patients
 - Ability to remember the 5 diagnostic criteria
 - Potential age range of KD patients

TABLE 1. Number of KD Patients Cared for by Study Participants

No. of KD Patients/TD	No. of General Pediatricians (n = 132)	No. of Pediatric Infectious Disease Subspecialists (n = 340)	<i>P</i> *
0–10	87 (65.9) [†]	21 (6.2)	<0.001
11–50	43 (32.6)	168 (49.4)	0.001
>50	2 (1.5)	151 (44.4)	<0.001

* χ^2 analysis.

[†]Numbers in parentheses, percent.

TABLE 2. Minimum Age of Patients in Whom the Diagnosis of KD Was Considered by Study Participants

Minimum Age	No. of General Pediatricians (n = 124)	No. of Pediatric Infectious Disease Subspecialists (n = 324)
Younger than 6 mo	53 (42.7)*	238 (73.5)
6–11 mo	29 (23.4)	53 (16.4)
12–18 mo	21 (16.9)	22 (6.8)
19–24 mo	11 (8.9)	6 (1.9)
3 yr	5 (4.0)	3 (0.9)
4 yr	2 (1.6)	2 (0.6)
Older than 5 yr	3 (2.4)	0 (0)

*Numbers in parentheses, percent.

TABLE 3. Maximum Age of Patients in Whom the Diagnosis of KD Was Considered by Study Participants

Maximum Age	No. of General Pediatricians (n = 124)	No. of Pediatric Infectious Disease Subspecialists (n = 325)
Younger than 5 yr	2 (1.6)*	5 (1.5)
5–6 yr	39 (31.5)	53 (16.4)
7–8 yr	23 (18.5)	23 (7.1)
9–10 yr	25 (20.2)	45 (13.9)
11–12 yr	14 (11.3)	37 (11.4)
13–14 yr	3 (2.4)	10 (3.1)
15–16 yr	5 (4)	30 (9.3)
17–18 yr	14 (10.5)	89 (27.5)
Older than 18 yr	0 (0)	32 (9.9)

Numbers in parentheses, percent

LIMITATIONS

- Was the question misunderstood?
- 53-58% response rate
- Cardiologists, rheumatologists, family practitioners not included
- Limited to San Diego area

COMPLICATIONS

- 1st pathological step: Coronary arteritis
- Coronary artery aneurysms → MI, heart failure, death
- Chronic CV outcomes: HLD, premature CAD, HTN
- Increased risk of allergic diseases
- Increased cancer risk beyond 6 years
- Increased risk of rheumatological disorders

COMPLICATIONS

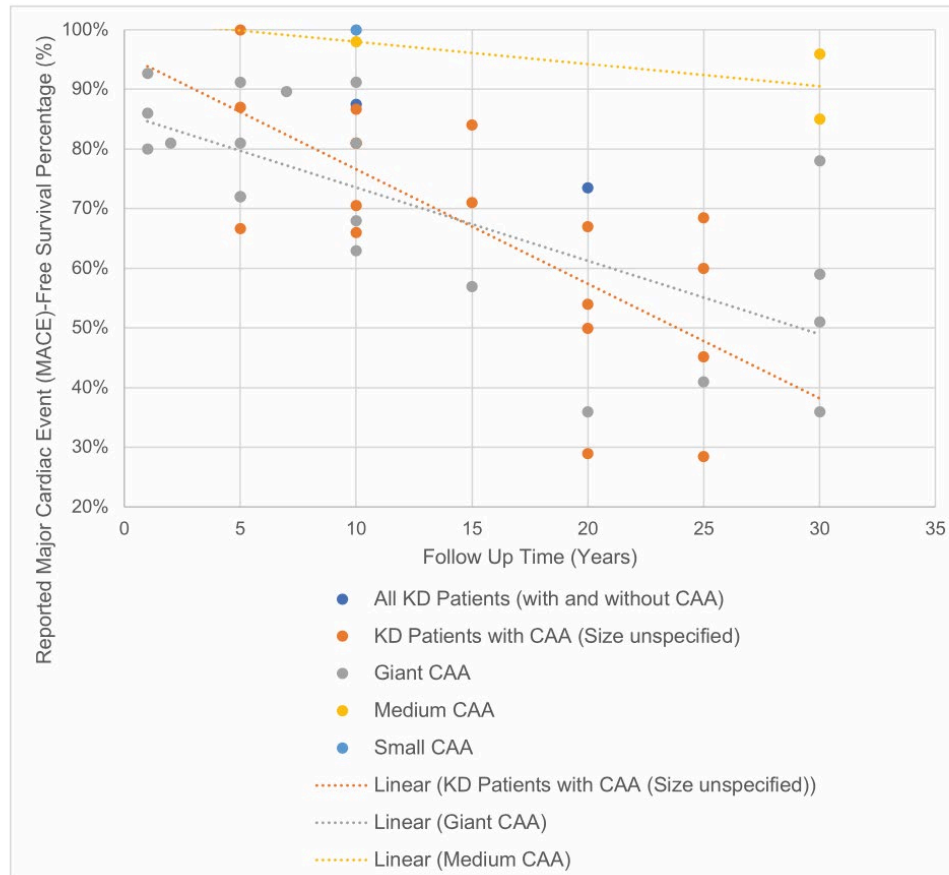
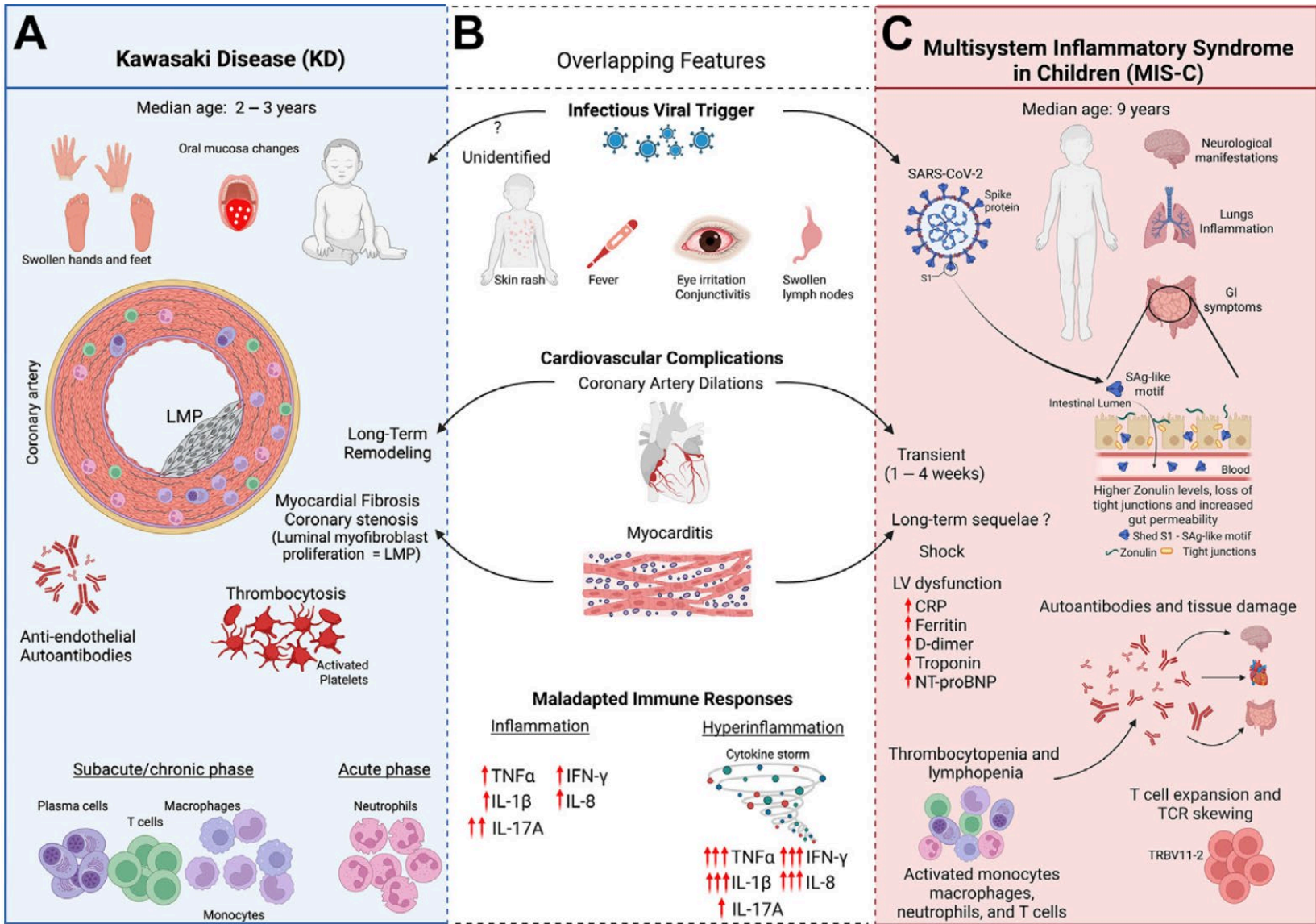


FIGURE 3
MACE-free survival in patients with KD over follow-up time.

KD vs. MIS-C

- Two distinctive diseases
- Triggered by different infectious agents
- Differ in their etiology, demography, epidemiology, clinical & laboratory findings, and pathology
- Share many common inflammatory characteristics



KD vs. MIS-C

KD

- Age < 5
- M:F 1.5:1
- Asian population

MIS-C

- Median age 9
- M-F 1:1 (age 0-4)
- M-F 2:1 (age 18-20)
- Hispanic or Black

KD vs. MIS-C

KD

- Fever > 5 days + 4/5 clinical findings
- Fever > 4 days if > 4 principal clinical findings are present
- Cardiac
 - Myocarditis → fibrosis
 - Coronary artery dilation (long-term sequelae)
- Inflammatory cytokine release

MIS-C

- 30% meet criteria for complete KD
- ↑↑↑ CRP, ferritin, D-dimer, troponin, NT-proBNP
- ↓ lymphocyte & platelets
- Cardiac:
 - LV systolic & diastolic dysfunction
 - Myocardial inflammation
 - Coronary artery dilation (transient)
- Cytokine Storm

2021 KD GUIDELINES

- American College of Rheumatology/Vasculitis Foundation
- 16 clinical PICO questions developed after review of 275 articles
- 2 voting panels:
 - Initial: 9 adult rheumatologists,
5 peds rheumatologists
2 patients
 - Second: 8 new peds rheumatologists
4 peds rheumatologists who previously participated
1 peds ID doc
1 peds cardiologist
- 70% of voting panel for consensus on recommendations

2021 KD GUIDELINES

- Strong recommendation:
 - Supported by moderate-high quality evidence
 - Applies to all or almost all patients
- Conditional recommendation
 - Supported by lower quality evidence
 - Balance between desirable & undesirable outcomes
 - Applies to majority of patients
 - Warrants shared decision-making approach

2021 KD RECOMMENDATIONS

1 Good Practice Statement

7 Strong Recommendations

4 Conditional Recommendations

1 GOOD PRACTICE STATEMENT

- **IVIG** is the **STANDARD OF CARE**

- Reduction in rate of coronary artery aneurysms
- Reduction in duration of fever

7 STRONG RECOMMENDATIONS

Pts with atypical KD

1. Do not delay IVIG.

Resolution of fever prior to day 10 is NOT an indication to withhold treatment in pts meeting criteria for incomplete or complete KD

Pts with acute KD & suspected or diagnosed MAS

2. IVIG + additional agents for MAS

Pts with acute KD

3. ASA (high vs. low dose)

7 STRONG RECOMMENDATIONS (CONT.)

Pts with acute KD with subsequent resolution of fevers

4. Continued daily monitoring of fevers

Pts with suspected incomplete KD + fever

5. Obtain the ECHO

Pts with unexplained shock physiology

6. Obtain the ECHO

Pts with unexplained MAS

7. Obtain the ECHO

4 CONDITIONAL RECOMMENDATIONS

Pts at **high risk for IVIG resistance** or developing coronary artery aneurysms

1. IVIG + glucocorticoid OR
2. IVIG + nonglucocorticoid immunomodulatory suppressive agents

HIGH RISK

Z Score in the LAD or RCA ≥ 2

Age < 6 months

Asian race

CRP ≥ 13 mg/dl

Pts with persistent fevers after initial treatment with IVIG

3. 2nd dose of IVIG > glucocorticoids

Pts with arthritis that persists after IVIG & do NOT have coronary artery aneurysms

4. NSAIDS

1 UNGRADED STATEMENT

Pts with persistent fevers after 2nd dose of IVIG

- Nonglucocorticoid immunosuppressive therapy OR glucocorticoids

Kawasaki Dz

Definition: Kawasaki disease (KD) is a vasculitis syndrome that can cause damage to blood vessels and the heart.
Most commonly affecting kids < 5 years of age.

Consider KD for patients with
≥ 5 days of daily fevers (reported/recorded ≥ 100.4)
AND
≥ 4 clinical features
OR
consistent lab findings

Inclusion Criteria

> 5 days of DAILY fever w/ clinical suspicion for KD

Exclusion

Fever < 5 days

Clinical features c/w:
Hemophagocytic lymphohistiocytosis
Macrophage Activation Syndrome
Sepsis
MISCI*

If child triggers sepsis → follow Sepsis Pathway

* Multisystem Inflammatory Syndrome in Children (MISCI)

Definition: Dysregulated autoimmune disorder post COVID-19 (~2-6 wks)

More commonly affecting age > 5 yrs

Clinically: mimics KD; involves MULT/organs +/- shock on presentation w/ mortality risk

See page 2 & expand workup to include EKG & additional labs to

1. Triage ESI 2 or 3
2. History & Physical Exam
Reminder clinical features are not usually present at same time
3. Proceed if complete vs **incomplete** KD concern persists

1. CBC with diff, CRP, ESR, CMP
2. Urinalysis (add culture if obtained via catheterization)
3. Blood culture

CRP ≥ 3 + ESR ≥ 40 & ≥ 2 of the following:

Decreased ALBUMIN ≤ to 3gm/dl

Increased WBC > 15,000

Increased Platelets > 450,000

Increased Pyuria ≥ 10/hpf

Anemia for age:

- < 4 years: < 11.0 g/dL

- 5-7yrs: < 11.5 g/dL

- 8-11yrs: < 12.0 g/dL

- Females 12-14yrs: < 12.0 g/dL

- Males 12-14yrs: < 13 g/dL

Increased Transaminases (AST > 45; ALT > 35)

Clinical Features in KD

Principal Clinical Features of KD

May not all be present at the same time

Oral changes

Erythema and cracking of lips, strawberry tongue, and/or erythema of oral and pharyngeal mucosa

Conjunctivitis

Bilateral bulbar conjunctival injection without exudate

Rash

Maculopapular, diffuse erythroderma, or erythema multiforme-like

Extremity changes

Erythema and edema of the hands and feet in acute phase and/or periungual desquamation in subacute phase

Lymphadenopathy

Cervical lymphadenopathy (≥ 1.5 cm diameter), usually unilateral

Lab Findings in KD

Elevated CRP

Elevated ESR

Anemia for age

Leukocytosis with neutrophils and immature forms

Thrombocytosis after 1 week

Sterile pyuria

Hyponatremia

Elevated serum transaminases

Elevated serum gamma glutamyl transpeptidase

Hypoalbuminemia

Abnormal plasma lipids

Pleocytosis of CSF

Leukocytosis in synovial fluid

Yes

Multiplex Viral PCR

ADMIT

Admit to pediatric hospital medicine
Likely course includes ECHO, IVIG, ASA

No

< 4 clinical features AND labs W/O KD concerns
Re-eval: if child is well appearing & PO tolerant

DISCHARGE

Discharge home with PMD follow up & strict ED return precautions

Created by: Anna Suessman, DO, MED; Todd Mastrovitch, MD. Published: July 1, 2024

TAKE AWAY

- 2021 ACR/vasculitis guidelines & recs supplement 2017 AHA guidelines.
- Must have high degree of suspicion for classic KD or atypical KD at extremes of age.
- If KD is on your list of differentials, just order the ECHO.
- Pts with incomplete KD have at least the same if not higher risk of coronary artery aneurysms as classic KD. Therefore, do not delay treatment with IVIG.
- Pts with coronary artery aneurysms are at high risk for major cardiac events.
- Increased risk for long-term non-cardiac diseases.
- Overall, survival appears largely unaffected after KD.

GOAL: PREVENTION

QUESTION

- A 4-year-old boy presents with red eyes and an erythematous rash on his extremities. Associated symptoms include a high fever for 4 days and mouth ulcers. He has no significant medical history and takes no medications. Vital signs are blood pressure 110/75 mm Hg, heart rate 119 bpm, oxygen saturation 96% on room air, and temperature 39.7 °C (103.5 °F). On physical examination, he has swollen lymph nodes on the left side of the neck. His oral mucosa is red, and his extremities reveal a red rash and edema. His eyes show bilateral painless conjunctival injection without exudate. Which of the following is the best initial management?
- 1. Intravenous immunoglobulin and low-dose aspirin
- 2. Intravenous immunoglobulin and high-dose aspirin
- 3. Intravenous immunoglobulin and high-dose corticosteroids
- 4. Intravenous immunoglobulin and infliximab

ANSWER

- The patient must have fevers for 5 or more days, with at least 4 of the following criteria to diagnose Kawasaki disease: Bilateral painless conjunctival injection without exudate, erythematous mouth, and pharynx, strawberry tongue or red, cracked lips, polymorphous exanthem, swelling of hands, and feet with erythema of the palms and soles, and cervical lymphadenopathy (over 1.5 cm in diameter).
- Patients with Kawasaki disease can develop coronary artery aneurysms if not managed promptly.
- Patients should receive high-dose intravenous immune globulin (IVIG) at 2 g/kg over 10- to 12 hours and high-dose aspirin (80 mg/kg/day to 100 mg/kg/day divided every 6 hours) until they have been afebrile for over 48 hours.
- The disease can also lead to heart failure, myocardial infarction, myocarditis, valvulitis, pericarditis with pericardial effusion, and rupture of the coronary arteries leading to hemopericardium and sudden death.



Thank you!

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