

INCLUSION CRITERIA

Fever for ≥ 4 days without an alternative explanation
OR infants ≤ 6 mo with fever ≥ 7 days without other explanation

EXCLUSION CRITERIA

- Known etiology without additional concerns for KD.
- ≥ 4 days of fever without additional concern for KD

MEASURES

Increase percentage of KD patients at high risk of IVIG resistance that receive steroids at admission

¹DIAGNOSTIC CRITERIA:

Fever ≥ 5 Days AND at least 4 of 5 principal clinical features.

KD can be diagnosed in the presence of ≥ 4 principal features, particularly when redness/swelling of hands/feet are present with 4 days of fever.

²Principal Clinical Features:

- Bilateral bulbar conjunctival injection without exudate
- Oral mucous membrane changes (erythema and/or fissuring of lips, erythema of oropharyngeal mucosa, and/or strawberry tongue)
- Polymorphous rash (maculopapular, diffuse erythroderma, or erythema multiforme-like)
- Cervical lymphadenopathy (at least 1 lymph node > 1.5 cm in diameter, usually unilateral)
- Peripheral extremity changes (erythema of palms/soles, and/or edema of hands/feet and/or periungual desquamation)

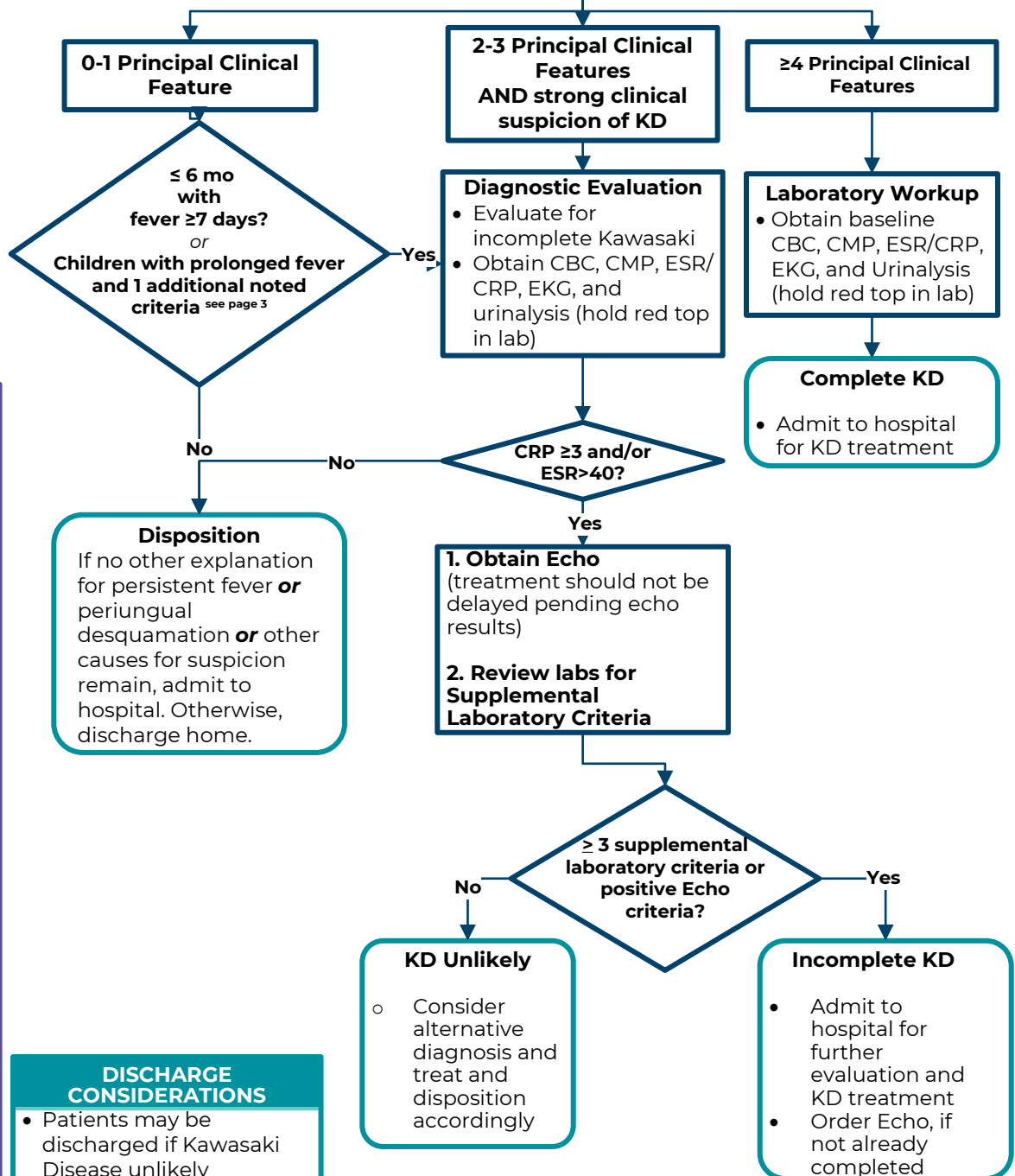
DISCHARGE CONSIDERATIONS

- Patients may be discharged if Kawasaki Disease unlikely
- Ensure follow-up with PCP in 24-48 hours
- If fever persists, will require serial clinical and laboratory re-evaluation. If periungual desquamation develops even after fever resolves, then obtain echocardiogram

INITIAL ASSESSMENT

- Perform history and physical examination

Assess for diagnostic criteria¹ and principal clinical features²



HIGH RISK CRITERIA FOR IVIG RESISTANCE FOR WHICH INITIAL TREATMENT INTENSIFICATION WITH STEROIDS OR INFLIXIMAB SHOULD BE CONSIDERED:

One of the following:

- Z-score of Left Anterior Descending (LAD) or Right Coronary Artery (RCA) ≥ 2.5 on baseline echo
- Age at fever onset ≤ 6 months

For patients of **Japanese descent**, use the Kobayashi criteria or similar validated criteria to determine risk. Kobayashi criteria (score ≥ 5 is considered high risk).

- Sodium < 133 mmol/L (2 points)
- Aspartate aminotransferase ≥ 100 international units/L (2 points)
- C-reactive protein ≥ 10 mg/dL (1 point)
- Neutrophils $\geq 80\%$ of the white blood cell count differential (2 points)
- Platelet count $\leq 300,000/\text{mm}^3$ (1 point)
- Days of illness at initial treatment ≤ 4 (2 points)
- Age ≤ 12 months (1 point)

For **Norther American patients**, use the following criteria (a score > 3 is considered high risk)

- Enlarged Coronary arteries on echocardiogram with a Z-score at baseline ≥ 2.00 (2 points)
- Age at fever onset < 6 months (1 point)
- Any Asian race reported (1 point)
- C-reactive protein > 13 mg/dL (1 point)

SUPPLEMENTAL LABORATORY CRITERIA

Need at least 3 of 6:

- Anemia for age
- Platelet count $\geq 450,000$ after the 7th day of fever
- Albumin ≤ 3.0 g/dL
- Elevated ALT level
- WBC count $\geq 15,000/\text{mm}^3$
- ≥ 10 WBC/hpf on urinalysis

POSITIVE ECHO CRITERIA

- Z-score of left or right coronary arteries ≥ 2.5 **OR**
- Coronary artery aneurysm **OR**
- ≥ 3 other suggestive features exist:
 - Decreased left ventricular function
 - Mitral regurgitation
 - Pericardial effusion
 - Z-score 2 to 2.5 in left or right coronary arteries

Infants and children with prolonged fever and 1 additional noted criteria

- Infants and children with prolonged fever and unexplained aseptic meningitis
- Infants or children with prolonged fever and unexplained or culture-negative shock
- Cervical lymphadenitis unresponsive to antibiotic therapy
- Retropharyngeal or parapharyngeal phlegmon unresponsive to antibiotic therapy

Medication	Route	Dose	Max. per Dose	Duration
Aspirin MODERATE dose	PO/PR	30-50 mg/kg/day divided Q6h		Until 48 hours after resolution of fever
Aspirin LOW dose	PO	3-5 mg/kg once daily	325 mg	6-8 weeks, assess at follow-up. Discontinuation dictated by cardiology
Famotidine	IV	< 3 months: 0.25 mg/kg/dose once daily ≥ 3 months: 0.25 mg/kg/dose twice daily	20 mg	Discontinue when off steroids and moderate dose aspirin
	PO	< 3 months: 0.5 mg/kg/dose once daily ≥ 3 months: 0.5 mg/kg/dose twice daily		
IVIG	IV	2 g/kg once		
Methylprednisolone/Prednisone/ Prednisolone – adjunctive	IV/PO	1 mg/kg/dose twice daily for 10 days, followed by 1 mg/kg/dose once daily for 5 days	30 mg	15 days total

Abbreviations

Abbreviation	Terminology	Abbreviation	Terminology
CBC	Complete Blood Count	EKG	Electrocardiogram
CMP	Complete Metabolic Panel	ESR	Erythrocyte Sedimentation Rate
CRP	C-Reactive Protein	mo	Month
ECHO	Echocardiogram	PCP	Primary Care Physician
ED	Emergency Department		

Disclaimer: The clinical pathways are based upon publicly available medical evidence and/or a consensus of medical practitioners at Nemours Children's Health ("Nemours") and are current at the time of publication. These clinical pathways are intended to be a guide for practitioners and may need to be adapted for each specific patient based on the practitioner's professional judgment, consideration of any unique circumstances, the needs of each patient and their family, and/or the availability of various resources at the health care institution where the patient is located.

VERSION MANAGEMENT

- Version 1.0 (11/11/2025): Go Live.

Questions about this pathway should be directed to clinicalpathways@nemours.org.

AUTHORS & REFERENCES

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