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**Kawasaki Disease: Diagnostic Criteria, Work-Up, and Treatment**

Kawasaki disease (KD, previously called mucocutaneous lymph node syndrome) is one of the most common vasculitides of childhood [[1](https://www-uptodate-com.york.ezproxy.cuny.edu/contents/kawasaki-disease-clinical-features-and-diagnosis/abstract/1)]. KD also occurs rarely in adults. It is typically a self-limited condition, with fever and manifestations of acute inflammation lasting for an average of 12 days without therapy [[2](https://www-uptodate-com.york.ezproxy.cuny.edu/contents/kawasaki-disease-clinical-features-and-diagnosis/abstract/2)]. However, complications such as coronary artery (CA) aneurysms, depressed myocardial contractility and heart failure, myocardial infarction, arrhythmias, and peripheral arterial occlusion may develop and lead to significant morbidity and mortality.

The etiology of Kawasaki disease still remains unknown. A number of microorganisms including bacteria, viruses, rickettsiae etc. were hypothesized as an etiology of the illness. Unfortunately, no specific agent that provides reproducible evidence has yet reported.

**Dx Criteria:**

Diagnostic criteria developed byTomisaku Kawasaki in 1967. Requires the presence of ***fever lasting ≥5 days***, combined ***with*** ***at least four*** of the five following physical findings, without an alternative explanation ([table 1](https://www-uptodate-com.york.ezproxy.cuny.edu/contents/image?imageKey=PEDS%2F67711&topicKey=ALLRG%2F6417&search=kawasaki+disease+in+pediatrics&rank=1%7E150&source=see_link)) [[1,2,47](https://www-uptodate-com.york.ezproxy.cuny.edu/contents/kawasaki-disease-clinical-features-and-diagnosis/abstract/1,2,47)]:

●Bilateral bulbar conjunctival injection ([picture 1](https://www-uptodate-com.york.ezproxy.cuny.edu/contents/image?imageKey=PEDS%2F78898&topicKey=ALLRG%2F6417&search=kawasaki+disease+in+pediatrics&rank=1%7E150&source=see_link))

●Oral mucous membrane changes, including injected or fissured lips ([picture 2](https://www-uptodate-com.york.ezproxy.cuny.edu/contents/image?imageKey=ALLRG%2F59319&topicKey=ALLRG%2F6417&search=kawasaki+disease+in+pediatrics&rank=1%7E150&source=see_link)), injected pharynx, or strawberry tongue ([picture 3](https://www-uptodate-com.york.ezproxy.cuny.edu/contents/image?imageKey=ID%2F68321&topicKey=ALLRG%2F6417&search=kawasaki+disease+in+pediatrics&rank=1%7E150&source=see_link))

●Peripheral extremity changes, including erythema of palms or soles, edema of hands or feet (acute phase) ([picture 4](https://www-uptodate-com.york.ezproxy.cuny.edu/contents/image?imageKey=ALLRG%2F72040&topicKey=ALLRG%2F6417&search=kawasaki+disease+in+pediatrics&rank=1%7E150&source=see_link)), or periungual desquamation (convalescent phase) ([picture 5](https://www-uptodate-com.york.ezproxy.cuny.edu/contents/image?imageKey=ALLRG%2F53452&topicKey=ALLRG%2F6417&search=kawasaki+disease+in+pediatrics&rank=1%7E150&source=see_link))

●Polymorphous rash

●Cervical lymphadenopathy (at least one lymph node >1.5 cm in diameter)

Rash and conjunctival injection are seen with many illnesses, but other KD features, such as red, cracked lips and redness and swelling of the hands and feet, are unusual in the illnesses in the differential diagnosis and should increase the suspicion for KD.

**Work-Up:**

***Physical exam:***

* ***Fever*** – typically above 101.3 and doesn’t respond well to antipyretics
* ***Conjunctivitis*** – b/l nonexudative conjunctivitis present in 90%, slit-lamp may be helpful
* ***Mucositis*** – cracked, red lips and “strawberry tongue” are characteristic
* ***Rash*** – polymorphous rash usually begins during first few days of illness; typically perineal erythema and desquamation first, followed by macular, morbiform, or targetoid skin lesions
* ***Extremity Changes*** – generally last to appear; indurated edema of dorsum of hands and feet and diffuse erythema of palms and soles
* ***Lymphadenopathy*** – cervical lymphadenopathy is the least consistent feature of KD, most commonly anterior cervical nodes
* Cardiovascular findings – CV findings are not dx’c of KD, but they support the dx since most conditions that mimic KD do not involve the heart.
* Arthritis – not included in the criteria but reported in 7.5-25% of cases
* Other findings include N/V/D, irritability, cough, rhinorrhea, decreased oral intake, and joint pain
* Infants at increased risk of CA aneurysms, possibly due to delay in treatment due to lack of complete dx criteria
* In adults, approx. ¼ adult KD cases occur in HIV pts. Cervical lymphadenopathy, hepatitis, and arthralgia more common in adults and meningitis, thrombocytosis, and CA aneurysms less common.

***Labs:*** Not included in the diagnostic criteria for typical KD. However, certain findings may support diagnosis, particularly for incomplete KD cases.

* Elevated ESR & CRP
* Increased ferritin in acute phase
* Lymphocytes drop during acute phase then rise dramatically during convalescence
* Platelets rise by second week
* Normocytic, normochromic anemia in peds
* Urinary WBC’s
* Thrombocytopenia, high TGC, low Na, elevated LFT’s, and monocytes/macrophages in CSF can be signs of subclinical MAS and may warrant further testing

**Treatment:**

***Recommended therapies include:***

* IVIG 2g/kg single-infusion over 8-12 hrs
* ASA 30-50 mg/kg PO daily divided into 4 doses
* IVIG resistance must be determined before tx; pts at high-risk for IVIG resistance are additionally treated with systemic glucocorticoids
  + Prednisone 2 mg/kg/day IV x 5 days, then 2 mg/kg/day PO x 5 days, tapering to 1 mg/kg/day x 5 days, then 0.5 mg/kg/day x 5 days or until afebrile
* Pts typically observed for 24 hrs (min 12) after completion of IVIG tx to confirm resolution of fever
* Timing of discharge may depend on CA (dilated or normal) and risk factors for ultimately failing IVIG tx

***Other tx for refractory KD:***

* Cyclosporine (a calcineurin inhibitor) has shown some efficacy for refractory KD and those at high risk of IVIG resistance
* TNF-alpha inhibitors (eg etanercept or infliximab) have been studied as both adjuvant therapy for KD and as monotherapy for refractory KD

Sources:

* <https://www-uptodate-com.york.ezproxy.cuny.edu/contents/kawasaki-disease-clinical-features-and-diagnosis?search=kawasaki%20disease%20in%20pediatrics&source=search_result&selectedTitle=1~150&usage_type=default&display_rank=1#H26262883>
* <https://www-uptodate-com.york.ezproxy.cuny.edu/contents/kawasaki-disease-initial-treatment-and-prognosis?search=kawasaki%20disease%20in%20pediatrics&source=search_result&selectedTitle=2~150&usage_type=default&display_rank=2>
* <https://www.ncbi.nlm.nih.gov/pubmed/18260321>