CLINICAL PATHWAY:

Kawasaki Disease and Incomplete Kawasaki Disease

THIS PATHWAY SERVES AS A GUIDE AND DOES NOT REPLACE CLINICAL JUDGMENT.

Inclusion Criteria:

Fever ≥ 4 days **and** *at least* 4/5 of the following clinical criteria, **OR**
Fever ≥ 5 days **and** *at least* 2/5 of the following **clinical criteria**:

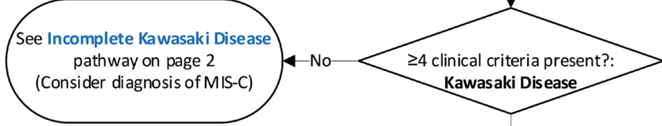
1. Bilateral bulbar non-exudative conjunctivitis
2. Mucosal changes (red cracked lips, strawberry tongue, erythema of oral and pharyngeal mucosa)
3. Polymorphous rash
4. Extremity changes (swelling and/or erythema, peeling occurs in convalescent phase)
5. Cervical adenopathy (≥ 1.5 cm diameter, usually unilateral)

Exclusion Criteria:

<2 mo old, exudative conjunctivitis, exudative pharyngitis, oral ulcers, bullous or vesicular rash, generalized adenopathy, splenomegaly, suspicion of Multi-System Inflammatory Disorder/MIS-C (follow the MIS-C Clinical Pathway), signs and symptoms can be easily explained by another condition

Initial Evaluation:

- CBC w diff, CRP, ESR, liver panel (without coags)
- UA with microscopy (clean catch or bag specimen)
- Consider chem 7



≥4 clinical criteria present?: **Kawasaki Disease**

- No → See Incomplete Kawasaki Disease pathway on page 2 (Consider diagnosis of MIS-C)
- Yes → **Admit to Hospital Medicine Service**

Treat:

- **IVIG** 2 g/kg x1 dose (max 100 g/dose). *Can start IVIG without obtaining ECHO first.*
- Medium dose **Aspirin PO** 30-50 mg/kg/day div q6hr, until afebrile x48hr
- If any high risk conditions[1] present, also add:
  - **Methylprednisolone IV** 1 mg/kg BID (max 60 mg/day) while febrile
  - When afebrile, change to **Prednisone/Prednisolone PO** 1 mg/kg BID (max 60 mg/day)
  - When CRP normalizes and patient completed 5 days of PO steroids at 1 mg/kg BID, begin steroid taper:
    - **Prednisone/Prednisolone PO** 1 mg/kg once daily x5 days
    - then 0.5 mg/kg once daily x5 days
    - then stop

Work up and Consults:

- Obtain Cardiology consult and ECHO[2]
- Daily CRP
- If high risk conditions present[1], consult ID

[1]**High Risk Conditions:**

- <6 months age
- Positive echocardiogram[2]
- Kawasaki Shock syndrome
- Prior history of Kawasaki Disease

[2]**ECHO is positive if any of these 3 conditions are met:**

- Z score of Left Anterior Descending (LAD) or Right Coronary Artery (RCA) ≥2.5
- Coronary artery aneurysm is observed
- ≥3 other suggestive features present (in discussion with Cardiology)

Fever recurs ≥36 hours after IVIG infusion is completed?

- No → Discharge home if criteria met
- Yes →

Treat:

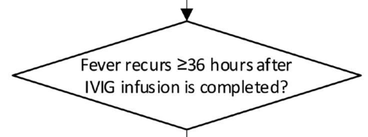
- Repeat **IVIG** 2 g/kg (max 100 g/dose) x1 dose
- Continue **Aspirin PO**
- Continue steroids, if already begun due to presence of high risk conditions[1]
- Consider starting steroids (dosing as above) if not already receiving

Consult

- Consider ID/Rheum consult

Labs:

- Daily CRP



Fever recurs ≥36 hours after IVIG infusion is completed?

- Yes → Consult ID/Rheum
- No → Discharge home

Discharge Criteria

- Afebrile x36 hours, well hydrated without need for IVFs

Discharge Instructions:

- **Aspirin PO** 3-5 mg/kg daily for about 6-8 weeks (as directed by Cardiology)
- Continue steroid taper, if indicated
- Avoid ibuprofen use while on ASA
- Delay live vaccines for 11 months post IVIG administration. Any live vaccines given within 2 weeks prior to IVIG administration should be repeated 11 months after IVIG dose
- Follow up outpatient with Cardiology in 2 weeks from onset of symptoms, then 6 weeks after disease onset (if ECHO positive[2], sooner follow up to be determined by Cardiology)
- Follow up with Rheumatology in 1-2 weeks if CRP remains elevated, or if child is sent home on steroids
- Follow up with Infectious Disease if needed
- PCP follow up within 2-3 days

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CLINICAL PATHWAY:

Kawasaki Disease and Incomplete Kawasaki Disease

THIS PATHWAY SERVES AS A GUIDE AND DOES NOT REPLACE CLINICAL JUDGMENT.

Incomplete Kawasaki

Fever ≥5 days **AND** *only* 2-3 of the following **clinical criteria**:

1. Bilateral bulbar non-exudative conjunctivitis
2. Mucosal changes (red cracked lips, strawberry tongue, erythema of oral and pharyngeal mucosa)
3. Polymorphous rash
4. Extremity changes (swelling and/or erythema, peeling occurs in convalescent phase)
5. Cervical adenopathy (≥ 1.5 cm diameter, usually unilateral)

OR

Infant with fever x7 days without source

Exclusion Criteria:

<2 mo old, exudative conjunctivitis, exudative pharyngitis, oral ulcers, bullous or vesicular rash, generalized adenopathy, splenomegaly, suspicion of Multi-System Inflammatory Disorder/MIS-C (follow the MIS-C Clinical Pathway), signs and symptoms can be easily explained by another condition

Initial Evaluation:

- CBC w diff, CRP, ESR, liver panel (without coags)
- UA with microscopy (clean catch or bag specimen)
- *Consider*: chem 7, blood culture, adenovirus, rapid strep

CRP <3 mg/dL and ESR <40 mm/hr

- Consider discharge
  - Follow up with the primary care physician day after discharge
- If patient warrants admission, admit to Hospital Medicine Service

- Consider daily CRP, ESR
- Closely monitor fevers
- Reassess for additional clinical criteria for Kawasaki or alternate diagnosis (see inclusion criteria on Kawasaki Pathway)

CRP >3 mg/dL and ESR >40 mm/hr, or meets additional Kawasaki disease criteria?

- Yes → Obtain ECHO and Cardiology consult. If positive[2], treat[3] (Refer to Kawasaki Pathway)
- No → Kawasaki unlikely. No treatment needed.

CRP ≥3 mg/dL and/or ESR ≥40 mm/hr

- Admit to Hospital Medicine Service
- Consider Infectious Diseases (ID) consult

<3 supplemental lab criteria[1] → Obtain ECHO and Cardiology consult → **ECHO positive?[2]**

- Yes → Treat[3] (Refer to Kawasaki Pathway)
- No → **Fever resolved?**
  - Yes → Kawasaki unlikely. No treatment needed.
  - No →
    - Consult Rheumatology
    - Consult ID if not already involved

≥3 supplemental lab criteria[1] →

- Treat[3] (Refer to Kawasaki Pathway)
- Obtain ECHO and Cardiology consult

[1] Supplemental lab criteria:

- Albumin ≤3
- Anemia for age
- ↑ ALT
- WBC ≥15,000
- UA ≥10 WBC
- Platelets ≥450,000 after 7 days of fever

[2] ECHO is positive if any of these 3 conditions are met:

- Z score of Left Anterior Descending (LAD) or Right Coronary Artery (RCA) ≥2.5
- Coronary artery aneurysm is observed
- ≥3 other suggestive features present (in discussion with Cardiology)

[3] Treat:

- **IVIG** 2 g/kg x1 (max 100 g/dose). *Can start IVIG without obtaining ECHO first.*
- Medium dose **Aspirin PO** 30-50 mg/kg/day div q6hr until afebrile x48hr
- If any high risk conditions present[4], consider adding:
  - **Methylprednisolone IV** 1 mg/kg BID (max 60 mg/day) while febrile
  - When afebrile, change to **Prednisone/ Prednisolone PO** 1 mg/kg BID (max 60 mg/day)
  - When CRP normalizes, begin steroid taper with **Prednisone/ Prednisolone PO**:
    - 1 mg/kg once daily x5 days
    - then 0.5 mg/kg once daily x5 days
    - then stop

[4] High Risk Conditions:

- <6 months age
- Positive echocardiogram[2]
- Kawasaki Shock syndrome
- Prior history of Kawasaki Disease

Discharge Criteria

- Afebrile x36 hours, well hydrated without need for IVFs

Discharge Instructions:

- **Aspirin PO** 3-5 mg/kg daily for about 6-8 weeks (as directed by Cardiology)
- Continue steroid taper, if indicated
- Avoid ibuprofen use while on ASA
- Delay live vaccines for 11 months post IVIG administration. Any live vaccines given within 2 weeks prior to IVIG administration should be repeated 11 months after IVIG dose
- Follow up outpatient with Cardiology in 2 weeks from onset of symptoms, then 6 weeks after disease onset (if ECHO positive[2], sooner follow up to be determined by Cardiology)
- Follow up with Rheumatology in 1-2 weeks if CRP remains elevated, or if child is sent home on steroids
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