Kawasaki Disease and Incomplete Kawasaki Disease

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What is a Clinical Pathway?

An evidence-based guideline that decreases unnecessary variation and helps promote safe, effective, and consistent patient care.

Objectives of Pathway

- Standardize care of patients with Kawasaki Disease and Incomplete Kawasaki Disease
- Reduce the incidence of coronary artery aneurysms
- Reduce the time to IVIG treatment
- Reduce inpatient length of stay
- If steroids are used, reduce the incidence of refractory Kawasaki Disease

Why is Pathway Necessary?

- Kawasaki Disease is one of the most common vasculitides of childhood, and is the most common cause of acquired heart disease in children in developed countries
- Estimated annual incidence: 20 per 100,000 children younger than five years in the United States
  - Prevalence is higher in children of Japanese or East Asian descent
- Complications such as coronary artery aneurysms, myocardial dysfunction, and heart failure may develop and lead to significant morbidity and mortality
- Given the high risk of delayed diagnosis and/or treatment, it is imperative to standardize care to expedite recognition and timely treatment of Kawasaki Disease

Kawasaki Disease Clinical Features

Fever for at least 4 days **and** *at least* 4/5 of the following clinical criteria

OR

Fever for at least 5 days **and** *at least* 2/5 of the following clinical criteria:

- Bilateral bulbar non-exudative conjunctivitis (typically limbic sparing)
- Mucosal changes (red cracked lips, strawberry tongue, erythema of oral and pharyngeal mucosa)
- Polymorphous Rash
- Extremity changes (swelling and/or erythema; peeling occurs in convalescent phase)
- Cervical lymphadenopathy of at least 1.5 cm diameter (usually unilateral)

Fevers may have resolved before admission or during hospitalization

See the Kawasaki Pathway (page 1)

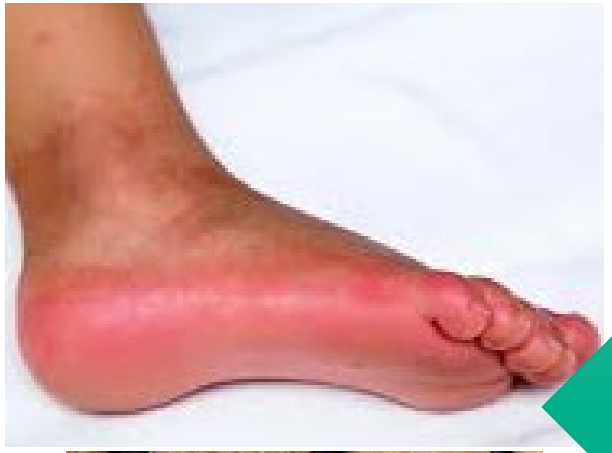
Kawasaki Disease Clinical Features

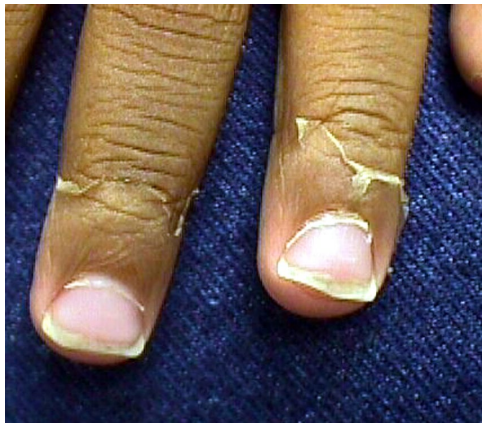
Conjunctivitis:
Bilateral bulbar conjunctival injection without exudate
*typically limbic sparing

Oral Changes:

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

Kawasaki Disease Clinical Features





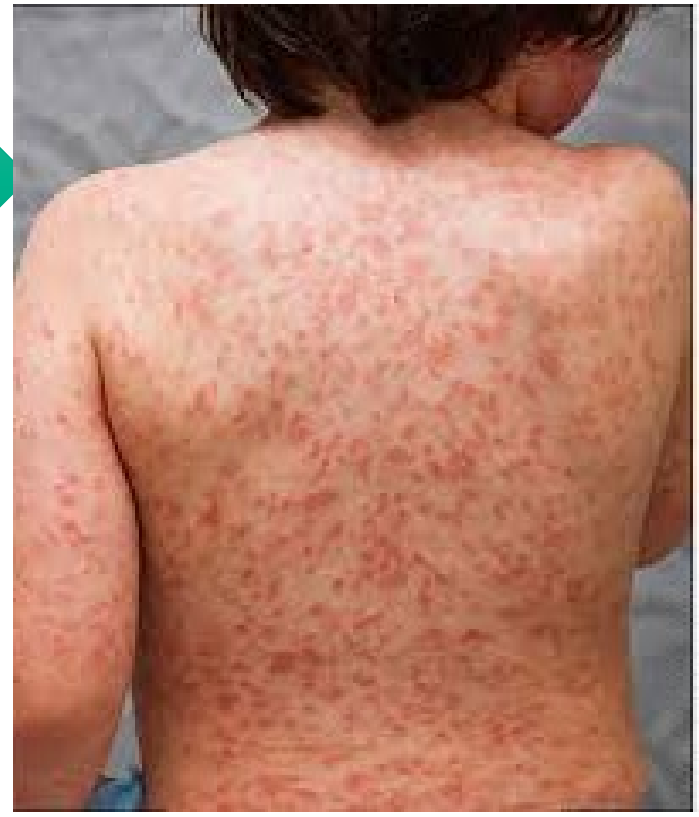
Extremity Changes:

Swelling and/or erythema; or desquamation in convalescent phase

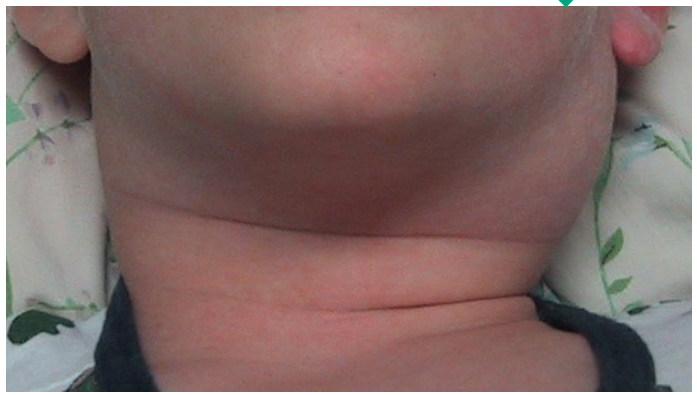
Rash:

Diffuse maculopapular rash

(There are many variations)



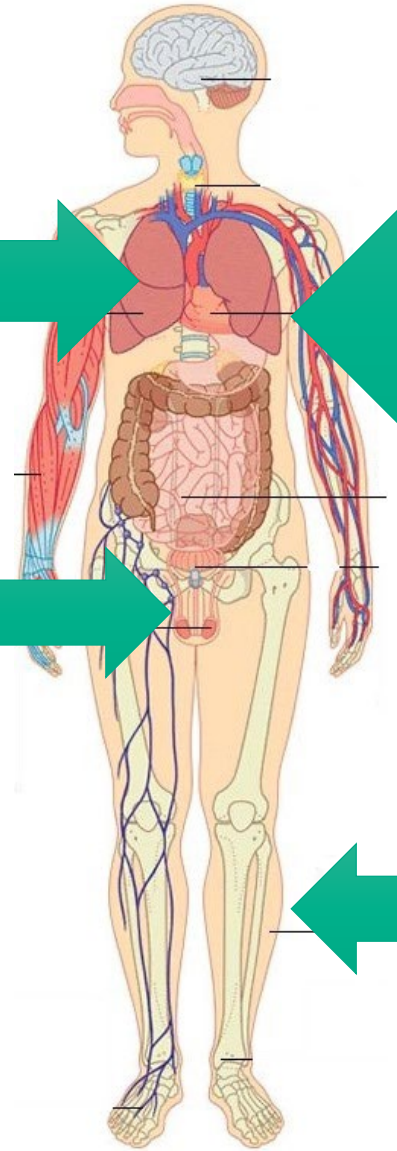
Cervical Lymphadenopathy: at least 1.5cm in diameter



Other Clinical Findings

Respiratory System:
- Peribronchial and interstitial infiltrates
- Pulmonary nodules

Genitourinary:
- Urethritis
- Hydrocele
- Orchitis



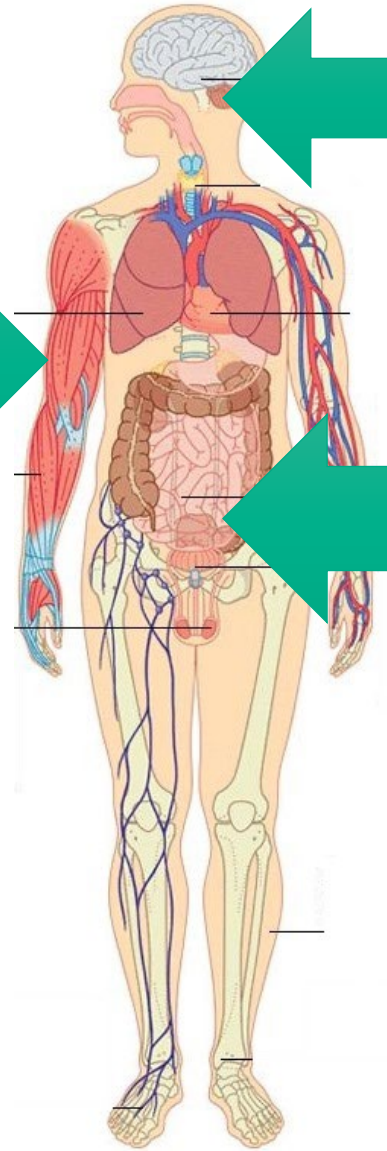
Cardiovascular System:
- Decreased LV ejection fraction (85% patients, generally transient
- Myocarditis (50-70% patients), pericarditis, shock
- Valvular dysfunction (25% patients), typically mitral valve
- Aneurysms of non-coronary arteries
- Peripheral gangrene
- Aortic root enlargement
- Small pericardial effusion
- EKG changes (prolonged PR interval, low voltage, non-specic ST and T wave changes

Skin:
- Erythema and induration at a previous BCG vaccine site

Other Clinical Findings

Musculoskeletal System:

- Arthritis, arthralgia (arthrocentesis will show aseptic purulent fluid with WBCs 125,000 to 300,000 per mm^3 but normal glucose)



Nervous System:

- Irritability/Encephalopathy
- Aseptic meningitis in about 30% of kids
- Transient Facial nerve palsy
- Temporary Sensorineural hearing loss in 1 of 5 kids; rarely permanent

Gastrointestinal:

- Abdominal pain, diarrhea, vomiting
- Hepatitis, jaundice
- Pancreatitis
- Gallbladder hydrops
- Splenomegaly is NOT seen in Kawasaki

Diagnosis: Incomplete Kawasaki

- Presentation of KD is not always classic
- Children who should be further evaluated for incomplete KD include:
  - Children with at least 5 days of fever but have *only* 2 or 3 clinical signs of KD, **or**
  - Infants with fever for 7 days or more with no identified source

Fevers may have resolved before admission or during hospitalization

See the Incomplete Kawasaki Pathway (page 2)

2026 Updates

- Fevers may have resolved before admission (or during hospitalization). Count the total number of days to determine if a patient meets criteria for Kawasaki or Incomplete Kawasaki
- Bilateral bulbar non-exudative conjunctivitis is typically limbic sparing
- Aspirin should not be started within 2 weeks of an influenza infection or a recent varicella infection (increased risk of Reye Syndrome)
- CRP monitoring does not need to be done daily. It would be necessary to have one at least prior to discharge, to help guide the steroid taper
- Discharge can be considered if the patient is afebrile for 24-36 hours *and* there is improvement in clinical status.
- Aspirin should be continued for 6-8 weeks

This is the Kawasaki Disease and Incomplete Kawasaki Disease Clinical Pathway.

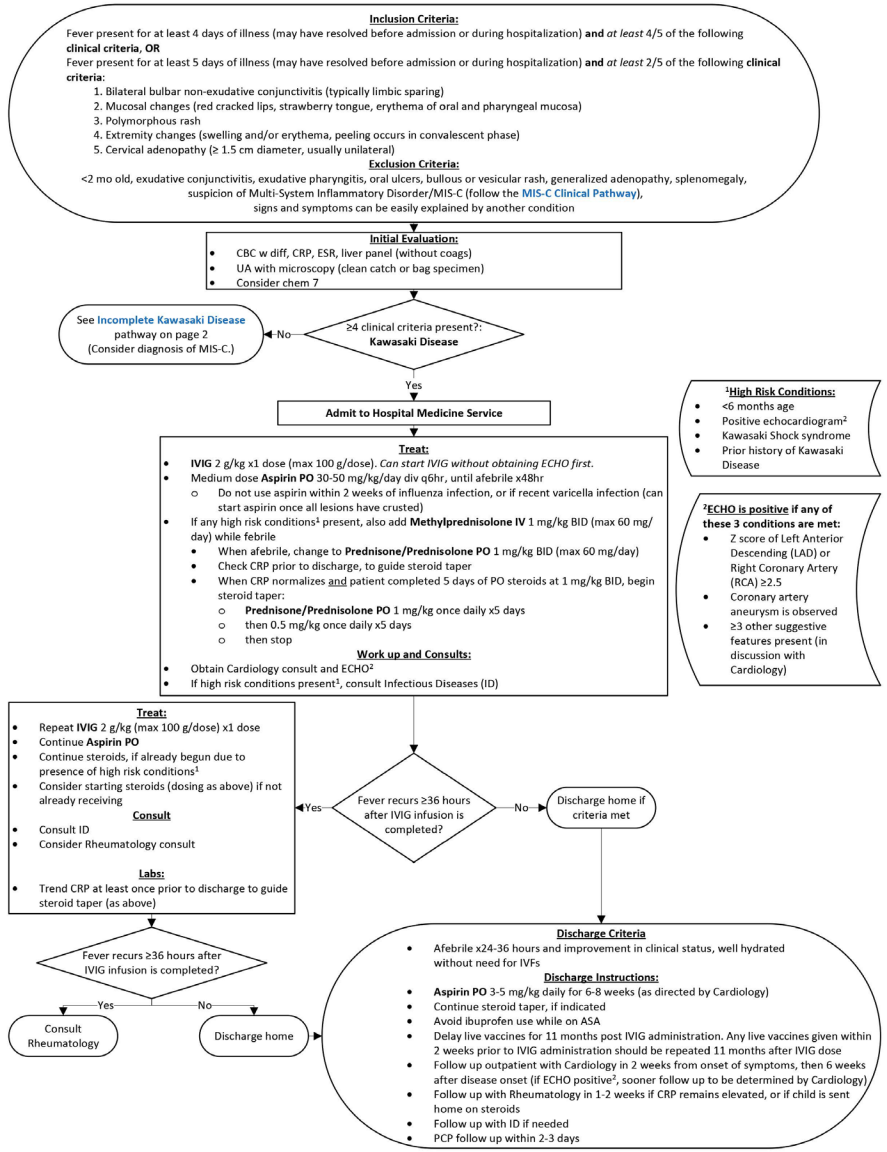
It is comprised of Kawasaki Disease (page 1) and Incomplete Kawasaki Disease (page 2).

We will be reviewing each component in the following slides.

CLINICAL PATHWAY:

Kawasaki Disease and Incomplete Kawasaki Disease

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



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Kawasaki Disease

Suspicion for Kawasaki disease includes the listed criteria.

2026 updates:

- Fevers may have resolved before admission (or during hospitalization). Count the total number of days.
- Bilateral bulbar non-exudative conjunctivitis is typically limbic sparing

Send screening labs and if 4 or more criteria for Kawasaki is present, proceed with the pathway. Otherwise, follow page 2 for Incomplete Kawasaki.

CLINICAL PATHWAY:
Kawasaki Disease and Incomplete Kawasaki Disease

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2026 Update

Inclusion Criteria:

Fever present for at least 4 days of illness (may have resolved before admission or during hospitalization) **and** *at least* 4/5 of the following **clinical criteria**, **OR**

Fever present for at least 5 days of illness (may have resolved before admission or during hospitalization) **and** *at least* 2/5 of the following **clinical criteria**:

1. Bilateral bulbar non-exudative conjunctivitis (typically limbic sparing) 2026 Update
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

start aspirin once all lesions have crusted)

Initial Evaluation:

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

(max 60 mg/

50 mg/day)

/kg BID, begin

then stop

Work up and Consults:

Cardiology consult and ECHO[2]

ons present[1], consult Infectious Diseases (ID)

[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)

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

No → See **Incomplete Kawasaki Disease** pathway on page 2 (Consider diagnosis of MIS-C.)

Yes →

- Continue
  presence of hig
- Consider starting steroids (dosing as above) if not already receiving

Consult

- Consult ID
- Consider Rheumatology consult

Labs:

- Trend CRP at least once prior to discharge to guide steroid taper (as above)

Fever recurs ≥36 hours after IVIG infusion is completed?

Yes → Consult Rheumatology

No → Discharge home

Fever recurs ≥36 hours after IVIG infusion is completed?

Yes → (see above)

No → Discharge home if criteria met

Discharge Criteria

- Afebrile x24-36 hours and improvement in clinical status, well hydrated without need for IVFs

Discharge Instructions:

- **Aspirin PO** 3-5 mg/kg daily for 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 ID if needed
- PCP follow up within 2-3 days

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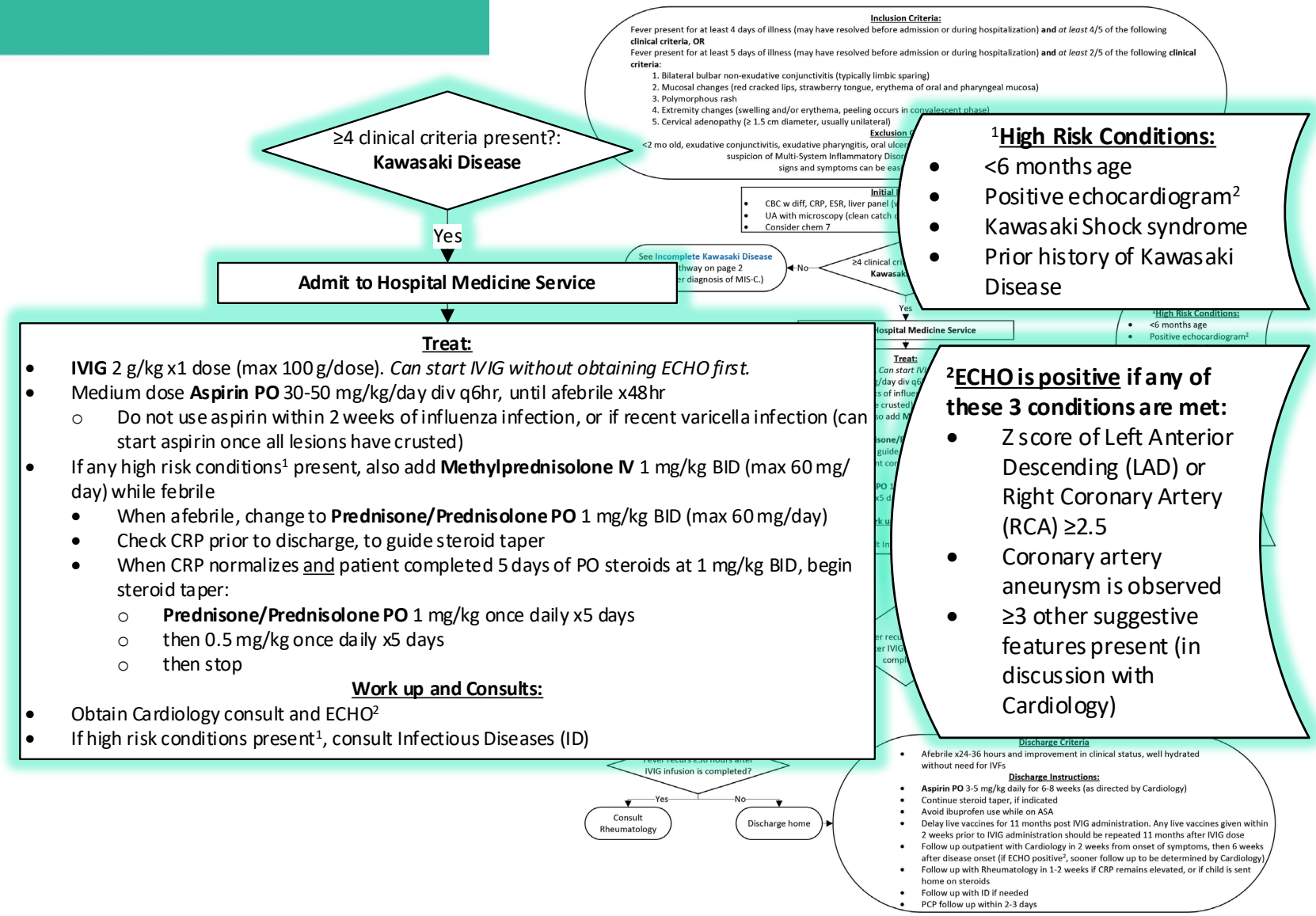
Kawasaki Disease

Goals of therapy are to:

- Reduce systemic inflammatory process
- Prevent coronary artery abnormalities
- If coronary artery abnormalities are present, then to minimize the peak dimension and any clots

CLINICAL PATHWAY:
Kawasaki Disease and Incomplete Kawasaki Disease

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



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Connecticut Children's

Kawasaki Disease

CLINICAL PATHWAY:
Kawasaki Disease and Incomplete Kawasaki Disease

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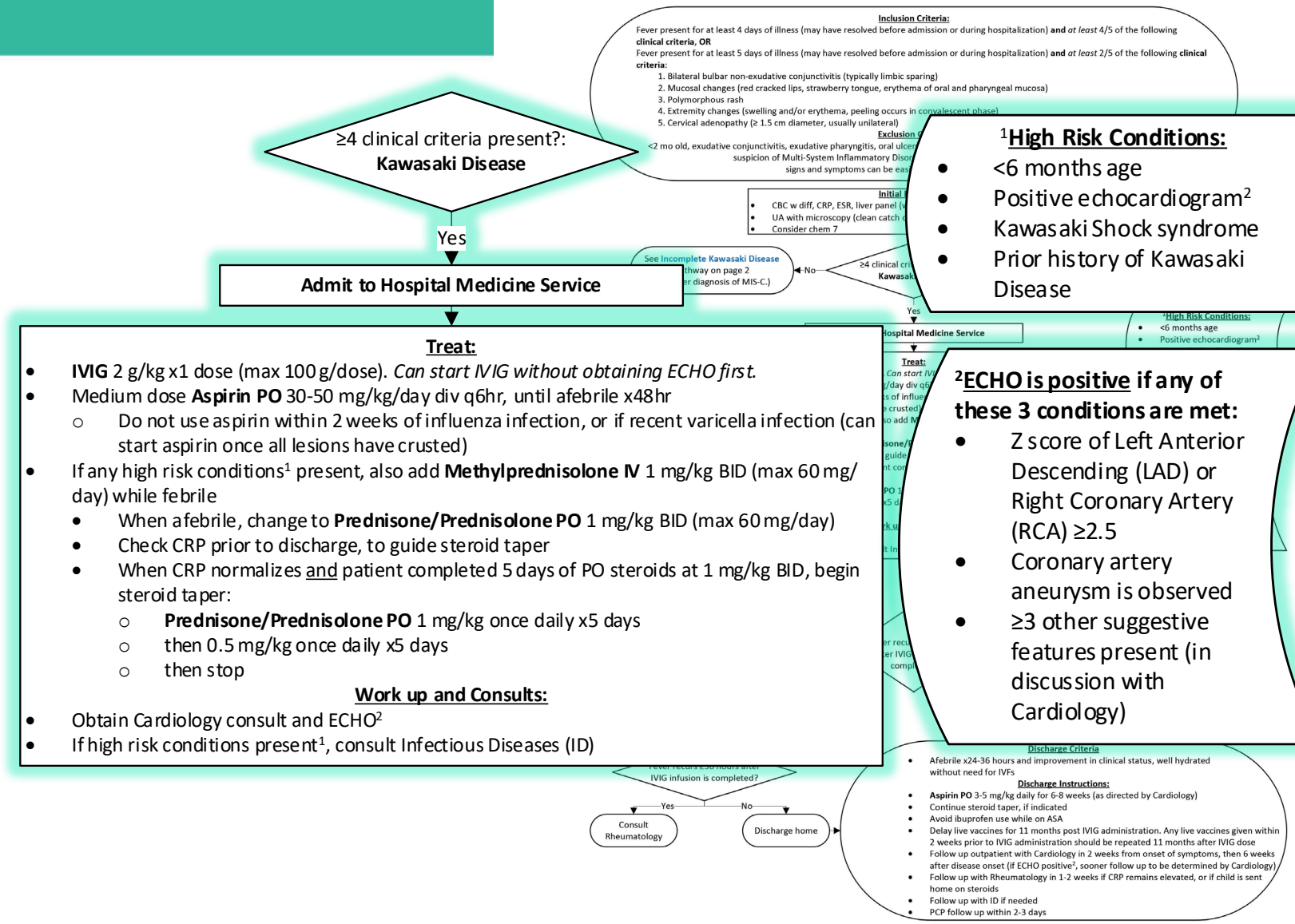
Treatment should not be delayed while the work up continues (e.g., ECHO, consults, etc.).

Cardiology consults are routine, and ECHO may wait until the morning if the patient is admitted overnight.



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Kawasaki Disease

Treatment includes IVIG and medium dose aspirin.

2026 updates:

- Aspirin should not be started within 2 weeks of an influenza infection or a recent varicella infection (increased risk of Reye Syndrome)
- CRP monitoring does not need to be done daily. It would be necessary to have one at least prior to discharge, to help guide the steroid taper

2026 Update

2026 Update

CLINICAL PATHWAY:
Kawasaki Disease and Incomplete Kawasaki Disease

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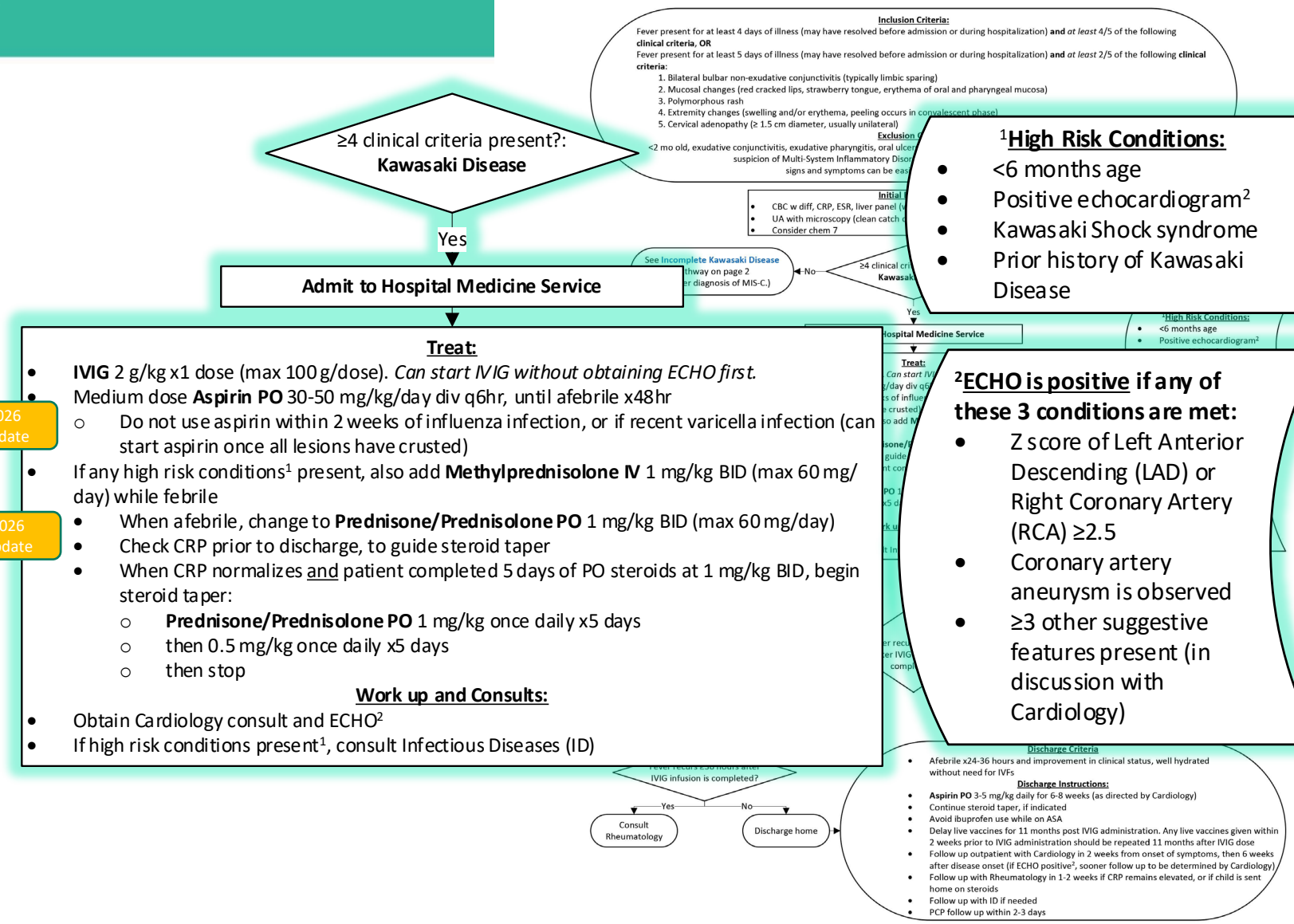

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Connecticut Children's

Kawasaki Disease

CLINICAL PATHWAY:
Kawasaki Disease and Incomplete Kawasaki Disease

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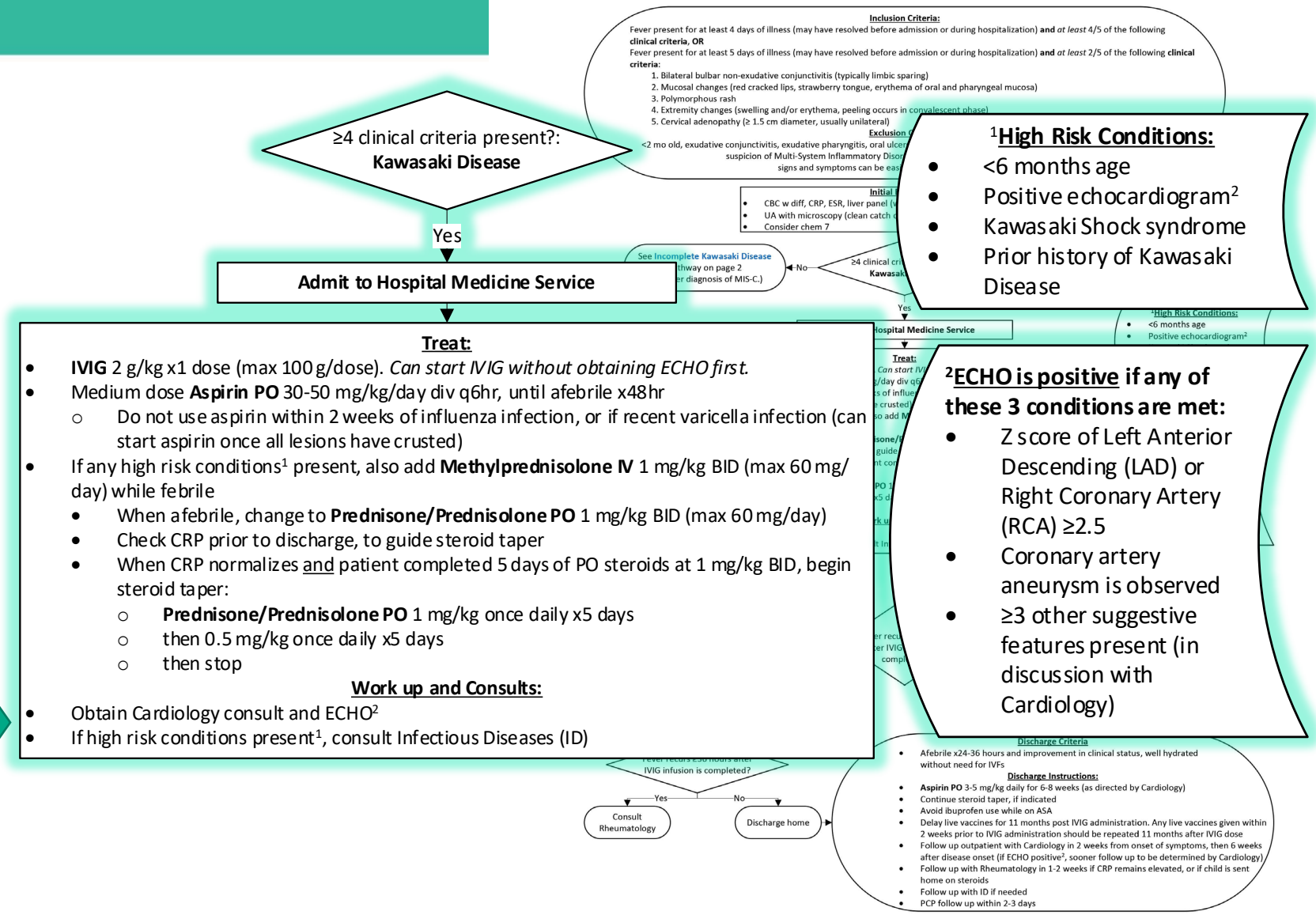
All patients should have cardiology consulted.

If any high risk conditions are present, consult ID and add steroids to the treatment plan.



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Connecticut Children's

Kawasaki Disease

If the fever recurs ≥36 *after* the IVIG infusion is completed, then IVIG may be repeated for a 2nd dose.

2026 updates:

- Consult ID for all of these cases (and consider rheumatology consult).
- CRP does not have to be daily – trend at least once prior to discharge.

If the patient's fever recurs ≥36 hours after the 2nd IVIG infusion is completed, consult rheumatology.

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Kawasaki Disease and Incomplete Kawasaki Disease

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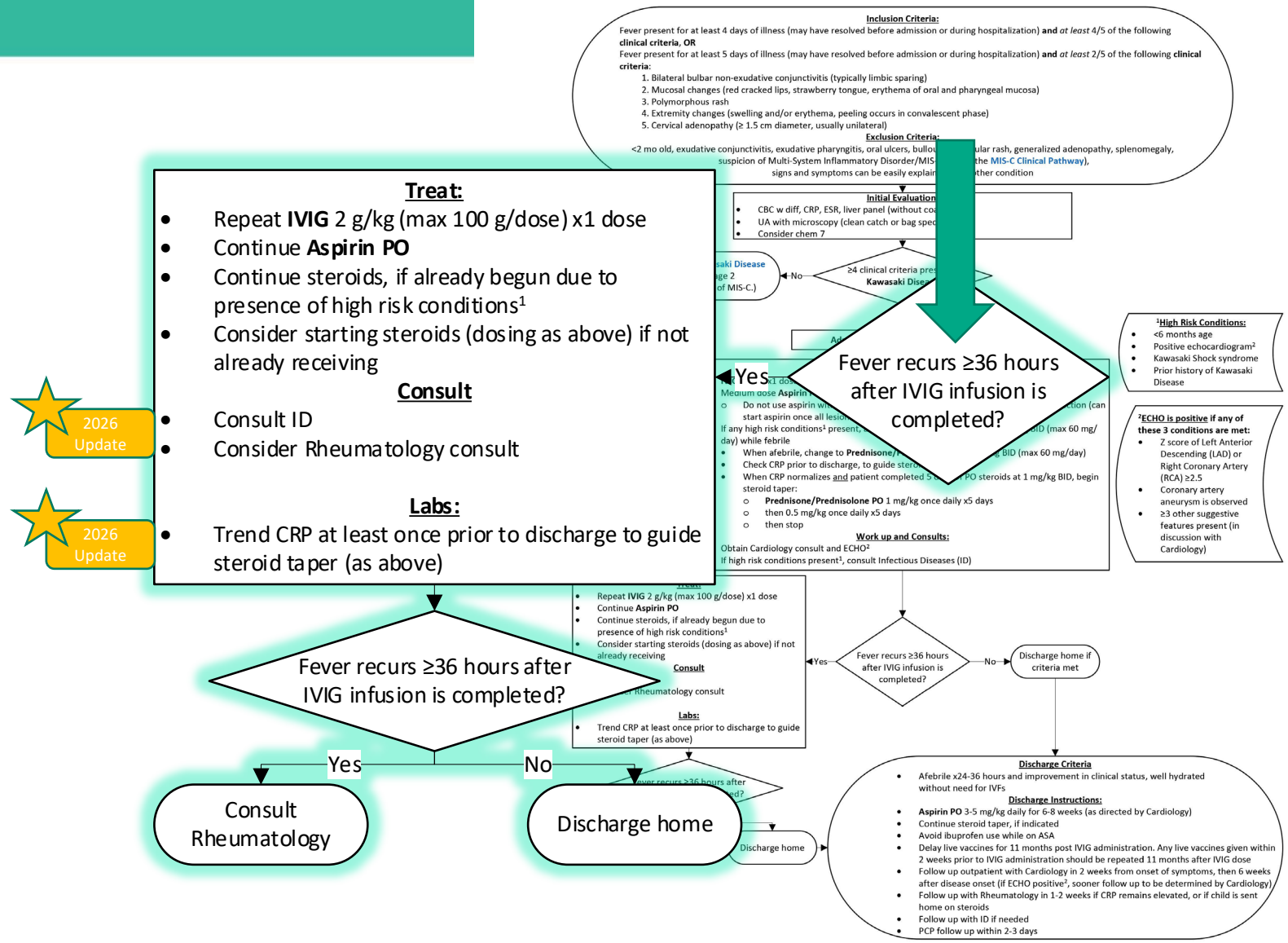

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Connecticut Children's

Kawasaki Disease

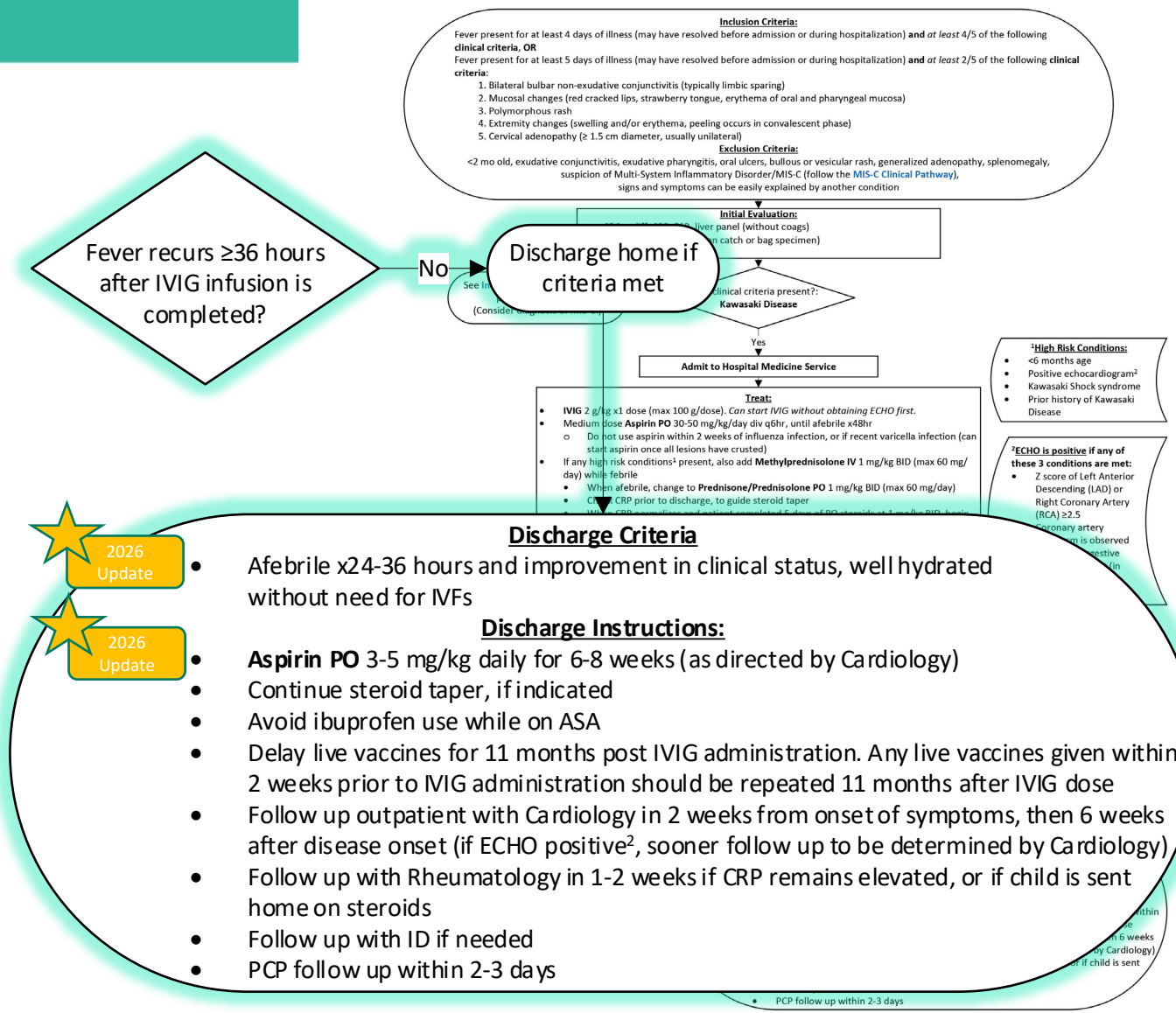
2026 updates:

- Discharge can be considered if the patient is afebrile for 24-36 hours *and* there is improvement in clinical status.
- Aspirin should be continued for 6-8 weeks (not “about 6-8 weeks”).

If the patient is discharged on steroids or the CRP remains elevated, they should follow up with Rheumatology.

Parental education prior to discharge is imperative.

- Emphasize the importance of continuing aspirin and/or steroids as directed, delaying live vaccines, and following up with necessary services.



Incomplete Kawasaki

This is the Incomplete Kawasaki Disease Clinical Pathway.

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Kawasaki Disease and Incomplete Kawasaki Disease

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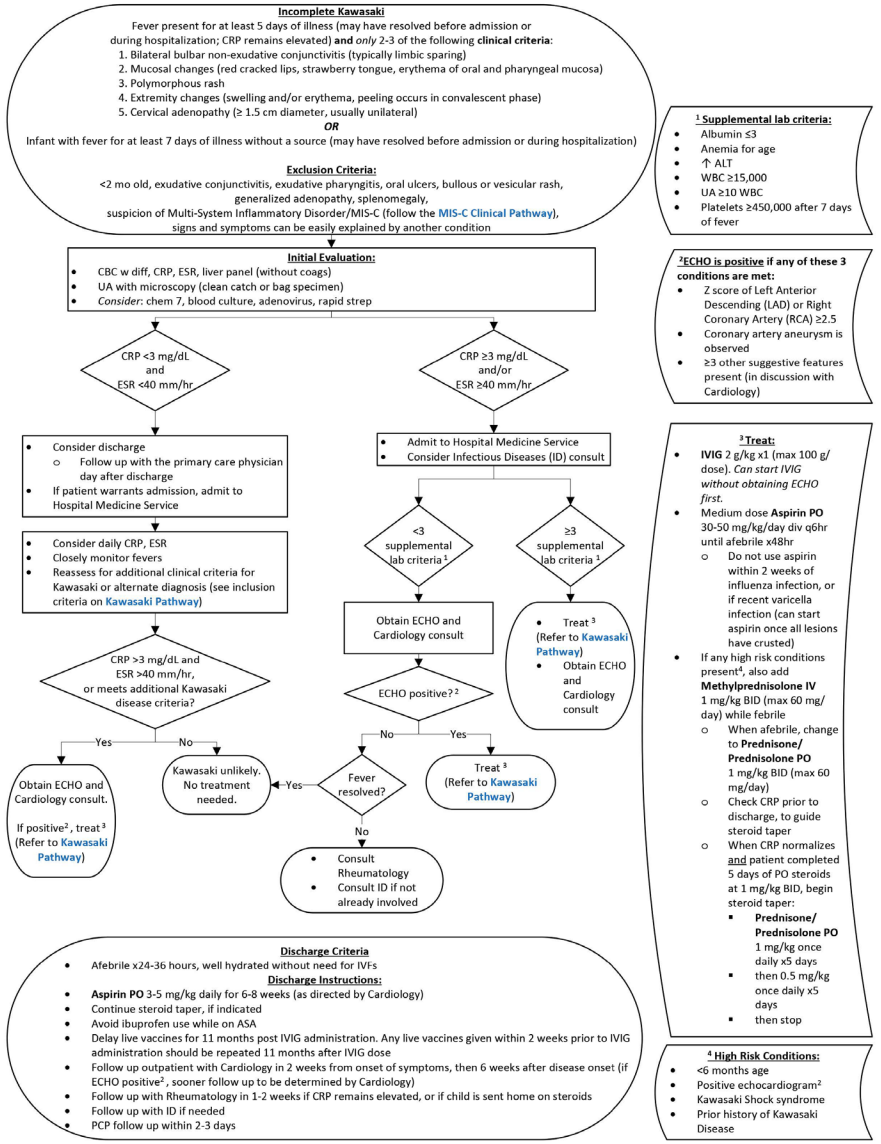

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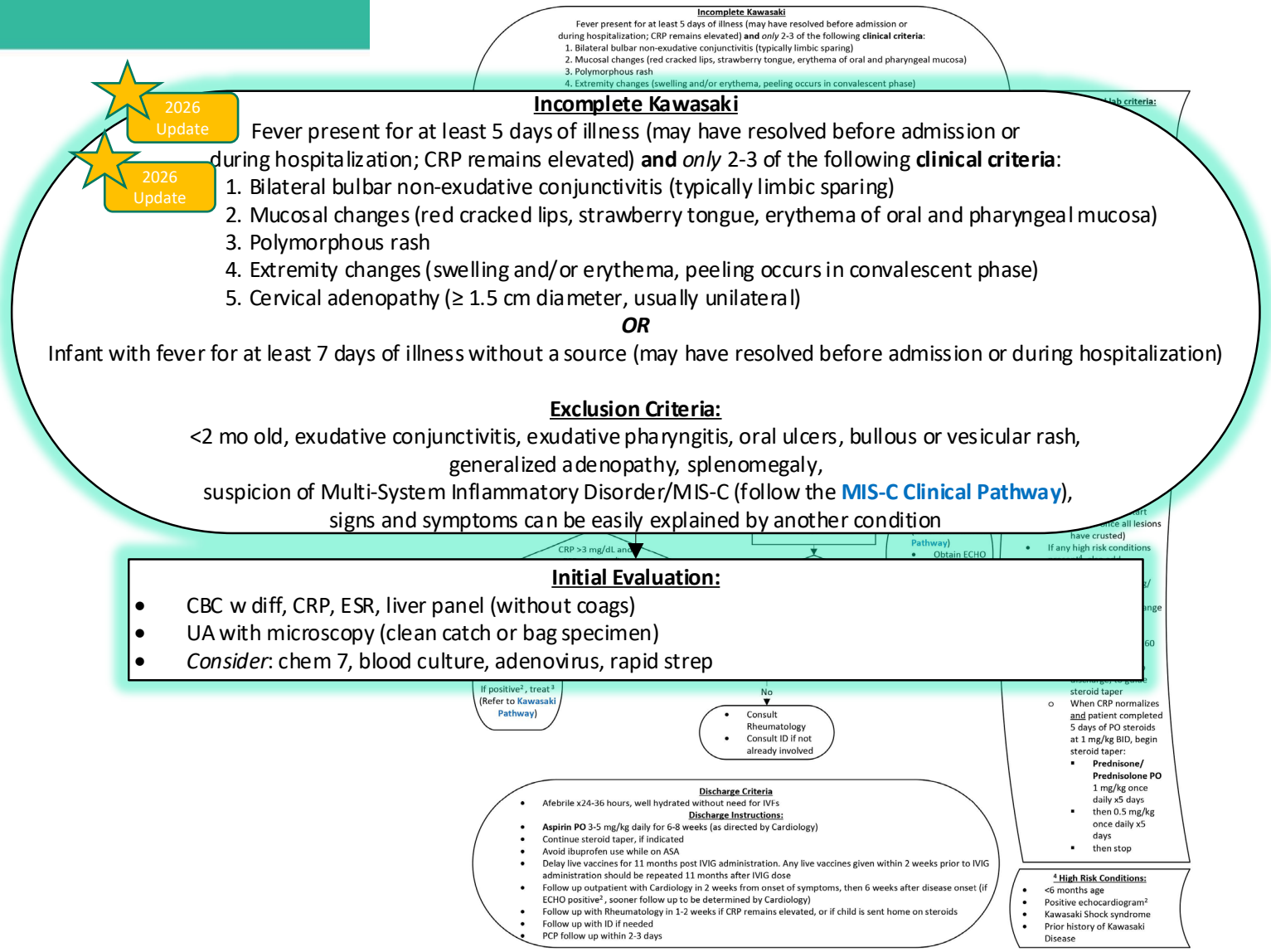
Incomplete Kawasaki

Incomplete Kawasaki Disease only meets 2-3 of the clinical criteria.

2026 updates:

- Fevers may have resolved before admission (or during hospitalization). Count the total number of days.
- Bilateral bulbar non-exudative conjunctivitis is typically limbic sparing

Initial labs will help determine if further work up/treatment is necessary.



Incomplete Kawasaki

Fever present for at least 5 days of illness (may have resolved before admission or during hospitalization; CRP remains elevated) **and** *only* 2-3 of the following **clinical criteria**:

1. Bilateral bulbar non-exudative conjunctivitis (typically limbic sparing)
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)

2026 Update

2026 Update

5. Cervical adenopathy (≥ 1.5 cm diameter, usually unilateral)

OR

Infant with fever for at least 7 days of illness without a source (may have resolved before admission or during hospitalization)

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

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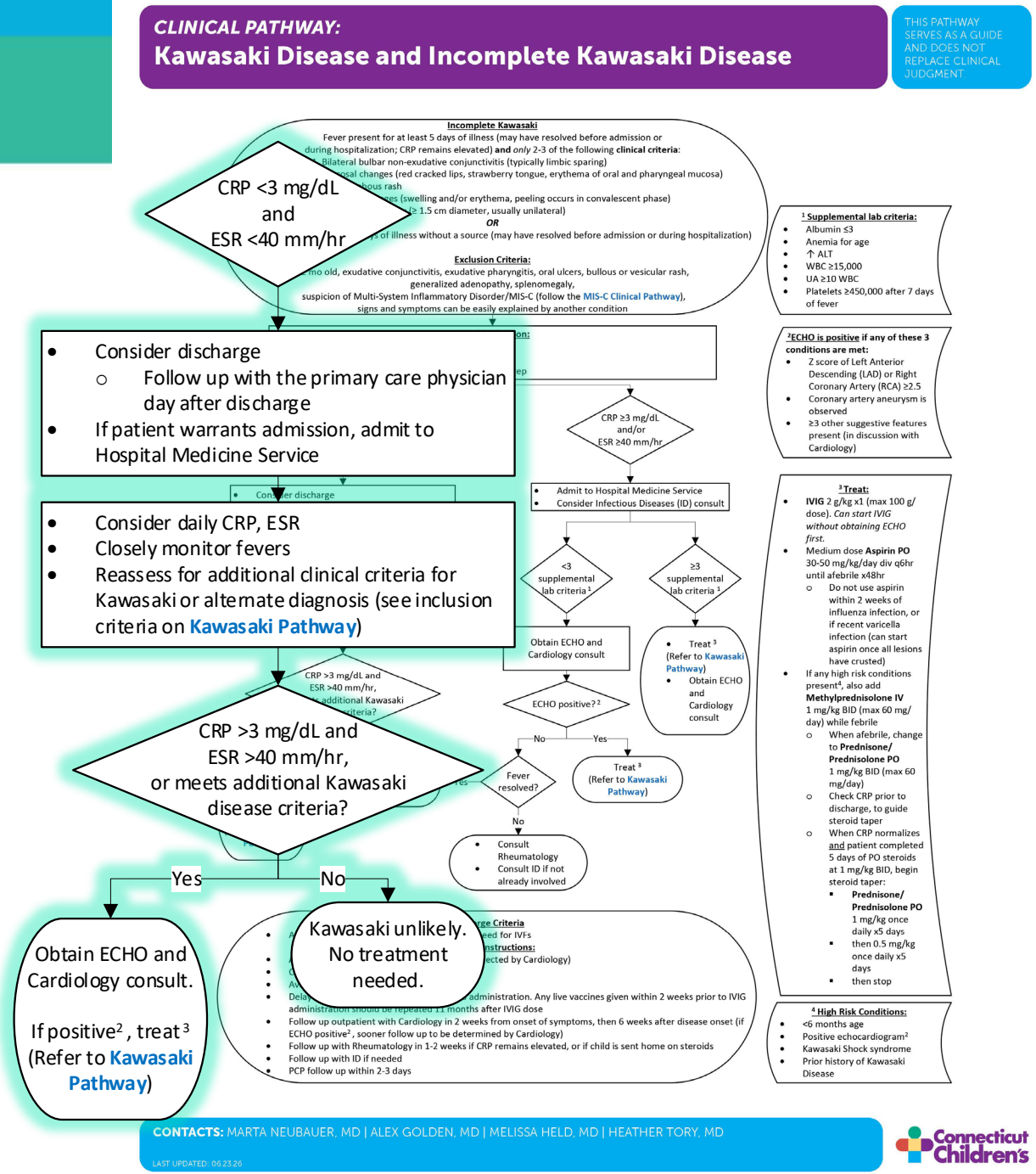
Incomplete Kawasaki

If inflammatory markers are NOT significantly elevated on presentation:

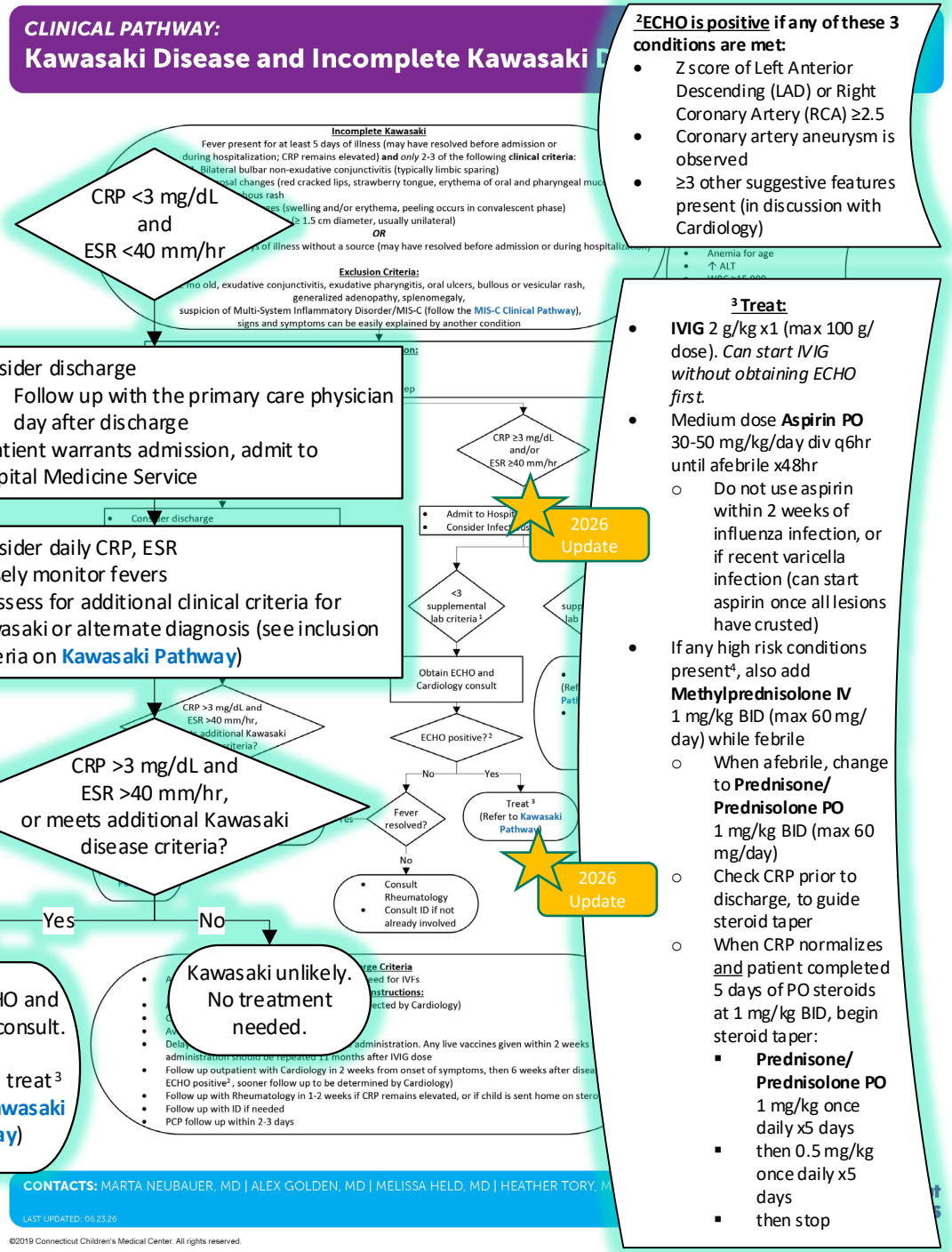
- Patient may be able to be discharged with close follow up with their PCP
- If the patient is admitted, monitoring for symptom progression and fevers should be done.
- If the patient meets Kawasaki criteria, or has elevated inflammatory markers, an ECHO and cardiology consult should be obtained.



Incomplete Kawasaki

If the ECHO is positive, then treatment should be initiated.

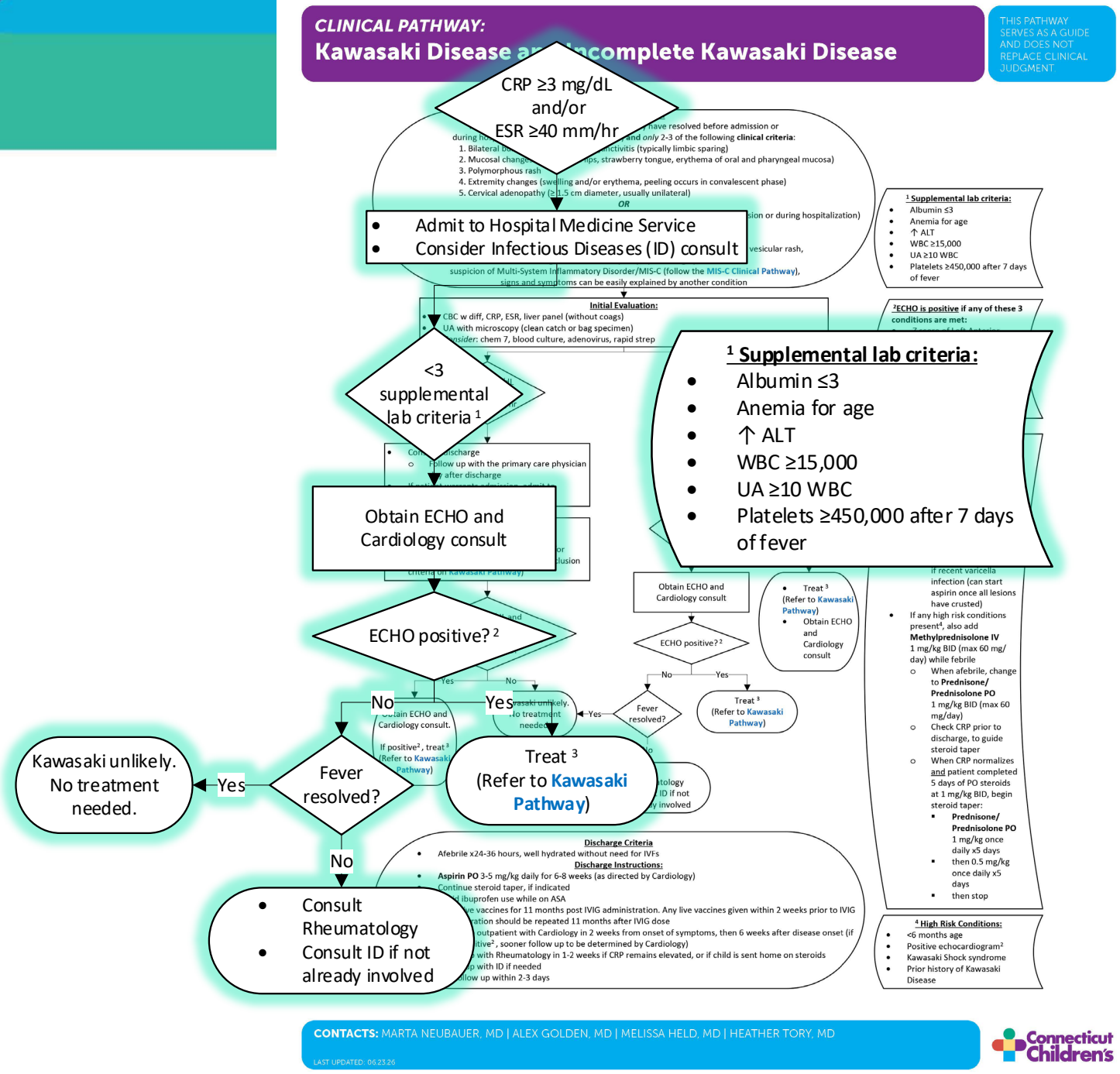
The treatment and recommendations remain the same as Kawasaki Disease.



Incomplete Kawasaki

If inflammatory markers **are** elevated at the time of initial evaluation, the patient should be admitted with consideration for ID consult.

If there are less than 3 supplemental lab criteria, an ECHO will determine the next steps and if treatment is needed.



Incomplete Kawasaki

If inflammatory markers **are** elevated at the time of initial evaluation and there are 3 or more supplemental lab criteria, then treatment and recommendations should follow Kawasaki Disease.

CLINICAL PATHWAY:
Kawasaki Disease and Incomplete Kawasaki Disease

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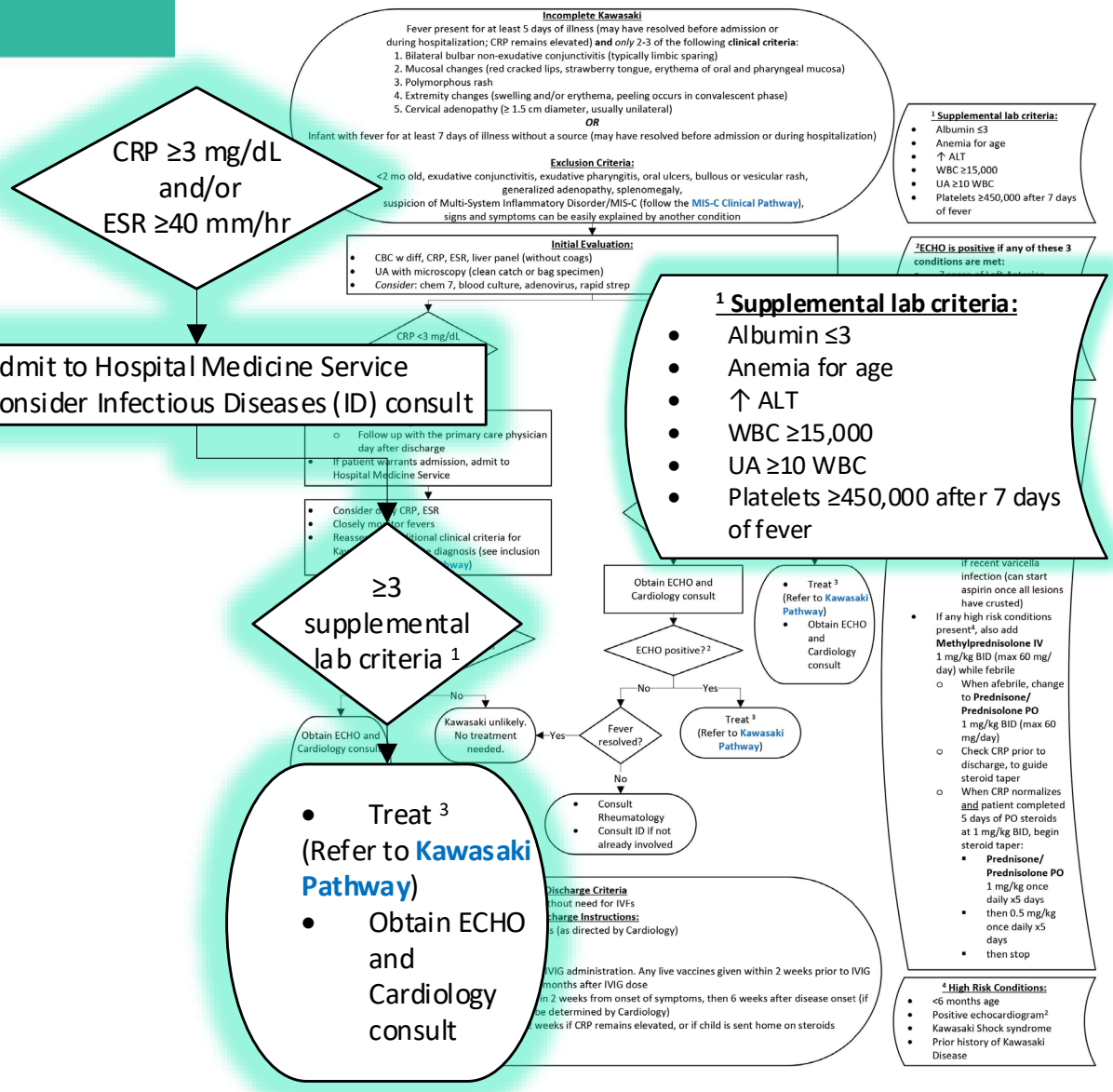

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Incomplete Kawasaki

Discharge criteria and instructions are the same as Kawasaki Disease.

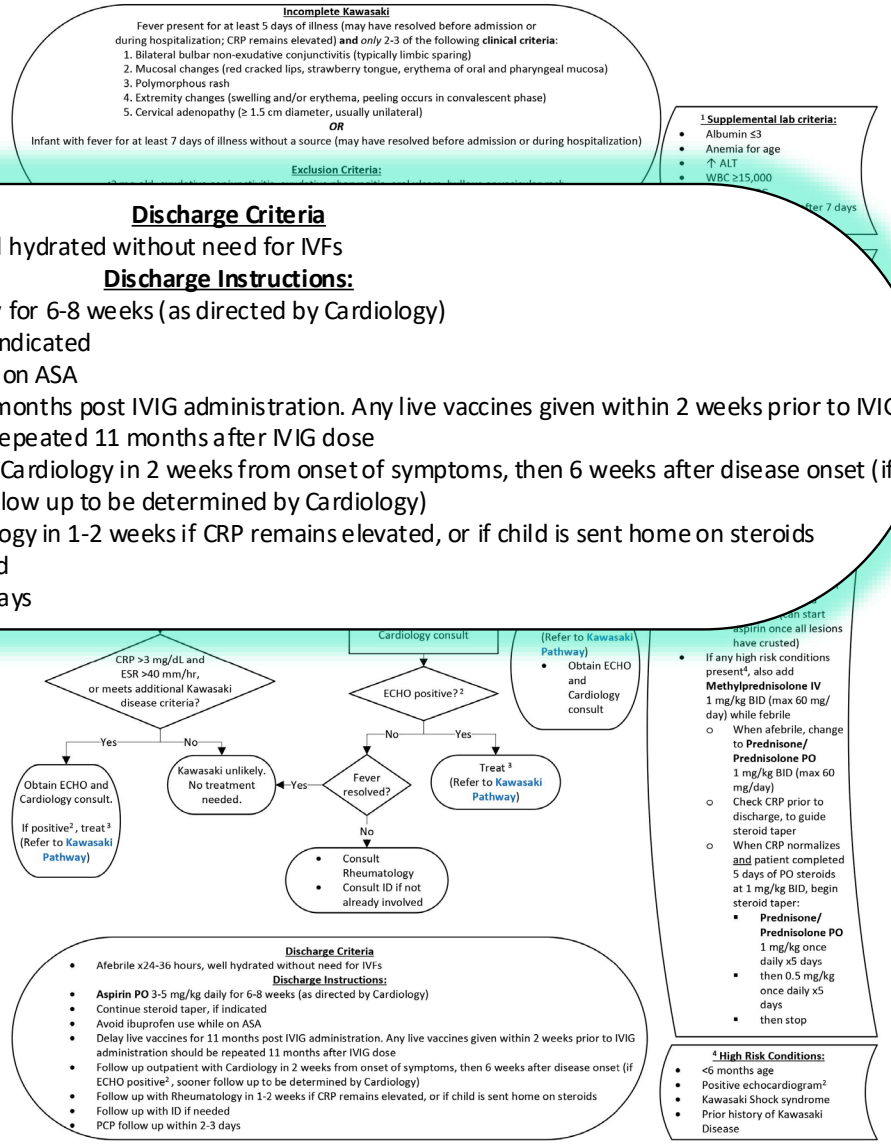
2026 Update

Discharge Criteria

- Afebrile x24-36 hours, well hydrated without need for IVFs

Discharge Instructions:

- **Aspirin PO** 3-5 mg/kg daily for 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 ID if needed
- PCP follow up within 2-3 days



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Review of Key Points

- Kawasaki disease may not present typically
- Fevers may have resolved before admission (or during hospitalization). Count the total number of days.
- Bilateral bulbar non-exudative conjunctivitis is typically limbic sparing
- Initial treatment and work-up includes IVIG, medium dose Aspirin, ECHO, and Cardiology consult
  - IVIG and aspirin should not be delayed while awaiting ECHO and Cardiology consult

Quality Metrics

- Percentage of patients with pathway order set usage
- Average time from admission to time of IVIG administration
- Number of patients with coronary artery aneurysms or ectasia at diagnosis
- Percentage of patients receiving medium dose aspirin in the acute phase of treatment
- Percentage of patients scheduled at discharge for follow up with a cardiologist
- Average length of stay (days)
- Number of patients readmitted due to Kawasaki Disease within 30 days

Pathway Contacts

- Marta Neubauer, MD
  - Pediatric Hospital Medicine
- Melissa Held, MD
  - Pediatric Infectious Diseases
- Heather Tory, MD
  - Pediatric Rheumatology
- Alex Golden, MD
  - Pediatric Cardiology

References

- Abrams JY, Belay ED, Uehara R, Maddox RA, Schonberger LB, Makamura Y. Cardiac Complications, Earlier Treatment, and Initial Disease Severity in Kawasaki Disease. *J Pediatr.* 2017 Sept;188:64-69.
- Baker AL, Lu M, Minich LL, et al. Associated symptoms in the ten days before diagnosis of Kawasaki disease. *J Pediatr.* 2009 Apr; 154(4):592-595.
- Chen S, Dong Y, Kiuchi MG, et al. Coronary Artery Complication in Kawasaki Disease and the Importance of Early Intervention: A Systematic Review and Meta-analysis. *JAMA Pediatr.* 2016 Dec;170(12):1156-1163.
- McCrindle BW, Rowley AH, Newburger JW, et al. Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease: A Scientific Statement for Health Professionals From the American Heart Association. *Circulation.* 2017 Apr;135(17):e927-e999.
- Shao, S., Luo, C., Zhou, K. *et al.* Predictive value of serum procalcitonin for both initial and repeated immunoglobulin resistance in Kawasaki disease: a prospective cohort study. *Pediatr Rheumatol Online J.* 2019 Nov 27;17(1):78. doi:10.1186/s12969-019-0379-5.

Thank You!

About Connecticut Children's Pathways Program

Clinical pathways guide the management of patients to optimize consistent use of evidence-based practice. Clinical pathways have been shown to improve guideline adherence and quality outcomes, while decreasing length of stay and cost. Here at Connecticut Children's, our Clinical Pathways Program aims to deliver evidence-based, high value care to the greatest number of children in a diversity of patient settings. These pathways serve as a guide for providers and do not replace clinical judgment.