

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?

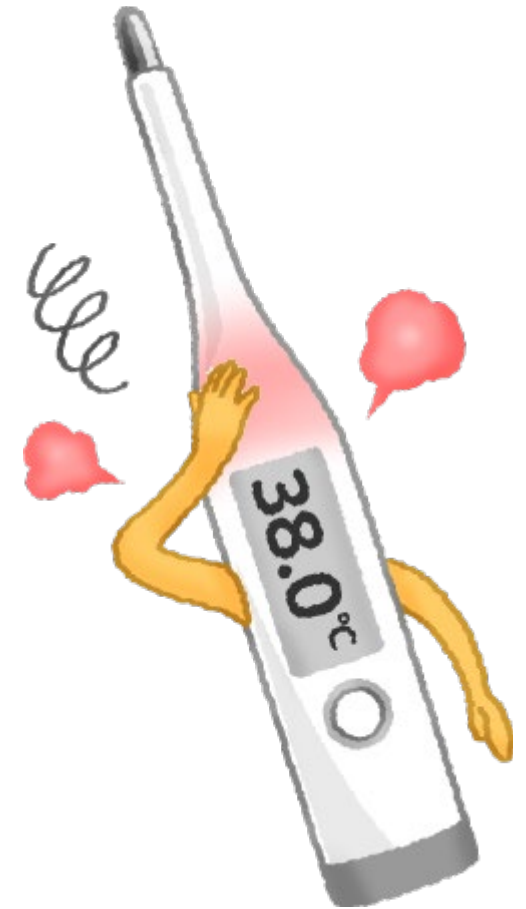


- Kawasaki Disease is one of the most common vasculitides of childhood, and is the most common cause of acquired heart disease in children in developed countries
- Estimated annual incidence of 20 per 100,000 children younger than five years in the United States, and prevalence is higher in children 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

MUST HAVE FEVER for at least 5 days **and**
at least 2/5 of the following clinical criteria:

- Bilateral bulbar non-exudative conjunctivitis
- 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 1.5cm diameter (usually unilateral)



See the [Kawasaki Pathway \(page 1\)](#)

Kawasaki Disease Clinical Features



Conjunctivitis:
Bilateral bulbar
conjunctival
injection without
exudate

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



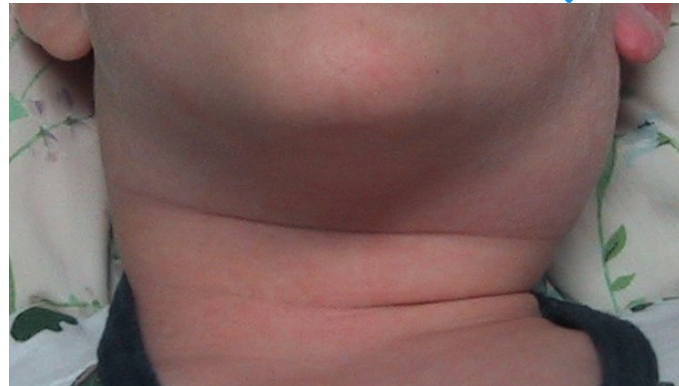
Kawasaki Disease Clinical Features



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:

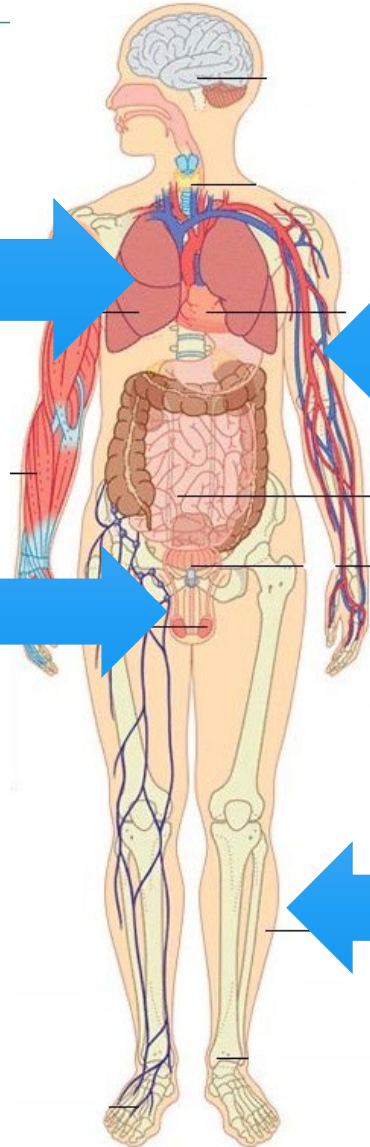
- Urethritis
- Hydrocele
- Orchitis

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-specific ST and T wave changes)

Skin:

- Erythema and induration at a previous BCG vaccine site



Other Clinical Findings

Musculoskeletal System:

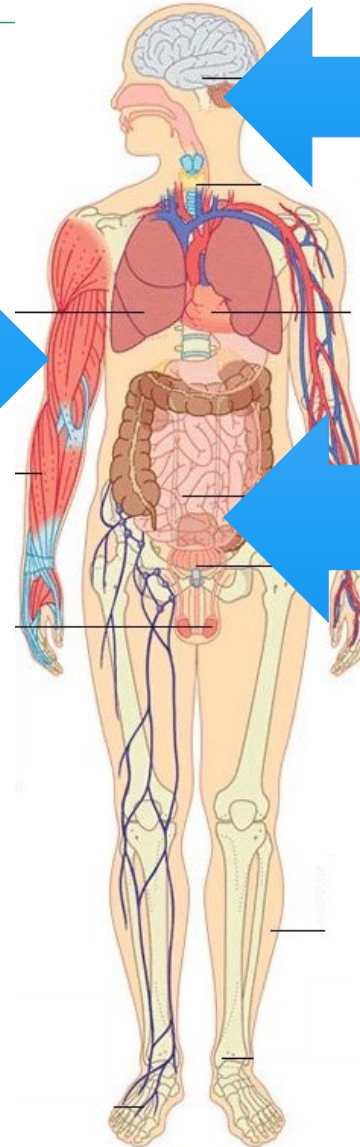
- Arthritis, arthralgia (arthrocentesis will show aseptic purulent fluid with WBCs 125,000 to 300,000 per mm³ but normal glucose)

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

See the [Incomplete Kawasaki Pathway \(page 2\)](#)

This is the Kawasaki Disease Clinical Pathway.

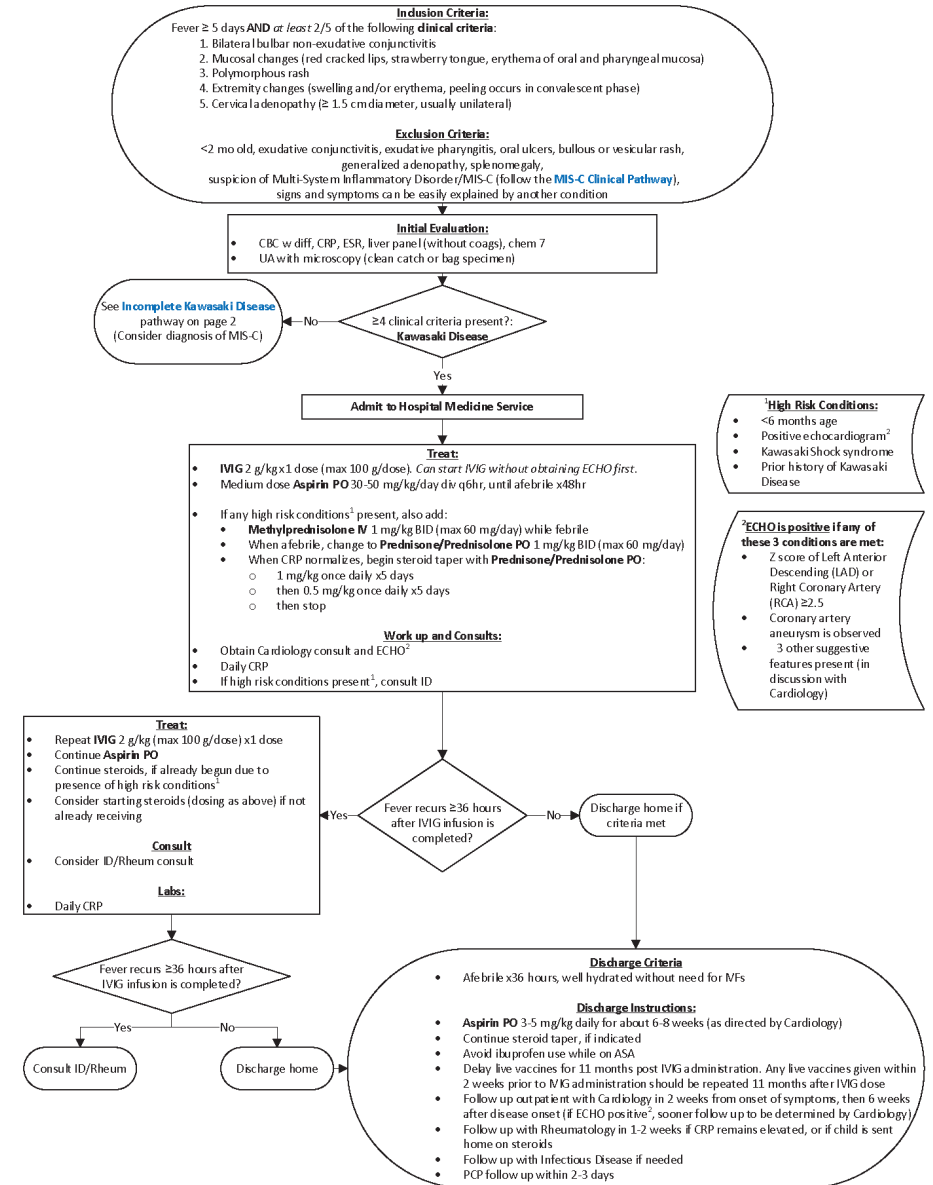
It is comprised of Kawasaki disease (page 1), and Incomplete Kawasaki disease (page 2).

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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CLINICAL PATHWAY: Kawasaki Disease and Incomplete Kawasaki Disease

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If a patient presents with clinical suspicion for **Kawasaki Disease (KD)**, the initial management is a thorough history and physical exam to determine what if any clinical criteria are present.

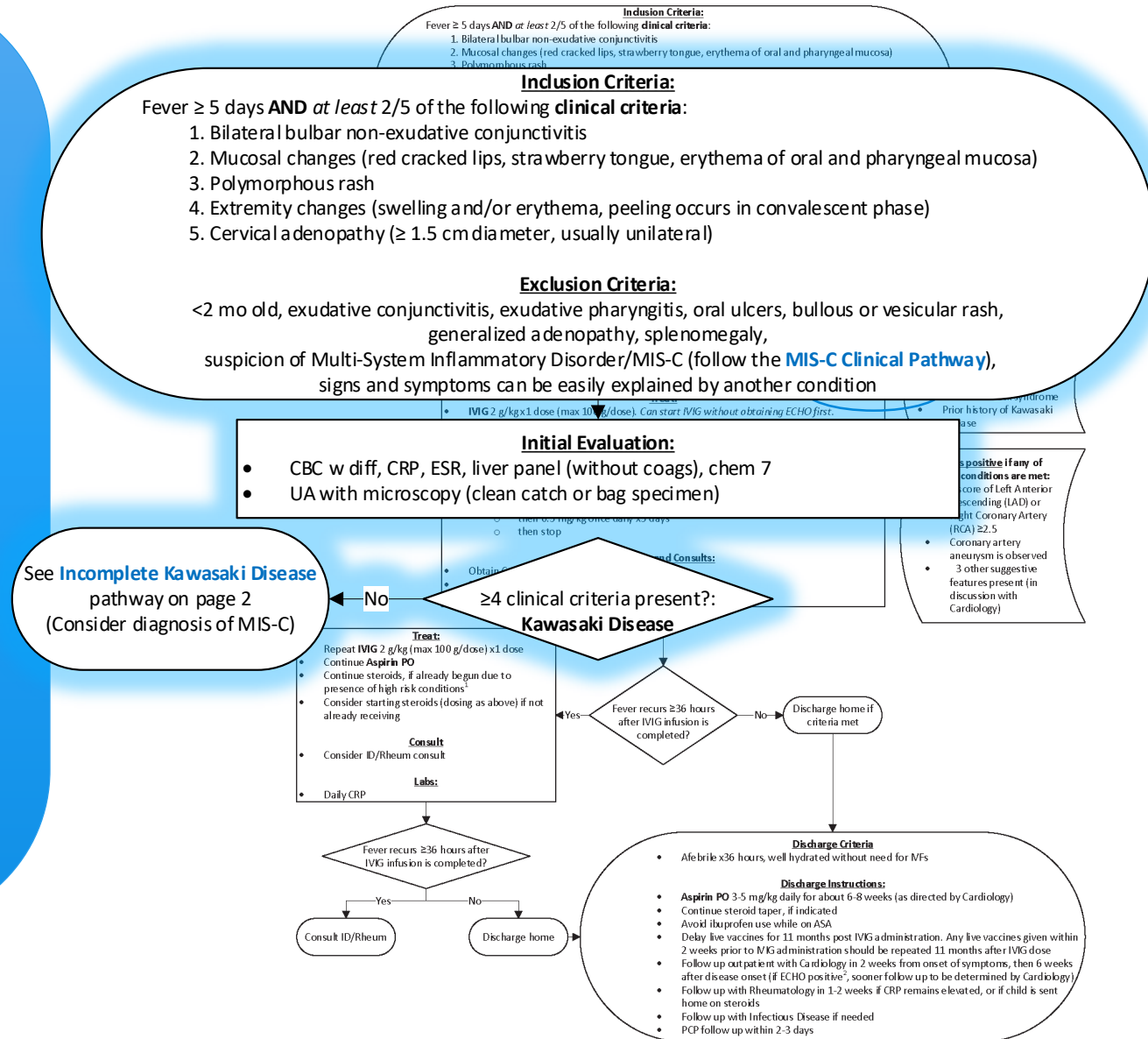
Screening labs should be done.

If a patient matches criteria for KD then you proceed with the main pathway.

If they do not meet criteria for full KD then Go to the **Incomplete Kawasaki Disease Pathway**



See the **Incomplete Kawasaki Disease Pathway** in later slides



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Kawasaki Disease

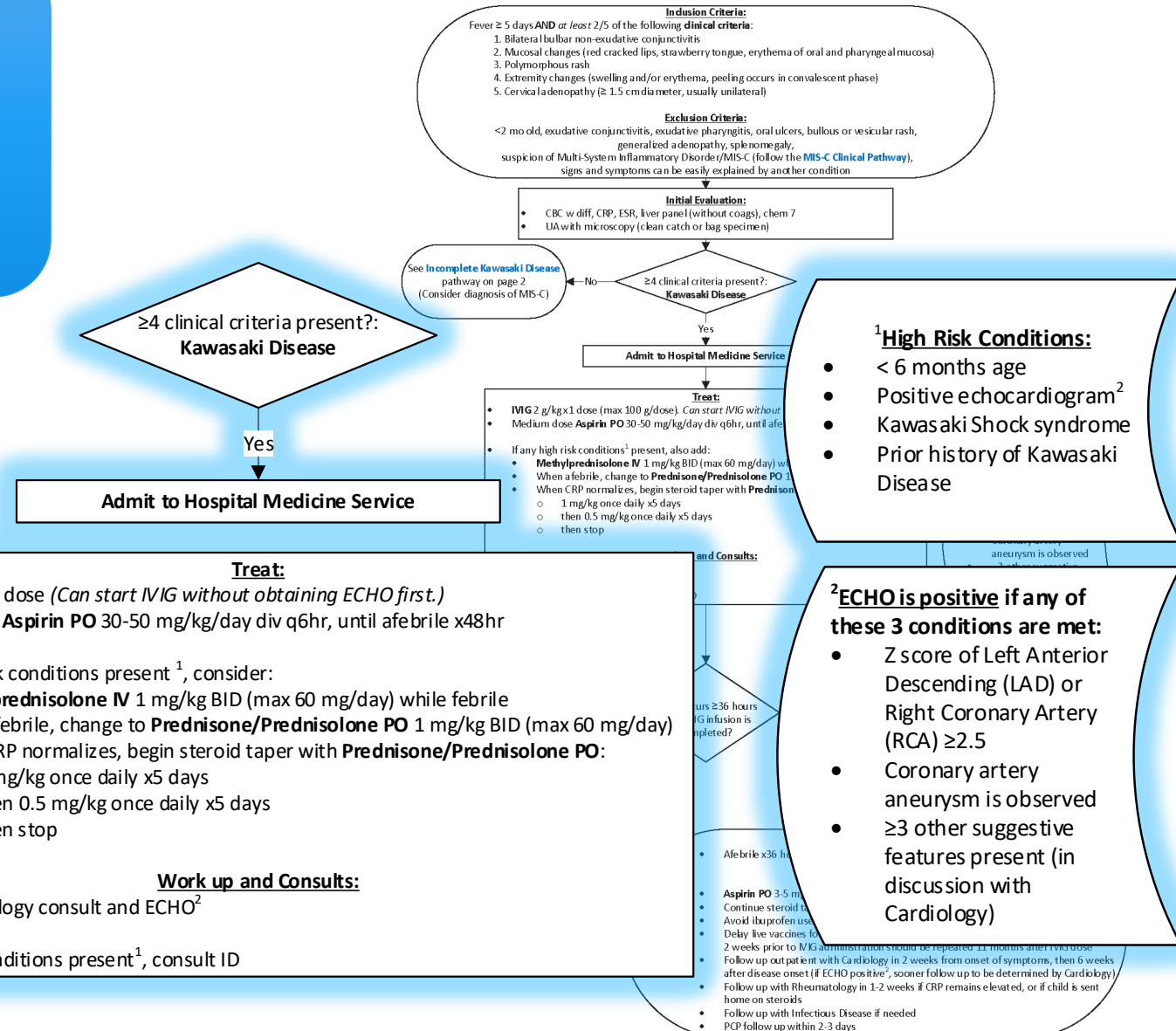
If a patient is identified as meeting criteria for **Kawasaki Disease**, treatment should not be delayed while the work up continues (ECHO, consults, etc.).

Cardiology consults will be routine, and ECHO may wait until the morning if patient admitted overnight

CLINICAL PATHWAY:

Kawasaki Disease and Incomplete Kawasaki Disease

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Kawasaki Disease

Treatment consists of IVIG and medium dose Aspirin

Goals of therapy are to:

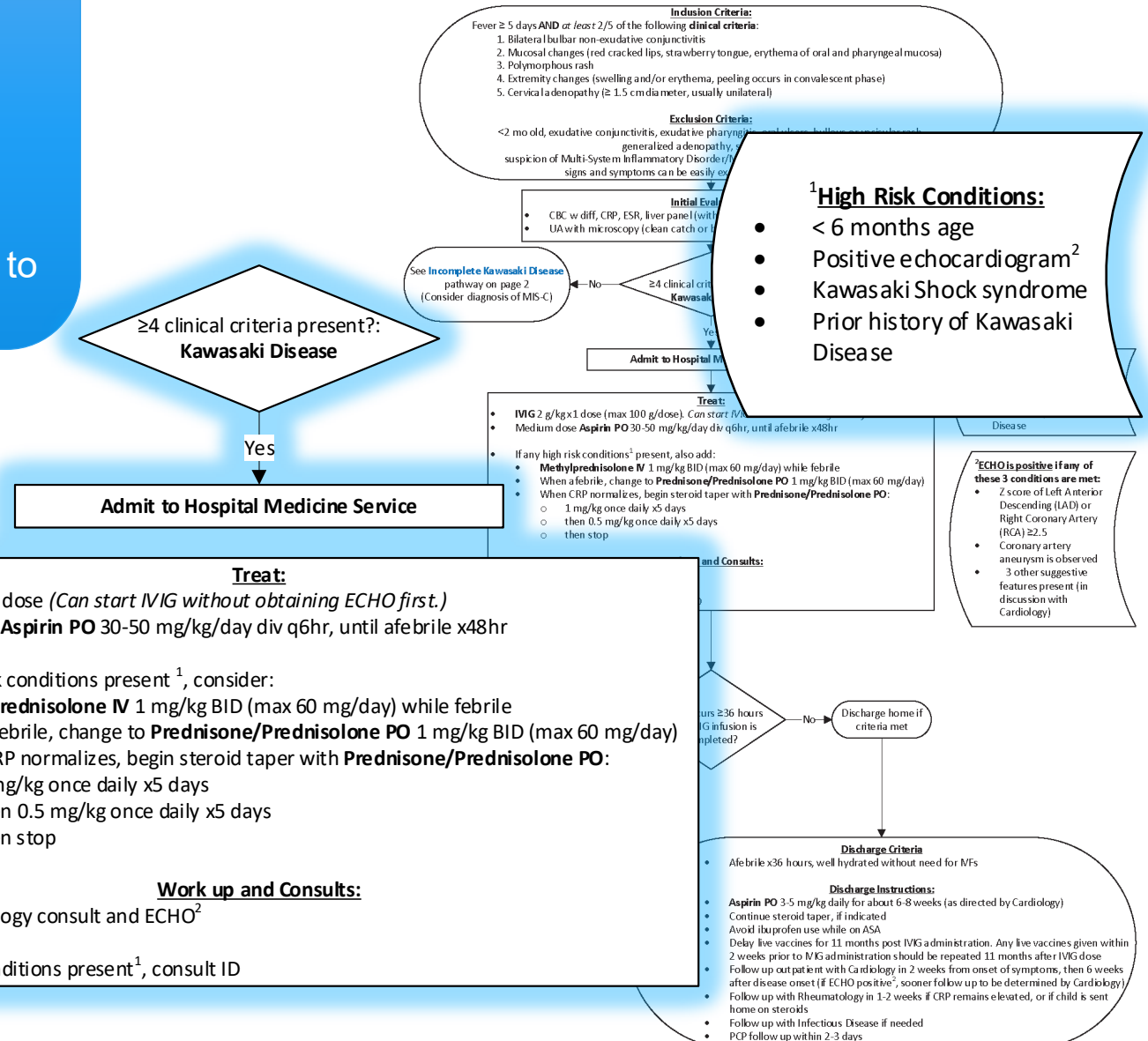
1. reduce the systemic inflammatory process
2. prevent coronary artery abnormalities
3. if coronary artery abnormalities are present, then to minimize the peak dimension and any clots

If a patient has any High risk characteristics, consult with Infectious Diseases and consider adding Methylprednisolone

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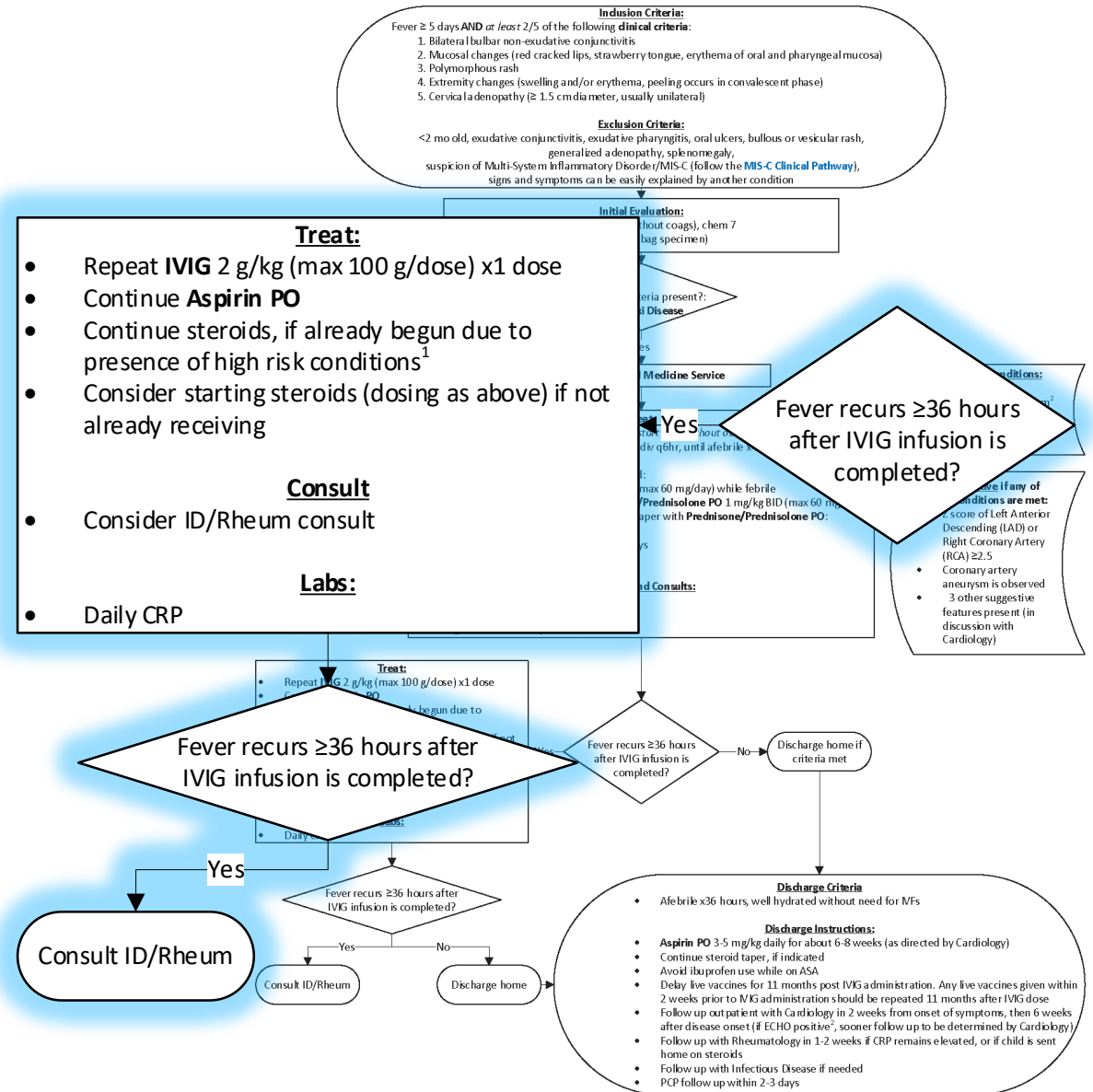
CLINICAL PATHWAY: Kawasaki Disease and Incomplete Kawasaki Disease

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Kawasaki Disease

IVIG treatment may be repeated for a second dose if the patient's fever recurs ≥ 36 hours after the IVIG infusion is completed.

Consider Infectious Disease and/or Rheumatology consults at any point, particularly if the patient's fever recurs ≥ 36 hours after the 2nd IVIG infusion is completed.



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Kawasaki Disease

Discharge can be considered once a patient is fever free for 36 hours and well hydrated.

Low dose Aspirin is continued on an outpatient basis as directed by Cardiology.

If patient is discharged on steroids, they should follow up with Rheumatology.

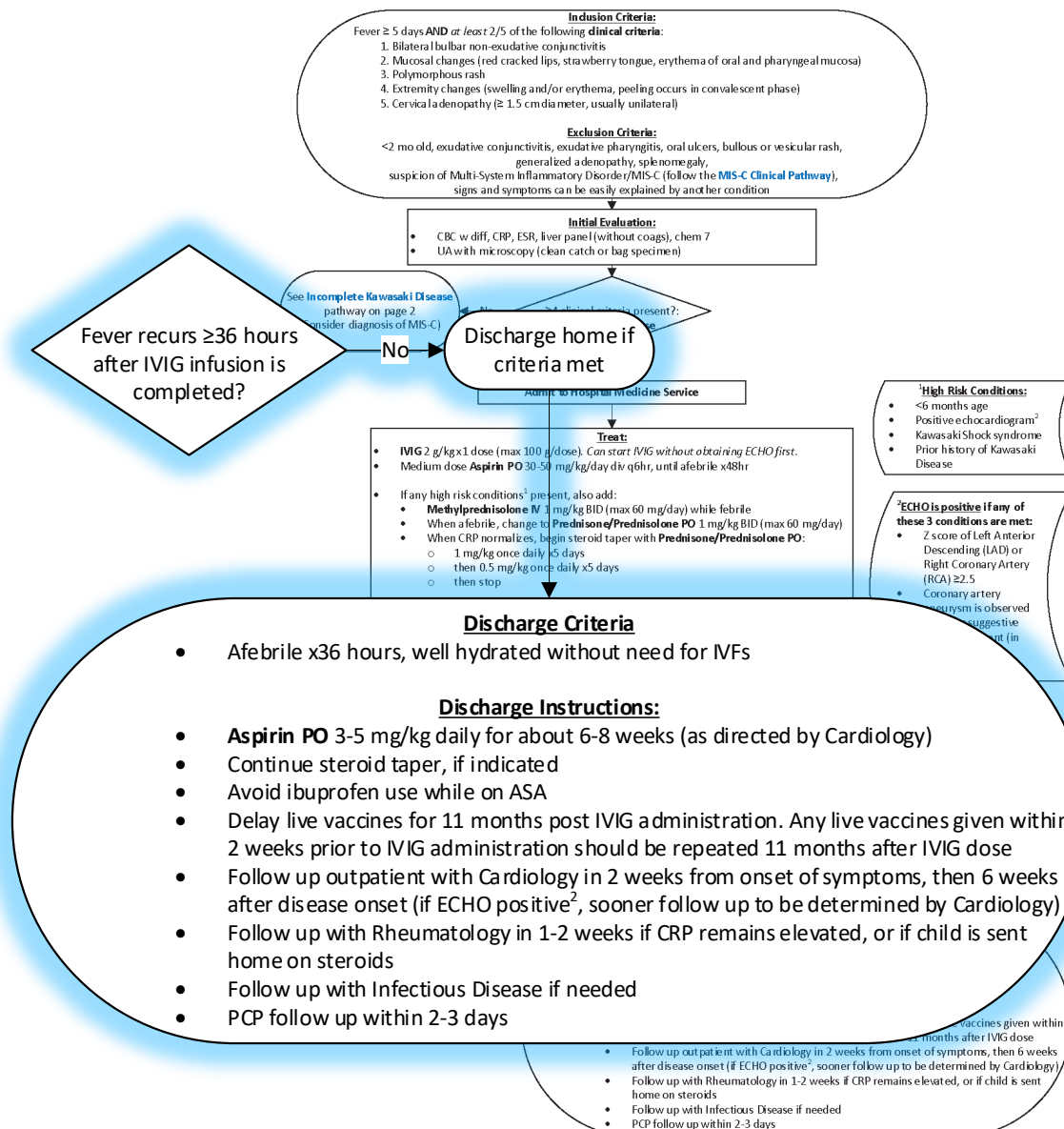
Parental education prior to discharge is imperative

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

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Kawasaki Disease and Incomplete Kawasaki Disease

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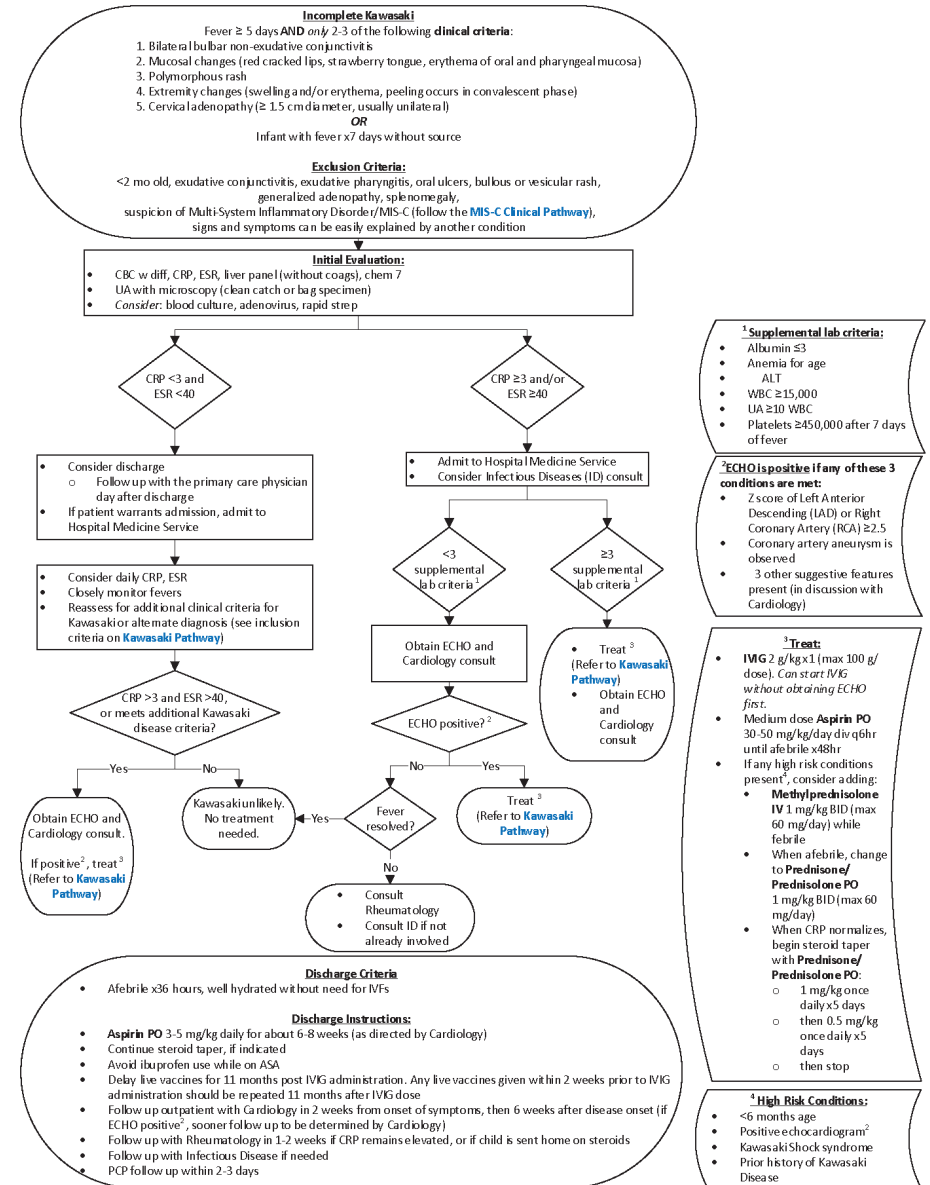
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CLINICAL PATHWAY: Kawasaki Disease and Incomplete Kawasaki Disease

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



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Incomplete Kawasaki Disease

If a patient meets criteria for incomplete Kawasaki Disease, the initial lab work done on presentation is used to guide further work up and possible treatment as necessary.

CLINICAL PATHWAY:

Kawasaki Disease and Incomplete Kawasaki Disease

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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

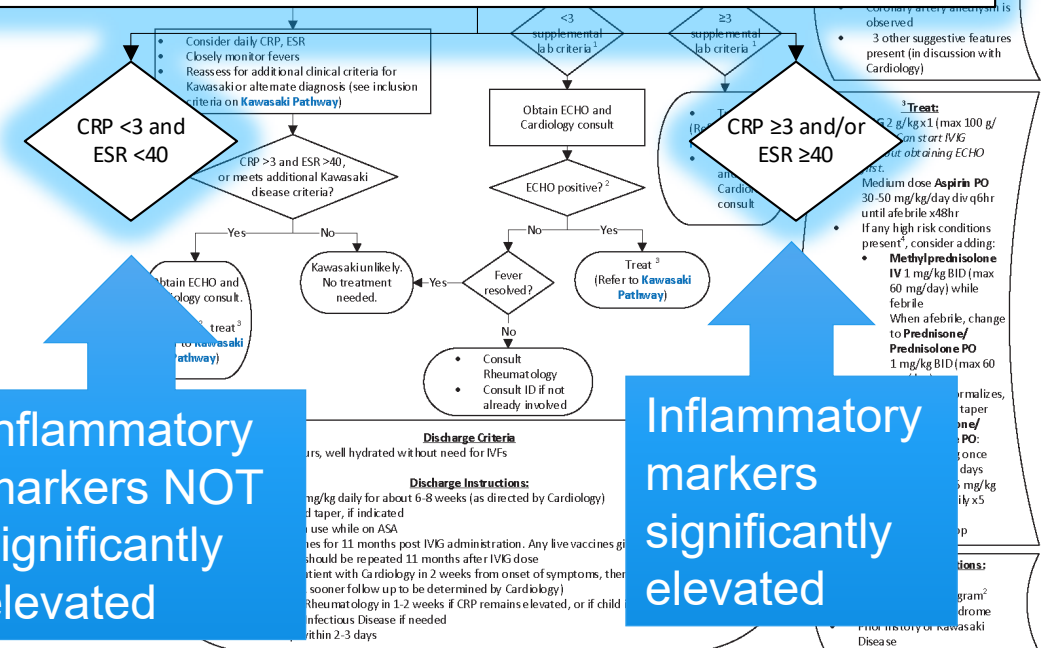
Infant with fever $\times 7$ days without source

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), chem 7
- UA with microscopy (clean catch or bag specimen)
- Consider: blood culture, adenovirus, rapid strep



Inflammatory
markers NOT
significantly
elevated

Inflammatory
markers
significantly
elevated

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Incomplete Kawasaki Disease

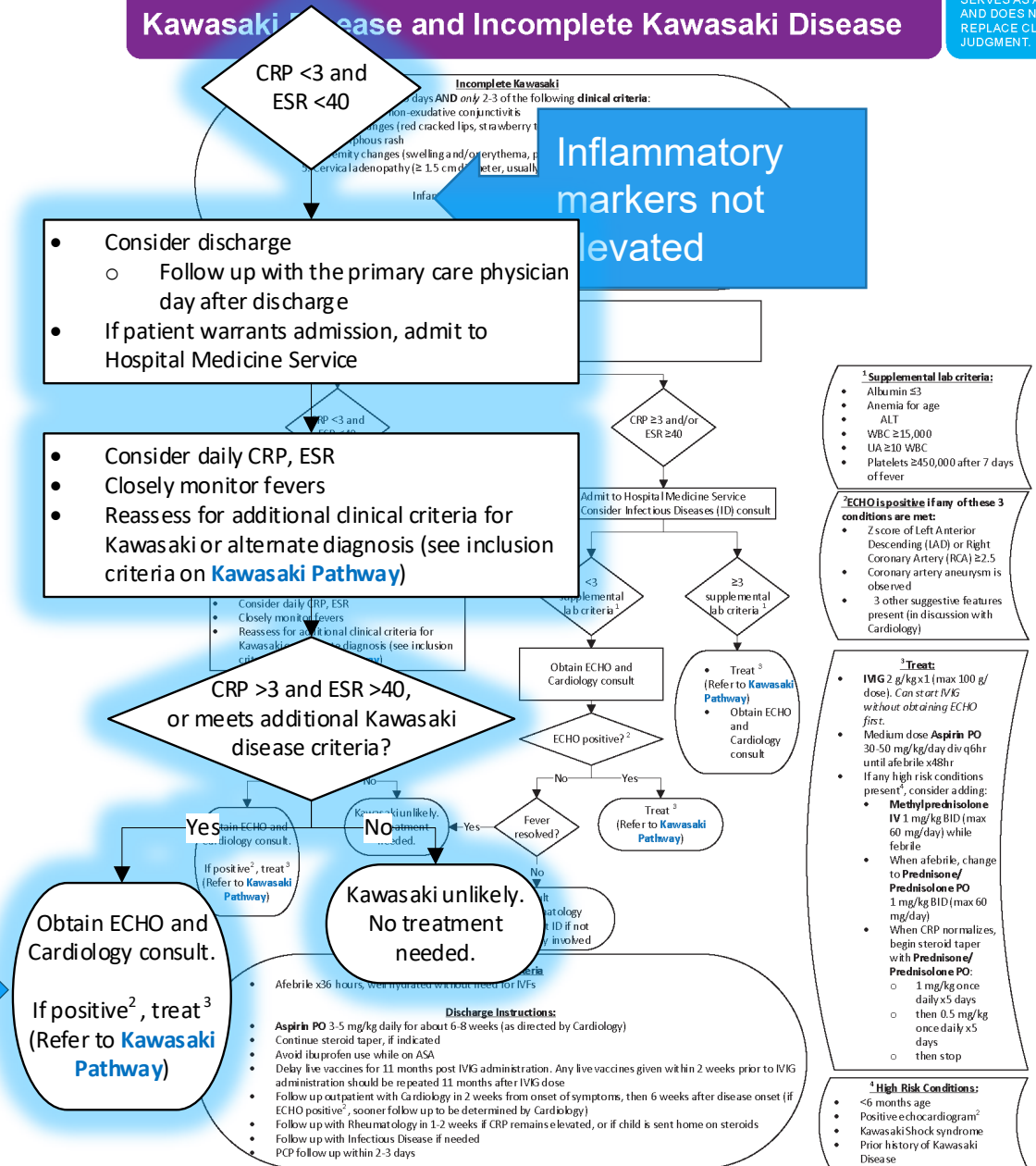
If inflammatory markers are NOT significantly elevated on presentation:

- 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.

If ECHO is positive – treatment is indicated.

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If inflammatory markers ARE significantly elevated at the time of initial evaluation:

- Patient should be admitted to PHM
- The patient's supplemental labs will determine the next steps.

THIS PATHWAY
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- Albumin ≤ 3
- Anemia for age
- \uparrow ALT
- WBC $\geq 15,000$
- UA ≥ 10 WBC
- Platelets $\geq 450,000$ after 7 days of fever

Elevated Inflammatory Markers

CRP ≥ 3 and/or
ESR ≥ 40

¹ **Supplemental lab criteria:**

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

- Z score of Left Anterior Descending (LAD) or Right Coronary Artery (RCA) ≥ 2.5
- Coronary artery aneurysm is

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

supplemental
lab criteria¹

Fewer than 3 supplemental lab criteria present

supplemental
lab criteria ¹

3 or more
supplemental
lab criteria
present

MD | HE

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If inflammatory markers ARE significantly elevated AND 3 or more supplemental laboratory criteria are met:

Elevated Inflammatory Markers

3 or more
supplemental
laboratory criteria

>> Treatment is indicated.

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

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

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

- Medium dose **Aspirin PO** 60 mg/kg/day divided b/w 4 hr while x48hr
- If high risk conditions present, consider adding:
 - **Methylprednisolone IV** 1 mg/kg BID (max 60 mg/day) while febrile
 - When afebrile, change to **Prednisone/ Prednisolone PO** 2 mg/kg BID (max 60 mg/day)
 - CRP normalizes, steroid taper **prednisone/ prednisolone PO:**
 - 1 mg/kg/day
 - q 5 days
 - then 0.5 mg/kg
 - then daily x5 days
 - then stop

- Positive echocardiogram²
- Kawasaki Shock syndrome
- Prior history of Kawasaki Disease

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Incomplete Kawasaki Disease

If inflammatory markers ARE significantly elevated but patient has less than 3 supplemental lab criteria present:

- Obtain an ECHO to determine next steps.

Elevated Inflammatory Markers

Less than 3 supplemental lab criteria

ECHO

CLINICAL PATHWAY: Kawasaki Disease and Incomplete Kawasaki Disease

THIS PATHWAY REQUIRES A GUIDE

¹ Supplemental lab criteria:

- Albumin ≤ 3
- Anemia for age
- \uparrow ALT
- WBC $\geq 15,000$
- UA ≥ 10 WBC
- Platelets $\geq 450,000$ 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)

Kawasaki unlikely. No treatment needed.

Treat ³ (Refer to Kawasaki Pathway)

- Consult Rheumatology
- Consult ID if not already involved

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 positive², 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

- present¹, 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 x 5 days
 - then 0.5 mg/kg once daily x 5 days
 - then stop

- ⁴ High Risk Conditions:
- <6 months age
 - Positive echocardiogram²
 - Kawasaki Shock syndrome
 - Prior history of Kawasaki Disease

Incomplete Kawasaki Disease

If inflammatory markers ARE significantly elevated but patient has less than 3 supplemental lab criteria present:

- If ECHO negative, evaluate fever curve.

Elevated Inflammatory Markers

Less than 3 supplemental lab criteria

If ECHO is negative: Proceed with work up based on clinical picture.

CLINICAL PATHWAY: Kawasaki Disease and Incomplete Kawasaki Disease

THIS PATHWAY REQUIRES A GUIDE

¹ Supplemental lab criteria:

- Albumin ≤ 3
- Anemia for age
- \uparrow ALT
- WBC $\geq 15,000$
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- Platelets $\geq 450,000$ 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)

Kawasaki unlikely. No treatment needed.

Treat ³ (Refer to Kawasaki Pathway)

- Consult Rheumatology
- Consult ID if not already involved

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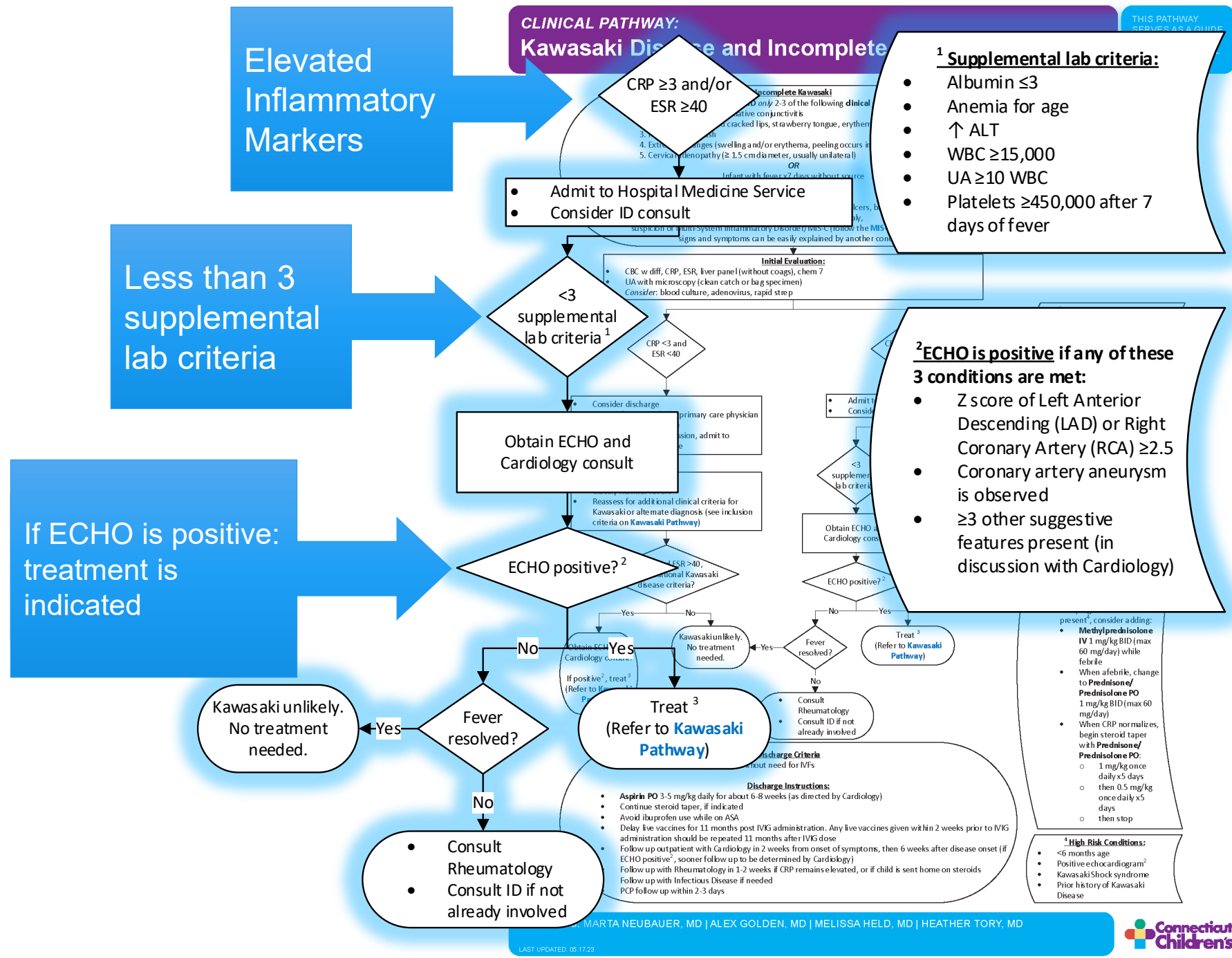
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If inflammatory markers ARE significantly elevated but patient has less than 3 supplemental lab criteria present:

- Less than 3 supplemental lab criteria

If ECHO is positive:
treatment is
indicated

Elevated Inflammatory Markers

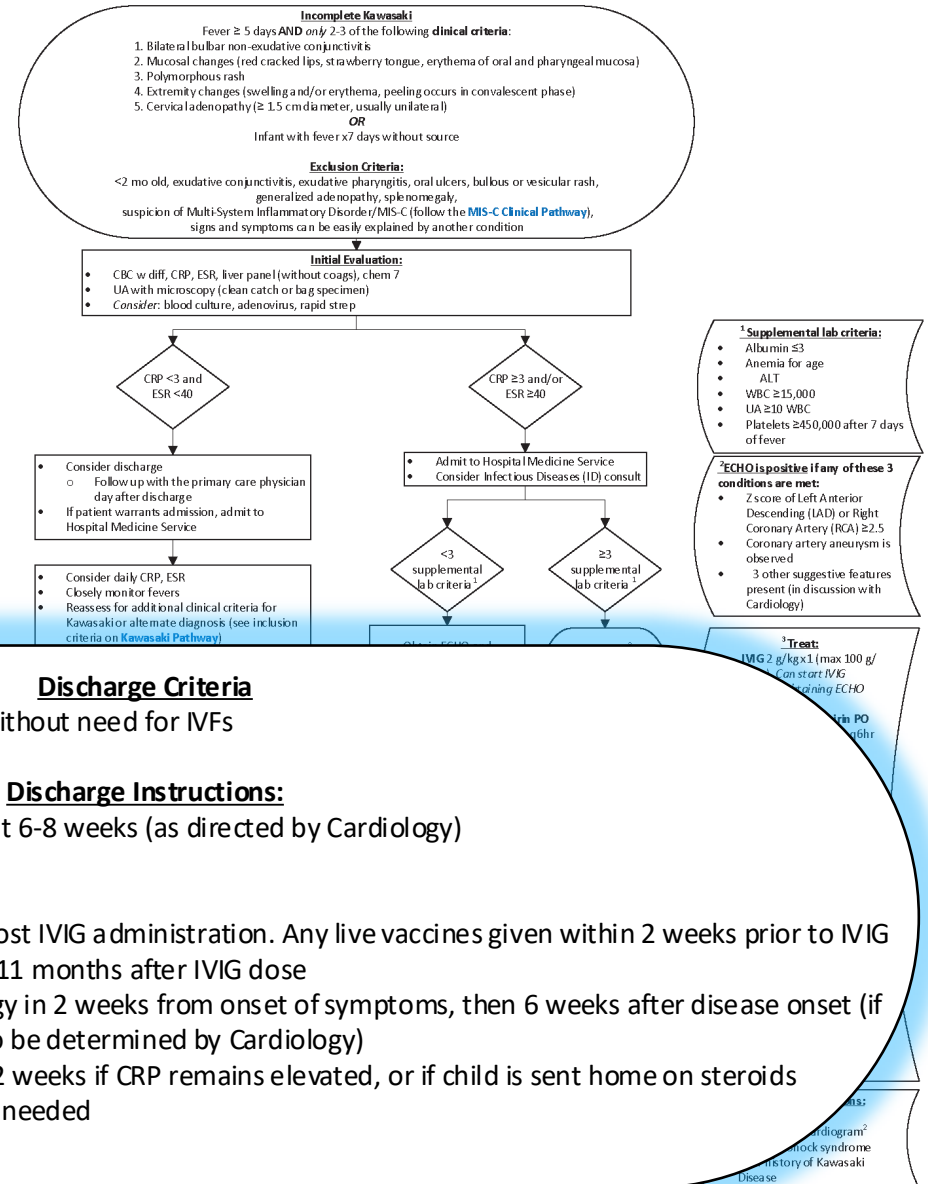


CLINICAL PATHWAY: Kawasaki Disease and Incomplete Kawasaki Disease

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Incomplete Kawasaki Disease

Discharge criteria and instructions are the same as for Kawasaki Disease.



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Review of Key Points



- **Kawasaki Disease** is defined as fever for ≥ 5 days **and** *at least* 2 out of 5 clinical features.
- **Incomplete Kawasaki Disease** is when a patient presents with 5 days of fever, but may only have 2 or 3 clinical features (or when an infant has 7 days of fever without a source)
 - Inflammatory labs, supplemental labs, and ECHO may be used to help guide management.
- 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



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Thank You!



About Connecticut Children's Clinical Pathways Program

The Clinical Pathways Program at Connecticut Children's aims to improve the quality of care our patients receive, across both ambulatory and acute care settings. We have implemented a standardized process for clinical pathway development and maintenance to ensure meaningful improvements to patient care as well as systematic continual improvement. Development of a clinical pathway includes a multidisciplinary team, which may include doctors, advanced practitioners, nurses, pharmacists, other specialists, and even patients/families. Each clinical pathway has a flow algorithm, an educational module for end-user education, associated order set(s) in the electronic medical record, and quality metrics that are evaluated regularly to measure the pathway's effectiveness. Additionally, clinical pathways are reviewed annually and updated to ensure alignment with the most up to date evidence. These pathways serve as a guide for providers and do not replace clinical judgment.