Kawasaki Disease: Diagnosis and Treatment

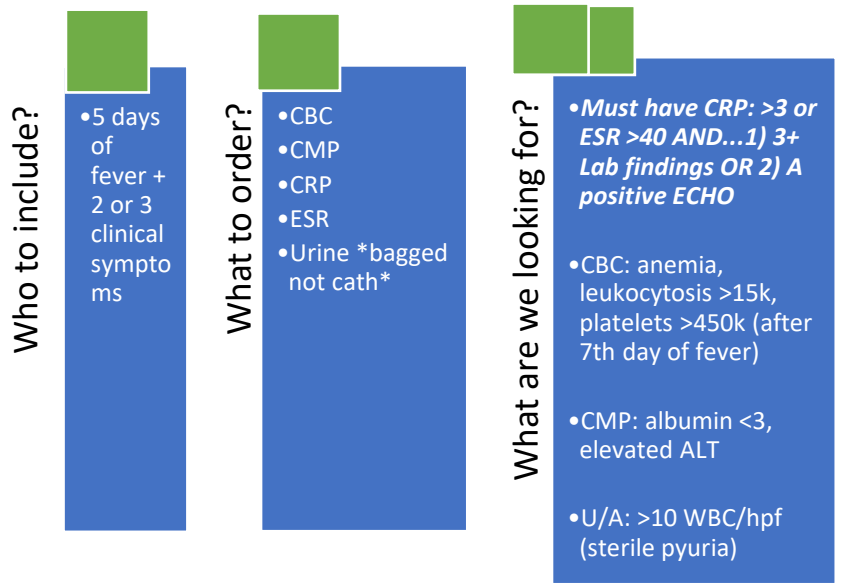
Background:
Kawasaki disease is an acute vasculitis of childhood that affects medium sized blood vessels. It's important to recognize because when left untreated, 25% of children will develop coronary artery aneurysms. With treatment, this number drops to 4%. Most common cause of acquired heart disease in pediatric populations in developed countries.

Pathophysiology:
Incidence in North America = 25 in 100,000 kids <5 years old, but incidence is higher in Japanese and Taiwanese populations. This leads us to believe that Kawasaki disease is a result of both a genetic predisposition AND an environmental trigger. The environmental trigger is unknown, but since Kawasaki is more common in winter and spring – aka virus season - in North America, the theory is that it is triggered by a viral illness. Once triggered by the environment, the Immune system goes bananas and starts attacking the medium sized vessels, causing a vasculitis.

Clinical Signs: Remember we are looking for systemic signs of inflammation in all the medium-sized arteries.

- Classic KD: Remember CONER THE KAWASAKI KID: **"F CONER"**
  - **Need: 5+ days Fever + 4 "CONER" criteria**
  - **F**ever
    - High fevers, usually 39 C and above, daily for 5+ days
    - Untreated, fevers persist for 1-3 weeks
  - **C**onjunctivitis
    - Nonexudative conjunctival injection
    - Bilateral
    - Limbic sparing: no redness in the limbic area, just around the iris. This is because there are fewer blood vessels there, and remember, this is a vasculitis
  - **O**ral Changes
    - Dry, cracked, peeling, erythematous lips
    - Strawberry tongue with prominent fungiform papillae
    - Diffuse erythema of the mucosa *no exudates*
  - **N**ode
    - *least common of the principal features*
    - Unilateral swelling, >1.5cm in diameter, and in anterior cervical triangle
    - Multiple nodes will likely be enlarged, but only one has to be >1.5cm diameter
  - **E**xtremity Changes
    - Erythema of palms and soles
    - Later, desquamation (within 2-3 weeks after onset of fever)
  - **R**ash
    - Erythematous rash occurs within 5 days of fever onset
    - Usually looks like a viral exanthem – diffuse, maculopapular, blanching rash on trunk and extremities, sometimes worse in the groin area – but can have a variety of appearances
- Atypical KD:
  - Also called "incomplete" Kawasaki – because not all classic criteria are met. This diagnosis is most common in infants, because they are at substantial risk for developing coronary artery anomalies but may only present with fever and otherwise subtle clinical findings. Additionally, these infants, particularly <6mo of age, are at high risk for development of coronary artery aneurysms, so definitely something to keep on your differential when assessing an infant with prolonged fever (>7 days) (or prolonged fever and pyuria).

- **Working up for *Atypical* Kawasaki Disease:**



- Differential:
  - Adenovirus (high fevers, rash, erythematous oropharynx, conjunctivitis)
  - Acute Unilateral Lymphadenitis (fever, unilateral node)
  - Measles (rash, fever)
  - MIS-C

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