KAWASAKI DISEASE

- Kawasaki disease, also known as Kawasaki syndrome,
- lymph node syndrome,
- mucocutaneous lymph node syndrome

Definition

- Idiopathic multisystem disease characterized by vasculitis of small & medium blood vessels, including coronary arteries.

- " A self-limited multisystem vasculitis of unknown etiology that predominantly affects children younger than 5 years. Jane Burns, MD*

- It is now the most common cause of acquired heart disease in children in the United States and Japan."

Epidemiology

- Median age of affected children = 2.3 years
- 80% of cases in children < 4 yrs, 5% of cases in children > 10 yrs
- Males:females = 1.7:1.(Males are more)
- Recurs in 3%
- Positive family history in 1% but 13% risk of occurrence in twins
- Overall U.S. in-hospital mortality ≈ 0.17%

Epidemiology

- Annual incidence of 4-15/100,000 children under 5 years of age
- The highest incidence occurs in Japan, 218.6 per 100,000 children <5 years of age
- more than one in 150 children in Japan
- Seasonal variation
  - More cases in winter and spring but occurs throughout the year

Etiology

- UNKNOWN
- Infectious agent most likely
  - Age-restricted susceptible population
  - Seasonal variation
  - Well-defined epidemics
  - Acute self-limited illness similar to known infections

- No causative agent identified
  - Bacterial, retroviral, superantigenic bacterial toxin
  - Immunologic response triggered by one of several microbial agents

Pathophysiology

Due to unknown etiology

Inflammation of small and medium sized blood vessels

coronary artery

more susceptible to damage

Aneurysms in 7th day of illness

Become largest diameter

Myointimal proliferation

pancarditis

Signs and symptoms of carditis
Tachycardia, a hyperdynamic precordium, a gallop rhythm
S3-- "lub-dub-ta"
S4 "ta-lub-dub"
or a flow murmur may be present;

Lab: CRP ESR , ECG , ECHO changes

Pathophysiology

Myointimal proliferation(affected vessels may try to heal to normal, but only size, but become thicker)

scarring, calcification,

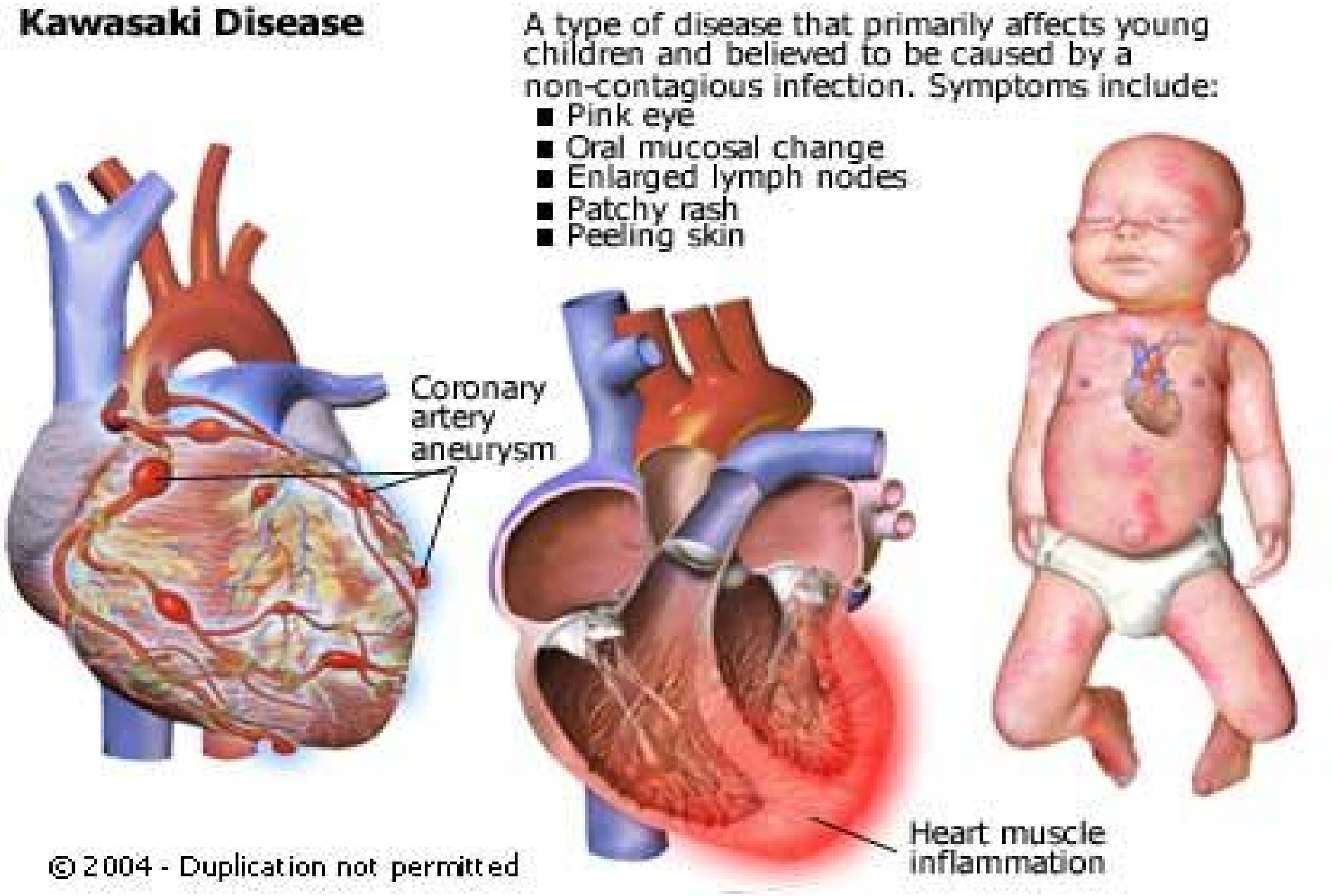
coronary thrombosis

Stenosis

MI

death

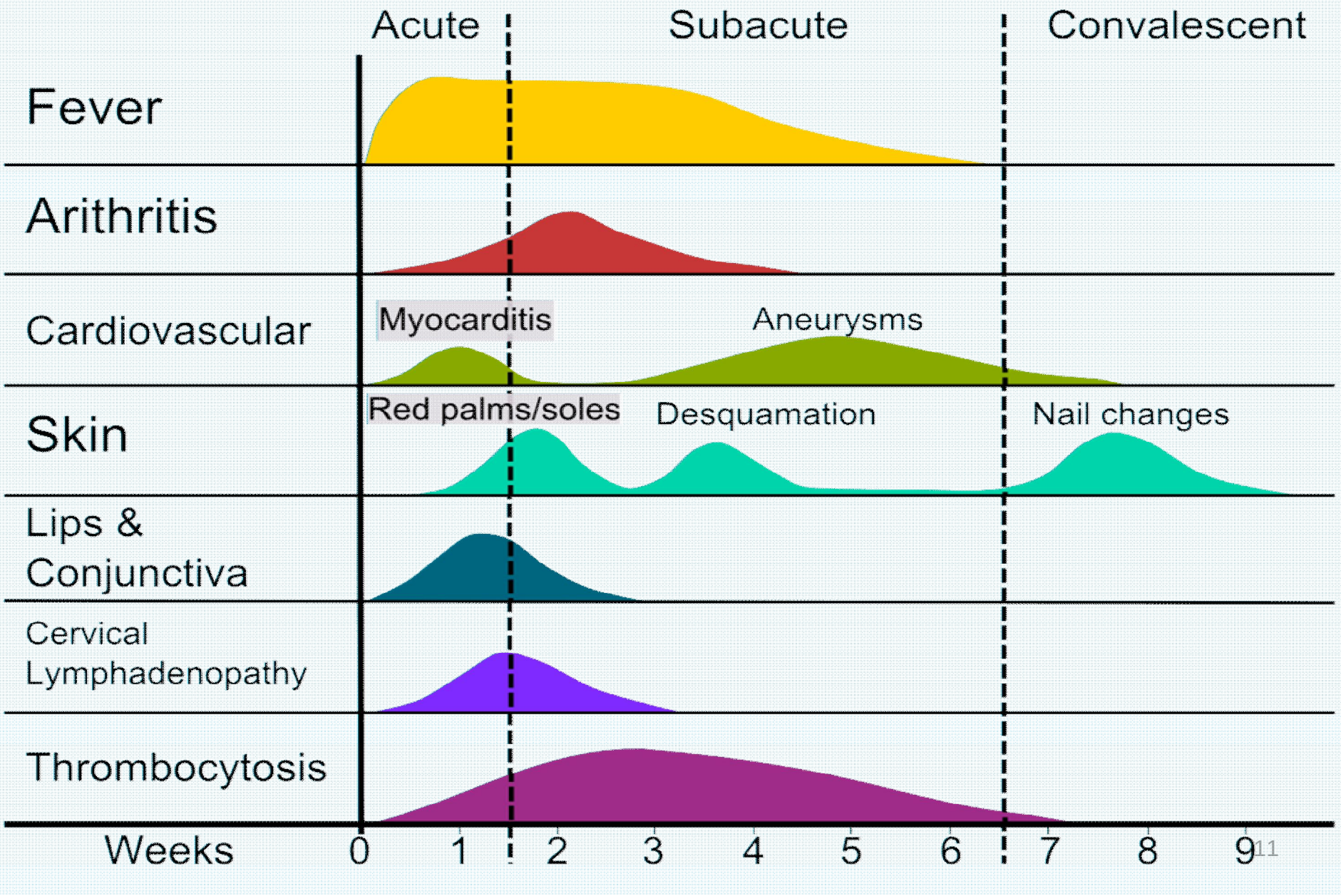
Signs and symptom



Signs and symptom in 3 Phases of Disease

- Acute
  - Febrile, irritable, toxic appearing
  - Oral changes, rash but never vesicular, edema/erythema of feet
- Subacute
  - Desquamation, may have persistent arthritis or arthralgias
  - Gradual improvement even without treatment
- Convalescent

Clinical manifestations of Kawasaki Disease



(A) Bilateral, nonexudative conjunctivitis with perilimbal sparing "conjunctival injection" (B) Strawberry tongue and bright red, swollen lips with vertical cracking and bleeding (C) Erythematous rash involving perineum (D) Redness of the palms, which is often accompanied by painful, brawny swelling of the back of the hands (E) Redness of the soles, and swelling of the back of the feet (F) Peeling of the skin of the fingers (G) Redness and induration at the site of a previous vaccination with Bacillus Calmette-Guérin (H) Perianal redness and peeling[5]

Signs and symptoms

- First symptom:- Intermittent high grade fever lasting more than 5 days. (37.8°C (100.0°F) or higher)
- Unresponsive to antibiotic and antipyretic therapy.
- Arthrits: Joint pain and swelling, frequently symmetrical involvement
- Cardiovascular:

Suggestive of myocarditis (50%)

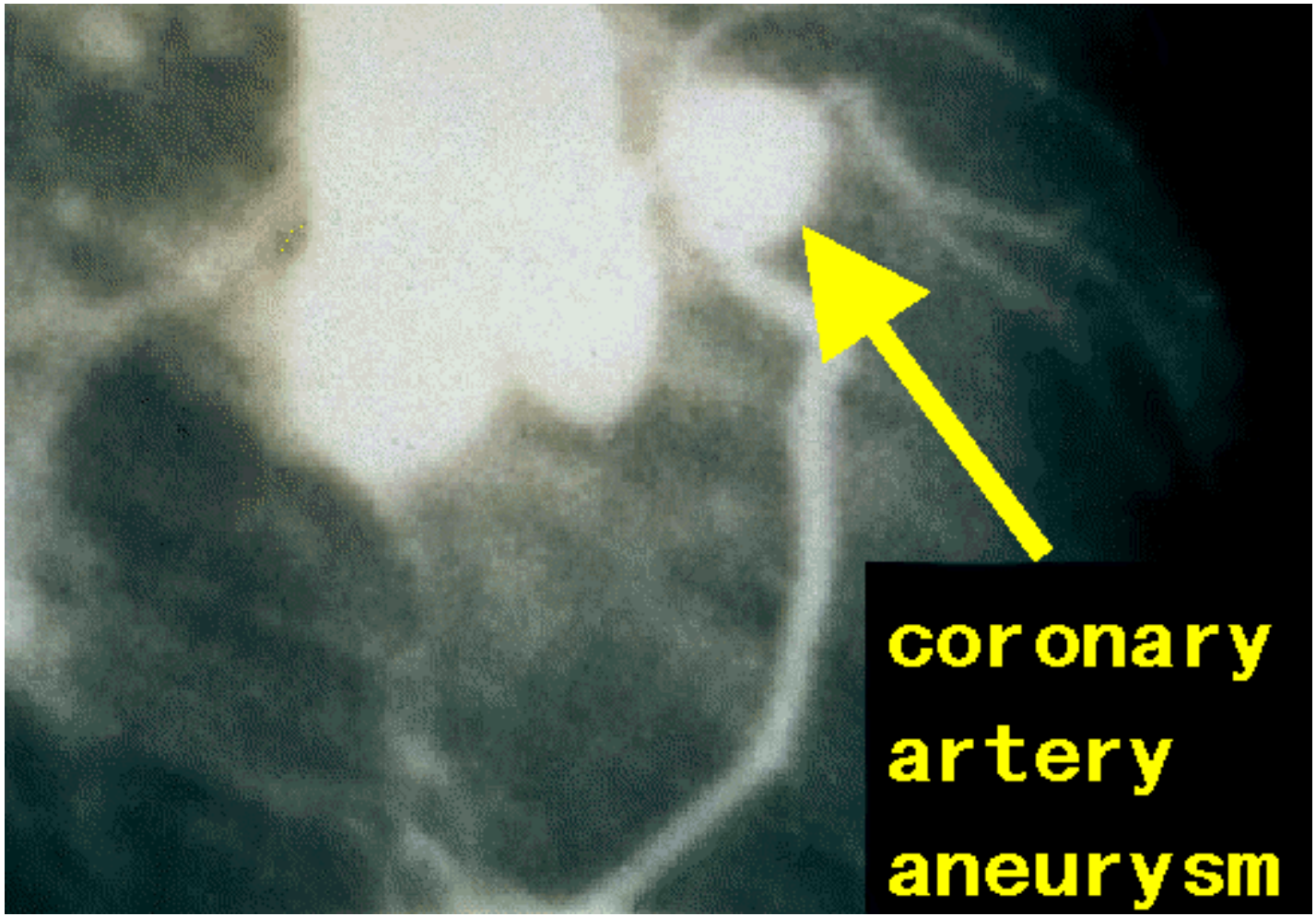
– Tachycardia, murmur, gallop rhythms

Suggestive of pericarditis (25%)

– Distant heart tones, pericardial friction rub, tamponade

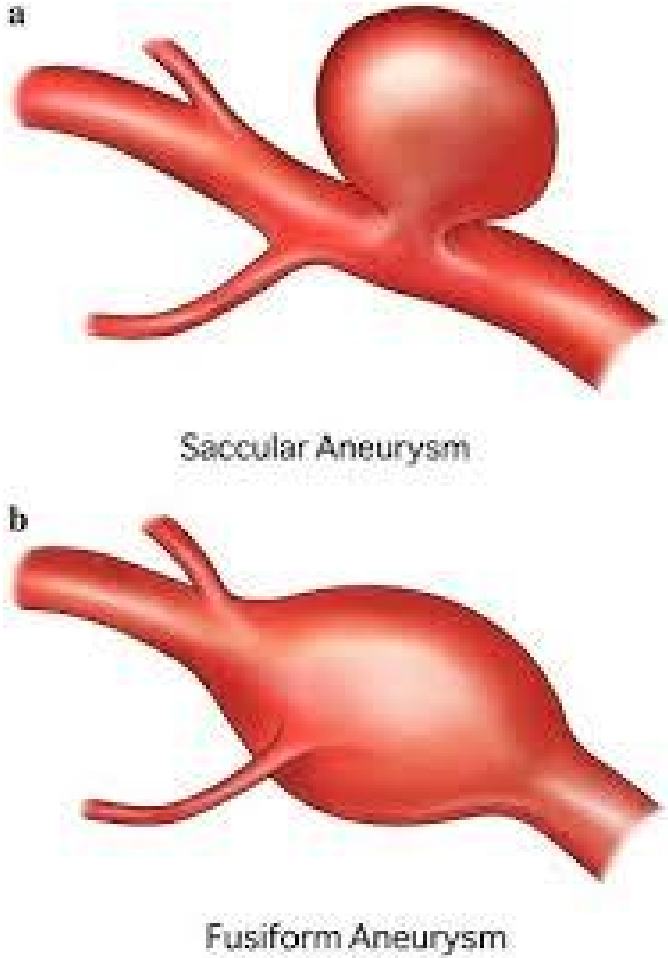
- Aneurism

- Normally, the walls of blood vessels are smooth and even. In Kawasaki disease, the muscular walls of the coronary arteries may become weakened.
- The pressure of the blood flowing through the arteries may cause these weak spots to balloon-out, just like a weak spot in a tire or inner tube.


coronary
artery
aneurysm

Coronary Aneurysms

- Size
  - Small = <5 mm diameter
  - Medium = 5-8 mm
  - Giant = ≥ 8 mm
    - Highest risk for sequelae
- Shape
  - Saccular
  - Fusiform



Coronary Aneurysms

- Patients most likely to develop aneurysms
  - Younger than 6 months, older than 8 years
  - Males
  - Fevers persist for greater than 14 days
  - Persistently elevated ESR
  - Thrombocytosis
  - Pts who manifest s/s of cardiac involvement

Coronary Aneurysms

- 50 % regress to normal
- 25 % become smaller
- 25 % do not regress
- 7-20 % develop stenosis or myocardial infarction cause death
- MI is principal cause of death in KD
  - 32% mortality
  - Most often in the first year
  - Majority while at rest/sleeping
  - About 1/3 asymptomatic

Signs and symptoms

- Skin: Erythema of palms & soles
  - macular-papular erythematous rash, and is characteristically located on the trunk, spread to face, extremities and perineum.
  - Often not pruritic, vesicular
  - 11% children, skin-peeling for many years
  - 99% of cases,
  - in perineum (60%)

- painful edema of hands/feet
  - Usually start 3-5 days after onset of fever
  - Subliungual desquamation in subacute

Signs and symptoms

- Nail changes :-
  One to two months after the onset of fever, deep transverse grooves across the nails may develop (Beau's lines)

Signs and symptoms

- Lips , moth and tongue:

In the first week, they may also have erythema and vertical cracking of lips, a strawberry tonque

Signs and symptoms

- Conjunctiva:- Red eye but no discharge

Signs and symptoms

lymphadenopathy,

- early stage lymphadenopathy, can be very transient.
- It may or may not be bilateral but is not generalized throughout the body.
- must be 1.5 cm in diameter or larger.
- About 90% of patients will have fever, conjunctival erythema, and oral changes and 70% will have lymphadenopathy.
- Affected lymph nodes are painless or minimally painful, nonsuppurative

Other system S&S

- Respiratory
  - Rhinorrhea, cough, pulmonary infiltrate
- GI
  - Diarrhea, vomiting, abdominal pain, hydrops of the gallbladder, jaundice
- Neurologic
  - Irritability, aseptic meningitis, facial palsy, hearing loss
- Musculoskeletal
  - Myositis, arthralgia, arthritis

Diagnostic evalvation

- History of fever not responding to antibiotic and antipyretics.
- Physical examination :- s/s,
- Suggestive of myocarditis (50%)

– Tachycardia, murmur, gallop rhythms

Suggestive of pericarditis

– Distant heart tones, pericardial friction rub, tamponade

- Blood:- Leukocytosis, Mild anemia, Thrombocytopenia/ Thrombocytosis, Elevated ESR, Elevated CRP, Hypoalbuminemia
- EKG changes :-Arrhythmias, Abnormal Q waves, Prolonged PR and/or QT intervals, ST-T–wave changes.

- ChestX ray–cardiomegaly,

Diagnostic evalvation

- ECHO:-
- Aneurism
- Myocarditis with dysfunction
- Pericarditis with an effusion
- Valvar insufficiency
- Coronary arterial changes

- Ultrasound or computerized tomography may show hydrops (enlargement) of the gallbladder.
- .

Criteria for Diagnosis of Kawasaki Disease
Fever of ≥5 days' duration associated with at least four† of these five changes
Bilateral nonsuppurative conjunctivitis
One or more changes of the mucous membranes of the upper respiratory tract, including throat redness, dry cracked lips, red lips, and "strawberry" tongue
One or more changes of the arms and legs, including redness, swelling, skin peeling around the nails, and generalized peeling
Polymorphous rash, primarily truncal
Large lymph nodes in the neck (>1.5 cm in size)
Disease cannot be explained by some other known disease process
†A diagnosis of Kawasaki disease can be made if fever and only three changes are present if coronary artery disease is documented by two-dimensional echocardiography or coronary angiography.

Diagnostic evalvation

- Classically, five days of fever plus four of five diagnostic criteria must be met establish the diagnosis.

The criteria are:

1. erythema of the lips or oral cavity or cracking of the lips;
2. rash on the trunk;
3. swelling or erythema of the hands or feet;
4. red eyes (conjunctival injection);
5. swollen lymph node in the neck of at least 15 mm.

Table 3. Differential diagnosis of Kawasaki disease[5]

Indications	Kawasaki disease	Stevens-Johnson syndrome	Streptococcal scarlet fever	Toxic shock syndrome	Systemic juvenile rheumatoid arthritis
Age (years)	Usually <5	Any age	Usually 2–8	Usually >10	2–5
Fever	Persistent	Prolonged	Variable, usually <10 d	Usually <10 d	Prolonged
Eyes	Nonexudative conjunctivitis, limbal sparing	Exudative conjunctivitis, keratitis	Normal	Conjunctivitis	Normal
Oral mucosa	Diffuse erythema, strawberry tongue	Erythema, ulceration, pseudomembrane formation	Pharyngitis, strawberry tongue	Erythematous	Normal
Extremities	Erythema of palms and soles, indurative edema, periungual desquamation (tends to be sheetlike)	Normal	Flaky desquamation	Swelling of hands and feet	Arthritis
Rash	Erythematous, polymorphous; targetoid or purpuric	Target lesions	Sandpaper rash, Pastia sign, circumoral pallor	Erythroderma	Transient, salmon pink
Cervical lymphadenopathy	At least one lymph node ≥1.5 cm	Normal	Painful swelling	Normal	Diffuse adenopathy
Characteristic lab results	Systemic inflammation, anemia, transaminitis, thrombocytosis after day 7	Associated herpes virus infection	Positive throat culture	Thrombocytopenia	Systemic inflammation, anemia
Other	Arthritis	Arthralgia, associated herpes virus infection (30%–75%)	Throat culture positive for group A streptococcus	Mental status changes, coagulopathy, shock	Pericarditis

Adapted from Yanagihara R, Todd JK. Acute febrile mucocutaneous lymph node syndrome. Am J Dis Child 1980 Jun;134(6):603–14, with permission of the American Medical Association.

Treatment for a Child with Kawasaki Disease

- There is no single medicine, which can treat or cure Kawasaki disease.
- Aspirin.
- Gamma Globulin

Treatment

- Aspirin
  - High dose (80-100 mg/kg/day QID) / until afebrile x 48 hrs &/or decrease in acute phase reactants
  - Decrease to low dose (3-5 mg/kg/day) for 6-8 weeks or until platelet levels normalize

Warfarin Sodium for those aneurism >8 mm.

Treatment

- IVIG : intravenous immune globulin
- 2g/kg as one-time dose over 10-12 hour IV
  - Mechanism of action is unclear
  - Significant reduction within first 10 days in CHF, aneurysm and Normal ESR
  - Efficacy unclear after day 10 of illness

Treatment

- IVIG
  - 70-90% defervesce & show symptom resolution within 2-3 days of treatment
  - Retreat those with failure of response to 1st dose or recurrent symptoms → Up to 2/3 respond to a second course

Treatment

- Aggressive support with diuretics & cardiac drugs for some patients with myocarditis
- Antibiotics while excluding bacterial infection

Patient Follow-Up Categories

- Five categories based on coronary arteries findings
  - No coronary changes at any stage of illness
  - Transient coronary artery ectasia (meaning "dilation" or "distention of a tubular structure"), resolved within 6-8 wks
  - Single Small/medium coronary aneurysm
  - One or more large or giant aneurysms or multiple smaller/complex aneurysms in same coronary artery, without obstruction
  - Coronary artery obstruction

Prognosis of Kawasaki Disease

- 95% recover completely.
- If no damage in the coronary arteries is seen on the echo tests, then complete recovery is most likely.
- This means that there is little chance of future problems.

Complications

- Cardiac complications are the most important aspect of kawasaki disease.
- Acute rheumatic fever .
- Coronary artery aneurysms occur as a sequela of the vasculitis
- Aneurysms
- Myocardial infarction secondary to thrombosis of a coronary artery aneurysm or to ruptureof a large coronary artery aneurysm. Death is most common two to 12 weeks

Nursing Diagnosis

- Alerted cardiac rhythm rhythm related to pancarditis
- Fear of death related to pancarditis
- Impaired mobiliy related to arthritis
- Altered body comfort related to disease process
- Altered body image
- Altered body image related changes in oral mucosal, tongue, rash, peeling of skin

Nursing Management

Monitoring

- Monitor pain level and child's response to analgesics.
- Institute continual cardiac monitoring and assessment for complications; report arrhythmias.
  - Take vital signs as directed by condition; report abnormalities.
  - Assess for signs of myocarditis (tachycardia, gallop rhythm, chest pain).
  - Monitor for heart failure (dyspnea, nasal flaring, grunting, retractions, cyanosis, orthopnea, crackles, moist respirations, distended jugular veins, edema).

Nursing Management

Monitoring

- Closely monitor intake and output, and administer oral and I.V fluids as ordered.
- Monitor hydration staus by checking skin turgor, weight, urinary output, specific gravity, and presence of tears.
- Observe mouth and skin frequently for signs of infection

Nursing Management

Supportive care

- Allow the child periods of uninterrupted rest. Offer pain medication routinely rather than as needed during stage I. Avoid NSAIDS if the child is in aspirin therapy.
- Perform comfort measures related to the eyes.
  - Conjunctivities can cause photosensitivity, so darken the room, offer sunglasses.
  - Apply cool compress.
  - Discourage rubbing the eyes.
  - Instill artificial tears to soothe conjunctiva.

Nursing Management

– *Supportive care*

- Monitor temperature every 4 hours. Provide sponge bath if temperature above normal.
- Perform passive range of motion exercises every 4 hours while the child is awake because movement may be restricted.
- Provide quiet and peaceful environment with diversional activities.
- Provide care measures for oral mucous membrane.
  - Offer cool liquids like ice chips and ice pops.
  - Use soft toothbrush only.
  - Apply petroleum jelly to dried, cracked lips.

Nursing Management

- Provide skin measures to improve skin integrity.
  - Avoid use of soap because it tends to dry skin and make it more likely to breakdown.
  - Elevate edematous extremities.
  - Use smooth sheets.
  - Apply emollients to skin as ordered.
  - Protect peeling of skin, observe for signs of infection.
- Offer clear liquids every hour when the child is awake.
- Encourage the child to eat meals and snack with adequate protein.

Nursing Management

- Infuse I.V fluids through a volume control device if dehydration is present
- Explain all procedures to the child and family.
- Encourage the parents and child to verbalize their concerns, fears, and questions.
- Practice relaxation techniques with child, such as relaxation breathing, guided imagery, and distraction.
- Prepare the child for cardiac surgery or thrombolytic therapy if complications develop.
- Keep the family informed about progress and reinforce stages and prognosis.


ThankYou!

Atypical or Incomplete Kawasaki Disease

- Present with < 4 of 5 diagnostic criteria
- Compatible laboratory findings
- Still develop coronary artery aneurysms
- No other explanation for the illness
- More common in children < 1 year of age

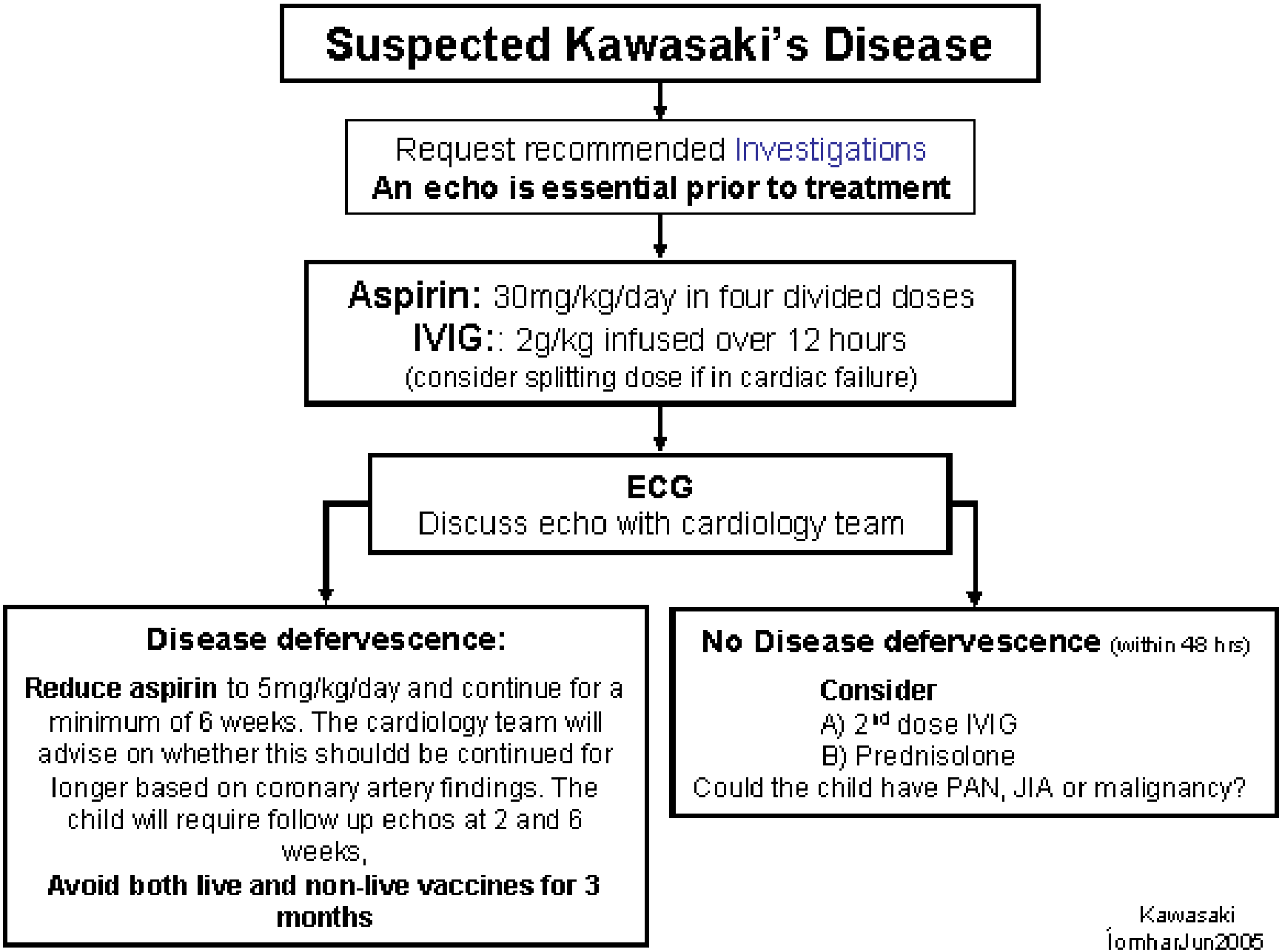
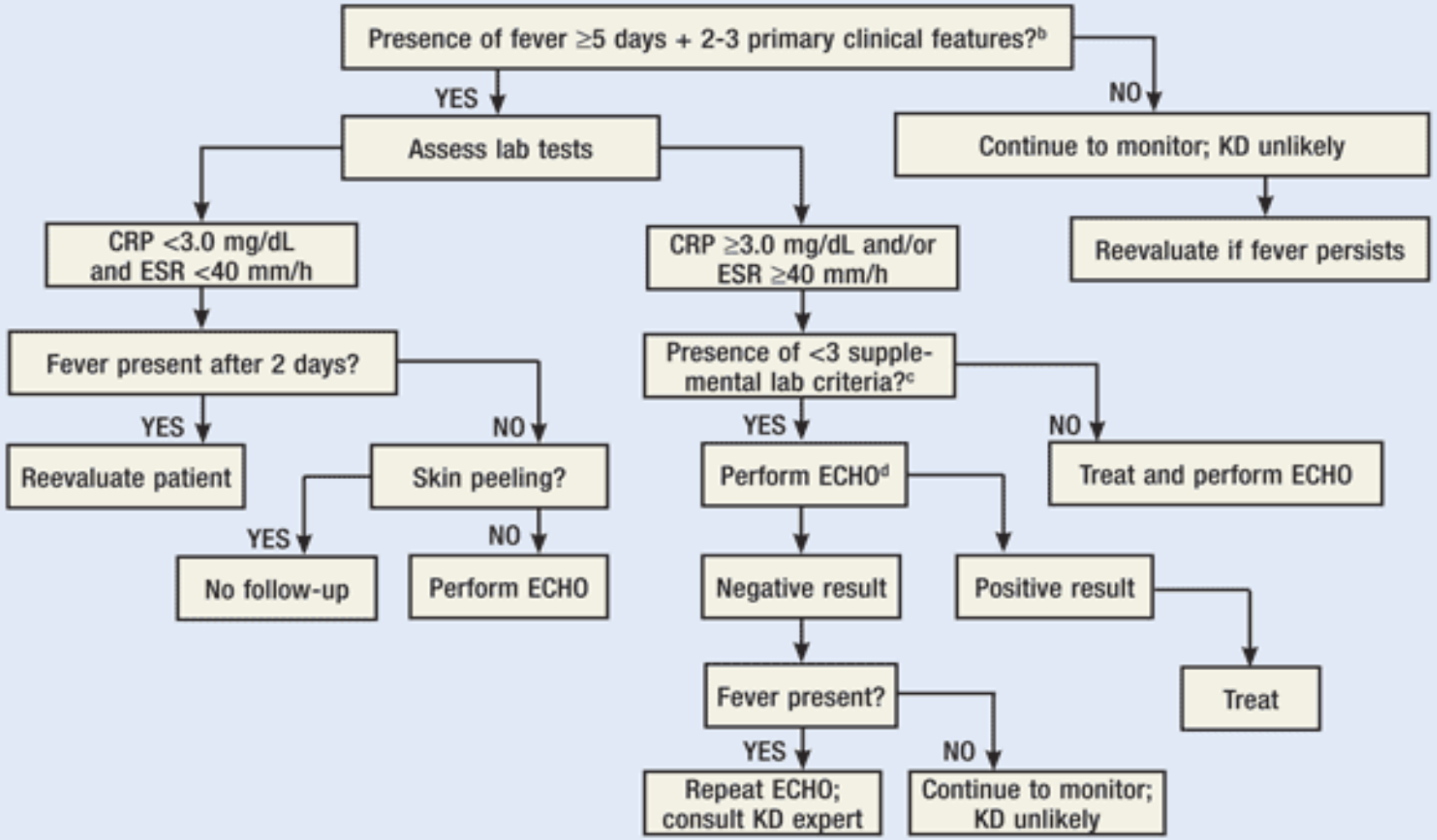

Suspected Kawasaki's Disease
Request recommended Investigations
An echo is essential prior to treatment
Aspirin: 30mg/kg/day in four divided doses
IVIG:: 2g/kg infused over 12 hours
(consider splitting dose if in cardiac failure)
ECG
Discuss echo with cardiology team
Disease defervescence:
Reduce aspirin to 5mg/kg/day and continue for a minimum of 6 weeks. The cardiology team will advise on whether this shouldd be continued for longer based on coronary artery findings. The child will require follow up echos at 2 and 6 weeks,
Avoid both live and non-live vaccines for 3 months
No Disease defervescence (within 48 hrs)
Consider
A) 2nd dose IVIG
B) Prednisolone
Could the child have PAN, JIA or malignancy?
Kawasaki
TomharJun2005

Figure 1. Assessment for Suspected Incomplete Kawasaki Disease[a]



[a] Algorithm is based on expert committee's opinions and is not evidence-based. [b] See **TABLE 1** *for primary clinical features. [c] These criteria include elevated ALT, albumin ≤3.0 g/dL, platelets ≥45,000/mm³ after 7 days, WBC count ≥15,000/mm³, anemia for age, and urine ≥10 WBCs/high-power field. [d] For positive ECHO, 3 conditions must be met: LAD or RCA z score ≥2.5, coronary arteries meeting JMH criteria for aneurysm, or existence of ≥3 other suggestive features (decreased LV function, lack of tapering, PE, mitral regurgitation, perivascular brightness, LAD or RCA z score 2-2.5).*

ALT: alanine transferase; CRP: C-reactive protein; ECHO: echocardiogram; ESR: erythrocyte sedimentation rate; JMH: Japanese Ministry of Health; KD: Kawasaki disease; LAD: left anterior descending artery; LV: left ventricular; PE: pericardial effusion; RCA: right coronary artery; WBC: white blood cell.

Source: Reference 2.