Acquired Heart Disease in Children

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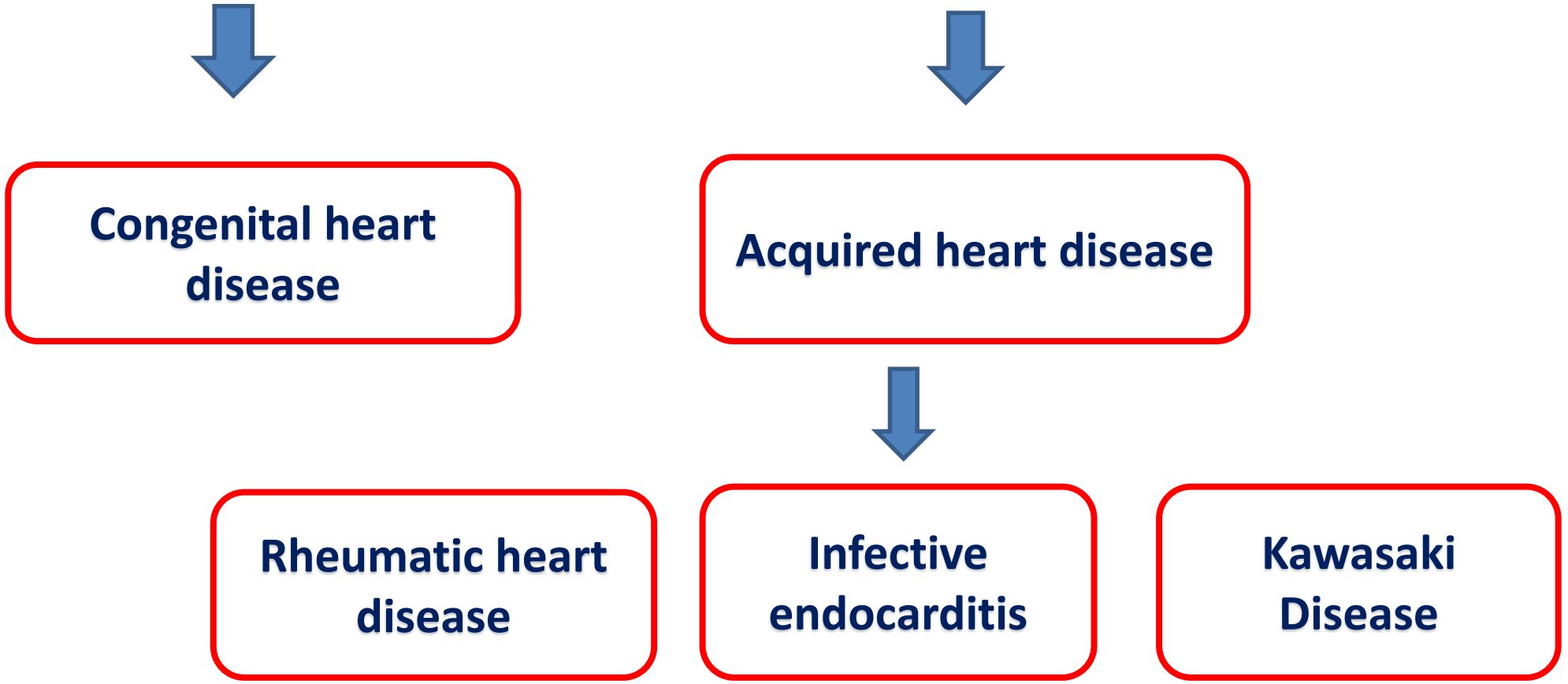
Heart Disease in Children



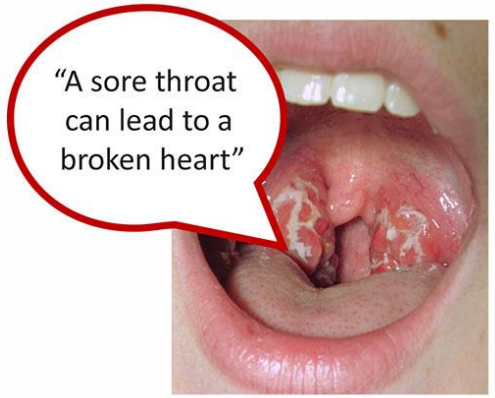
Acute rheumatic fever and rheumatic heart disease

Rheumatic Fever
Introduction

- Leading cause of acquired heart disease in children in developing countries
- Incidence is higher in areas with overcrowding
- Disease may lead to serious long term cardiac sequale if not properly treated

Rheumatic Fever
Aetiology

- Type II immune reaction following an infection of group A streptococcal pharyngitis
- Usually occurs in 2-4 weeks, following pharyngitis
- Rheumatogenic strains of GAS infection (serotypes 1, 3, 5, 6, and 18)
- Children of 5-15 years are at a greatest risk



Rheumatic Fever
Diagnosis

Clinical diagnosis

- Modified Jones criteria are used. Essential, major and minor criteria

- For diagnosis

  essential criteria

  &

  2 major criteria OR 1 major with 2 minor criteria

Rheumatic Fever - Diagnosis

Essential criteria + Major criteria + Minor criteria

Evidence of streptococcal infection

- Positive throat swab culture
- Positive streptococcal antigen test
- Positive anti strep antibodies

- Arthritis
- Carditis
- Chorea
- Erythema marginatum
- Subcutaneous nodules

- Arthralgia
- Fever
- Elevated ESR/ CRP
- Prolonged PR

Rheumatic Fever – Essential Criteria

Rheumatic Fever – Essential Criteria

Evidence of group A Streptococcal infection (GAS)

- Positive throat swab culture for GAS
- Positive antigen test for GAS
- Positive antibodies for GAS

    ASOT

    Anti- Dnase B

    Antihyaluronidase

If only a single antibody is measured (usually antistreptolysin O) sensitivity is 80-85% If all three are measured sensitivity rises up to 95-100%

Rheumatic Fever – Major Criteria
Clinical Features

Rheumatic Fever – Arthritis

- Present in 75% of patients with acute rheumatic fever
- Earliest feature to occur
- Involve **larger joints**, particularly the knees, ankles, wrists, and elbows
- Involvement of the spine, small joints and hips is uncommon
- Migratory polyarthritis
- Non deforming arthritis

Rheumatic Fever – Arthritis

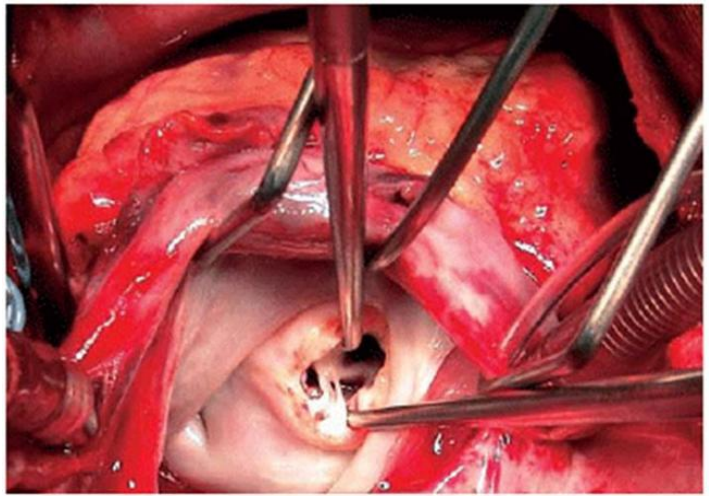
- Response to even small doses of salicylates
- Mono-articular arthritis is unusual

(unless anti-inflammatory therapy is initiated prematurely, aborting the progression of the migratory polyarthritis)

Inverse relationship between the severity of arthritis and the severity of cardiac involvement

Rheumatic fever – Carditis

- Most serious manifestation
- Aschoff nodules are characteristic on histology
- Manifested in 50-60% of patients
- Pancarditis (endocarditis, pericarditis and myocarditis)
- Endocarditis is a universal feature



Tachycardia, new onset murmur with Fever ? Rheumatic carditis

Carditis

- Carditis is always associated with a murmur of valvulitis
- Mostly isolated mitral valvular disease or combined aortic and mitral valvular disease
- Valvular insufficiency is characteristic of both acute and convalescent stages of acute rheumatic fever
- Valvular stenosis usually appears several years or even decades after the acute illness

Carditis

- **Mitral regurgitation** - high-pitched apical holosystolic murmur radiating to the axilla
- **Aortic insufficiency -** characterized by a high-pitched diastolic murmur at the lower left sternal border

- Right sided valvular lesions are rare
- Recurrent disease with initial carditis will have higher risk of subsequent carditis
- Mitral insufficiency may regress but not aortic insufficiency
- Long term – chronic progressive cardiac disease

Eryethema marginatum

Eryethema marginatum

- Pink or red macules, or papules which spread outwards in a circular shape, the edges become raised and red, and the centre clears.
- Not itchy or painful. They fade and reappear within hours and may persist intermittently for weeks to months.

- Occurs primarily on the trunk and extremities, but not on the face

Subcutaneous nodules

- Firm nodules approximately 1 cm in diameter along the extensor surfaces of tendons near bony prominences
- Non tender
- Associated with significant rheumatic heart disease

Chorea

- Occurs in 10-15% patients
- usually presents as an isolated, frequently subtle, neurologic behaviour disorder
- Emotional lability, incoordination, poor school performance, uncontrollable movements, and facial grimacing, exacerbated by stress and disappearing with sleep, are characteristic.
- Occasionally unilateral

Chorea

- **Milkmaid's grip:** Irregular contractions of the muscles of the hands while squeezing the fingers
- **Jack in the box sign**: darting movements of the tongue upon protrusion
- **Spooning:** the arms are extended, the wrist will flex and the metacarpophalangeal joints overextend
- **Pronator sign:** hyperpronation of the hands, with the palms facing outward when the arms are held over the head
- Permanent neurological sequale are rare

Minor criteria

- Arthralgia (in the absence of polyarthritis as a major criterion)

- Fever

Minor criteria

- Elevated acute-phase reactants (e.g., C-reactive protein, erythrocyte sedimentation rate)

- Prolonged PR interval on ECG (1st degree heart block)

(A prolonged P-R interval alone does not constitute evidence of carditis or predict long-term cardiac sequelae)

Rheumatic Fever – Diagnosis

Conditions in which Jones criteria need not to be fulfilled

- **Chorea** may occur as the only manifestation of acute rheumatic fever
- Characteristic **Rheumatic heart disease** as the only manifestation in patients who 1st comes to medical attention months after the onset of acute rheumatic fever
- **Recurrences** of acute rheumatic fever

Rheumatic Fever – Treatment

- **Bed rest** – need longer period if carditis with cardiomegaly/heart failure

- **Antibiotic therapy** – regardless of throat culture results

  Oral penicillin / erythromycin 10 days / single IM Benzathine penicillin to eradicate the organism

  After the initial course, start on long term prophylaxis

Rheumatic fever – Treatment

Carditis with no cardiomegaly or CCF

Aspirin only

Aspirin100 mg/kg/day in 4 divided doses for 3-5 days

75 mg/kg/day in 4 divided doses for 4 weeks

Carditis with cardiomegaly or CCF

Aspirin + Steroids

Prednisone 2 mg/kg/day in 4 divided doses for 2-3 wk

tapering dose by 5 mg/24 hr every 2-3 days
At the beginning of the tapering of the prednisone - Aspirin : 75 mg/kg/day in 4 divided doses for 6 weeks

Treatment of Sydenham chorea

- Anti-inflammatory agents are usually **not** indicated
- Sedatives may be helpful early in the course of chorea
- Phenobarbitone, sodium valproate, haloperidol or chlorpromazine

Rheumatic fever – Complications

- Chronic progressive rheumatic valvular heart disease
- Mitral stenosis, aortic stenosis and regurgitation
- Chorea and arthritis do **not** result in long term sequale

Rheumatic Fever – Prognosis

- Depends on the severity of the initial episode, and the presence of recurrences
- 70% of patients with carditis during the initial episode recover with no residual heart disease
- More severe the initial cardiac involvement, greater the risk for residual heart disease

Rheumatic Fever – Prevention

Primary prevention:

- Identification and treatment of the GAS that causes acute pharyngitis

Rheumatic fever – Prevention

Secondary prevention:

Continuous antibiotic prophylaxis to prevent recurrences

CATEGORY	DURATION
Rheumatic fever without carditis	5 yr or until 21 yr of age, whichever is longer
Rheumatic fever with carditis but without residual heart disease (no valvular disease*)	10 yr or until 21 yr of age, whichever is longer
Rheumatic fever with carditis and residual heart disease (persistent valvular disease*)	10 yr or until 40 yr of age, whichever is longer, sometimes lifelong prophylaxis

Rheumatic Fever – Antibiotic prophylaxis

Drug	Dose	Route
Penicillin G benzathine	600,000 U for children, ≤60 lb 1.2 million U for children >60 lb, every 4 wk	Intramuscular
or		
Penicillin V	250 mg, twice a day	Oral

Rheumatic Fever – Antibiotic prophylaxis

- Patients who have had acute rheumatic fever are susceptible to recurrent attacks following reinfection

- Patients with chorea, even in the absence of other manifestations of rheumatic fever, require long-term antibiotic prophylaxis

Rheumatic Fever – Follow up

- Good Compliance on antibiotic prophylaxis

In the presence of chronic valvular disease

- Good dental hygiene
- Antibiotic prophylaxis for major and minor surgeries

Kawasaki disease

Kawaski disease - Introduction

- First described in 1967 by Dr Tomisaku Kawasaki
- He reported 50 cases of a distinctive illness in children seen in Japan
- Febrile illness with mucocutaneous changes due to vasculitis
- Predominantly affects medium sized arteries. Predilection for coronary arteries
- Leading cause of acquired heart disease in developed countries

Kawaski Disease -Epidemiology

- Higher incidence in East Asia (Japan, Korea, Taiwan)
- MALE > FEMALE
- Common at younger age but uncommon less than 6 months
- 80% occurs in less than 5 year age group
- Median age of the illness is 2-3 years
- Coronary artery aneurysms occur in 20-25% of untreated patients

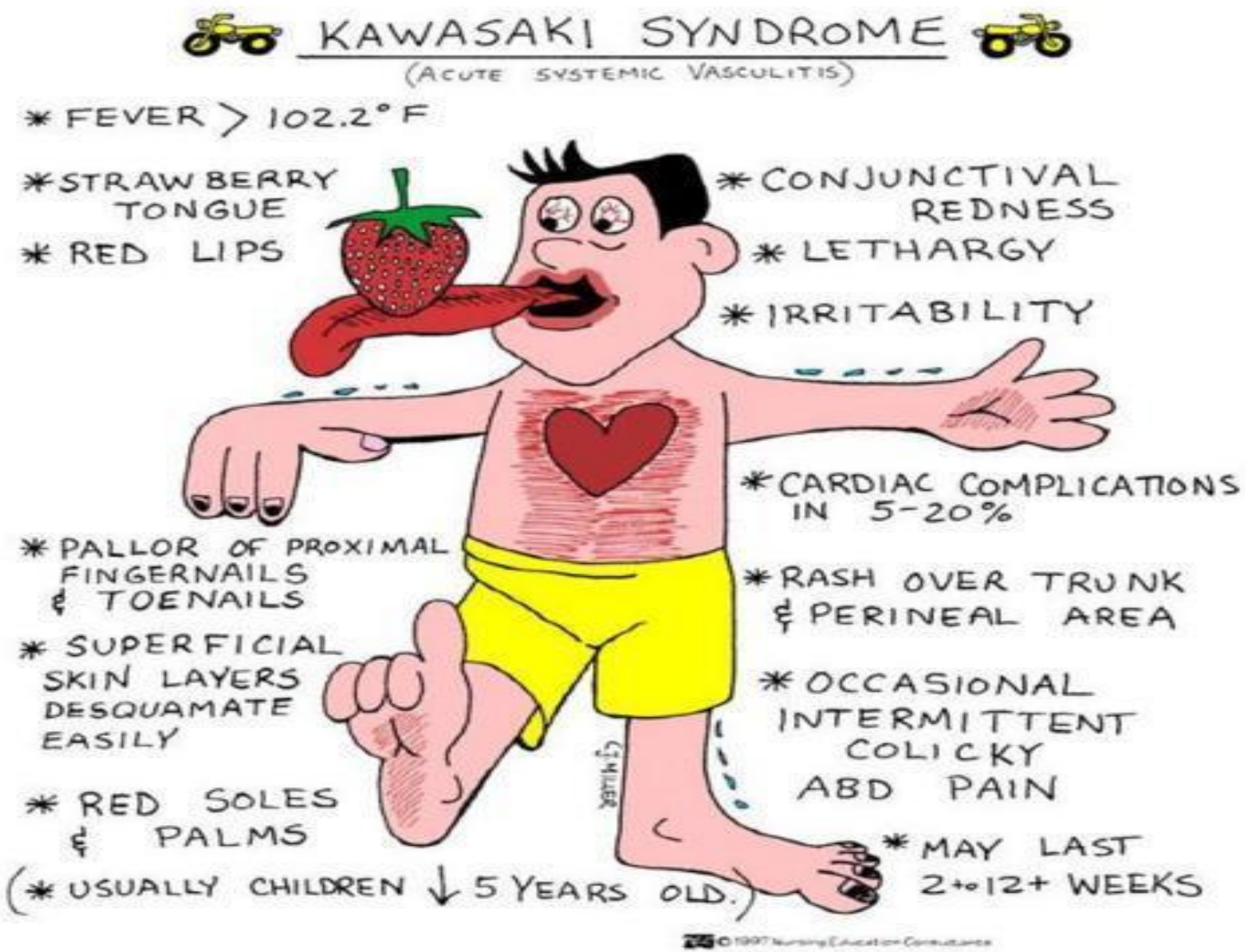
Kawasaki Disease - Aetiology

Unknown

Suggested theory

? Infectious trigger in genetically susceptible children

Kawasaki Disease – clinical features



Kawasaki disease – clinical features

Fever :

- High grade(equal or >101F)
- Abrupt onset, unremitting
- Associated with irritability
- Unresponsive to antibiotics
- Duration 1- 2 week (may persist 2-4 weeks)

Kawasaki Disease – clinical features

Conjuctival injection:

- Bilateral non-exudative, bulbar conjunctival injection with limbal sparing
- Seen in 90% of patients
- Painless
- Associated with anterior uveitis in 70%

Kawasaki Disease – clinical features

Oral mucosal changes:

- Erythema of the oral and pharyngeal mucosa with strawberry tongue and dry, cracked lips

- Seen in 90% of patients

Kawasaki Disease – clinical features

Changes in extremities:

- Seen in 50- 85% of patients
- Acute - oedema and erythema of the hands and feet
- Sub acute - periungual peeling of fingers and toes in weeks 2 and 3.
- Linear nail creases (Beau's lines)

Kawasaki Disease – clinical features

Skin eruption :

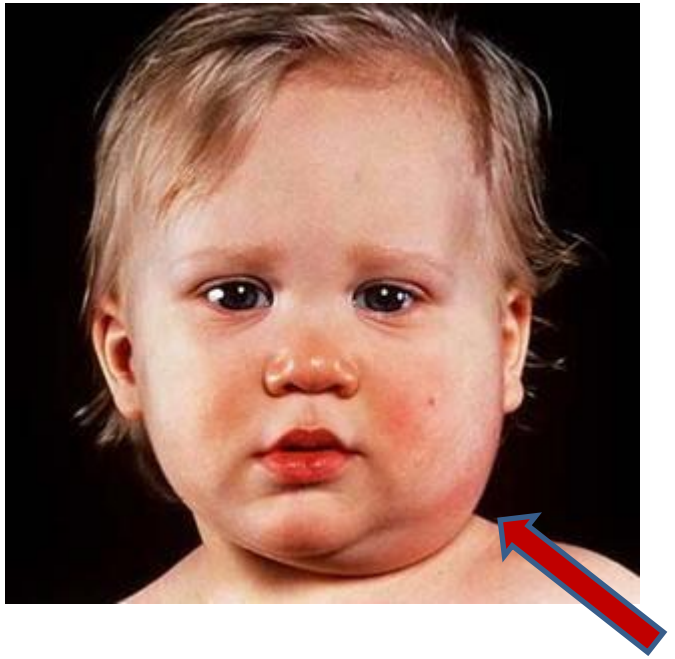
- Rash of various forms (maculopapular, erythema multiforme, or scarlatiniform) with accentuation in the groin area
- Seen in 70-90% of patients

Figure 3: Perineal desquamation in the acute phase of the disease

Kawasaki Disease – clinical features

Lymphadenopathy:

- Non-suppurative cervical lymphadenopathy
- Usually unilateral



Kawasaki Disease – Diagnosis

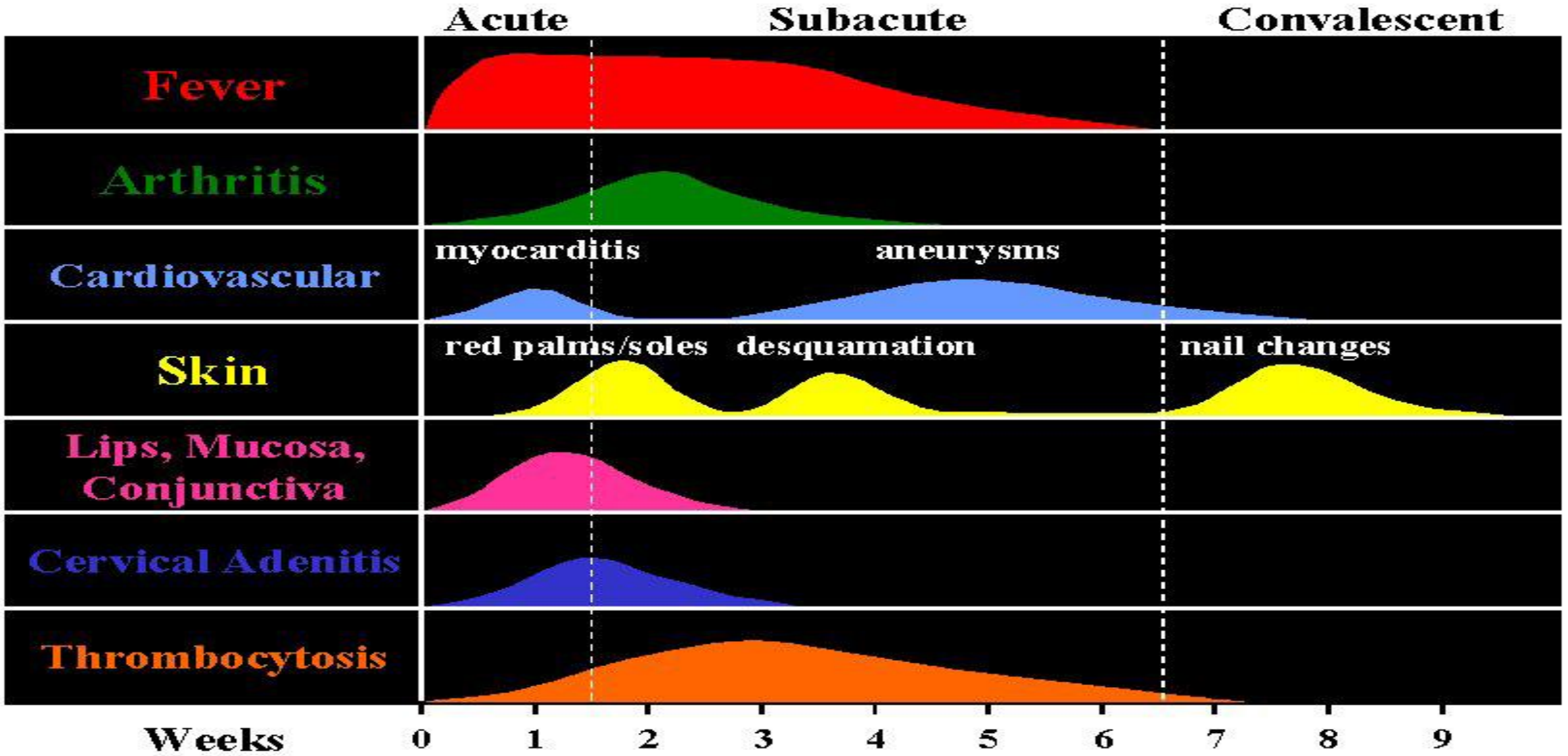
Clinical diagnosis

The acronym “FEBRILE” is used to remember the criteria as follows:

Fever for 5 days or more with at least 4 of 5 criteria

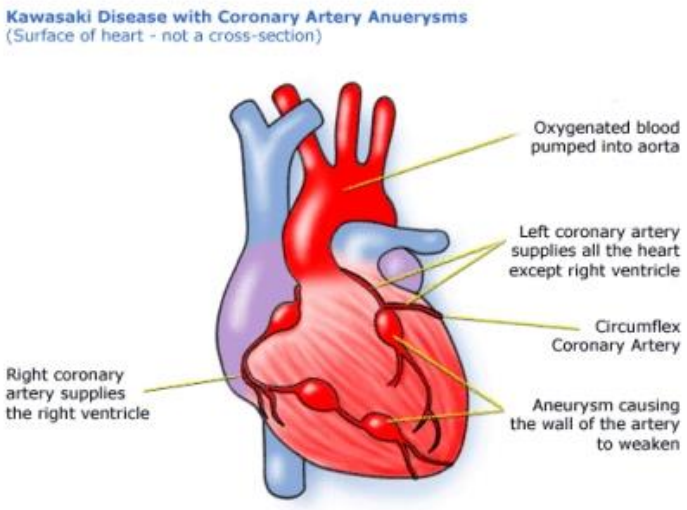
- Enanthem
- Bulbar conjunctivitis
- Rash
- *Internal organ involvement (**not part of the criteria**)*
- Lymphadenopathy
- Extremity changes

Clinical Manifestations of Kawasaki Disease



Cardiac involvement in Kawasaki Disease

- Coronary artery aneurysms - detected in 2-3 weeks
- Myocarditis
- Pericarditis
- Impaired contractility



Kawasaki Disease - Investigations

Full Blood Count

- Neutrophil leuckocytosis with immature forms
- Normochromic normocytic anaemia
- Thrombocytosis after 1st week

Inflammatory markers

- Elevated CRP
- Elevated ESR

Kawasaki Disease - Investigations

Urine Full Report

- Sterile pyuria on microscopy

Liver profile

- Elevated transaminases
- Hyperbilirubinaemia

USS abdomen

- Hydrops of the gall bladder

Cerebro-spinal Fluid

- Pleocytosis, high protein, Hypoglycorrhachia

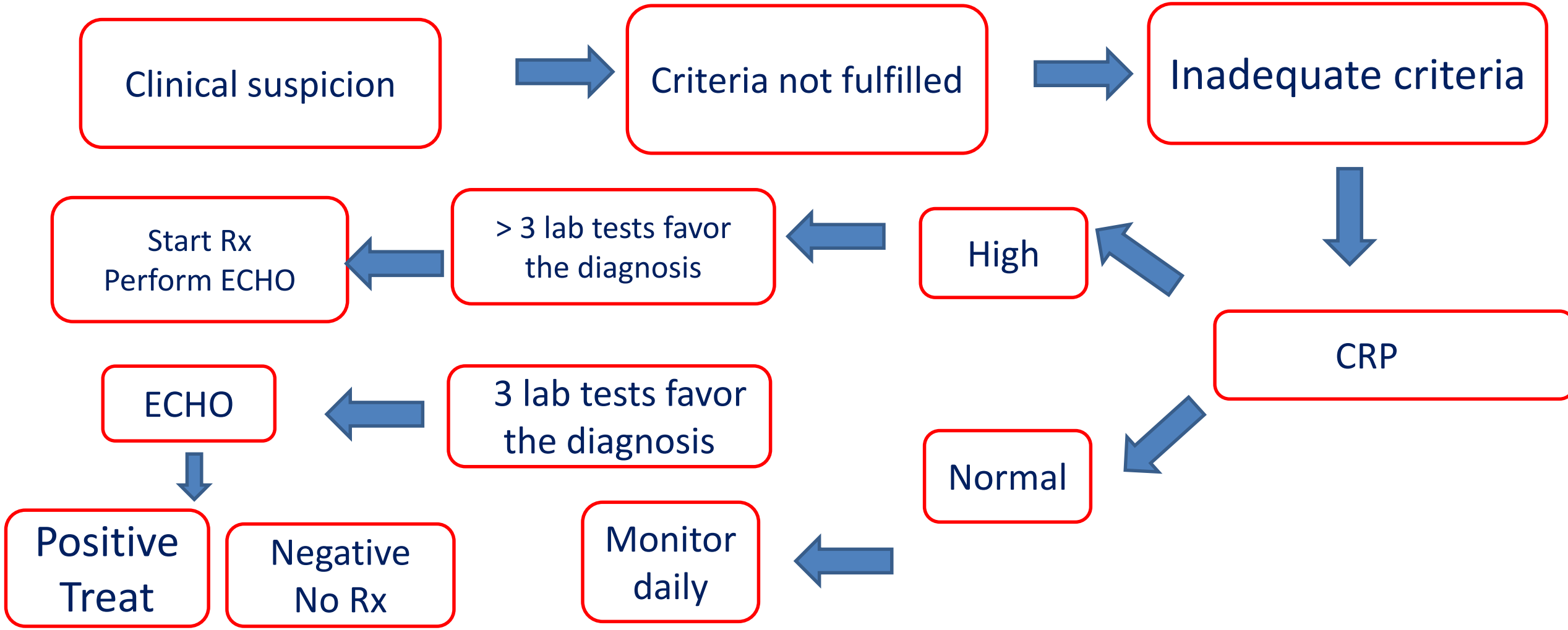
Kawasaki Disease - Investigations

Echocardiography:

- Is mandatory to visualise the presence of coronary artery aneurysms
- Should be performed at diagnosis and again after 2-3 weeks of illness and 6-8 weeks after the onset of the disease
- In patients without coronary abnormalities repeat ECHO at least annually

Atypical / Incomplete Kawasaki

- Criteria for diagnosis of the disease is not fulfilled
- Usually 2 or 3 criteria



Risk factors for coronary artery aneurysms

- Young age <1 year
- Male gender
- Pronged fever or recurrence of fever after an afebrile period of 48 hours
- Cardiomegaly
- Neutrophilia > 12 000/microlL PCV> 35%, thrombocytopenia < 350,000/microlL, hepatic transaminase elevation, hyponatremia, hypoalbuminemia < 3.5, and elevated C-reactive protein levels

Kawasaki Disease - Treatment

- Intravenous immunoglobulin (IVIG) 2 g/kg over 10-12 hours as a single dose
- High-dose aspirin (80-100 mg/kg/day divided)
- Treatment should be started as soon as possible after diagnosis (ideally within 10 days of disease onset)
- Prompt treatment will reduce the incidence of aneurysms up to 2-4%

Live vaccines (MMR & Varicella should be postponed for 11 months) following IVIG therapy

Kawasaki Disease - Treatment

- The dose of aspirin is usually decreased from anti-inflammatory to antithrombotic doses (3-5 mg/kg/day as a single dose) after the patient has been afebrile for 48 hours

- Aspirin is continued for its antithrombotic effect until 6 to 8 weeks after illness then discontinued in patients who have had ECHO

- Patients with coronary artery abnormalities continue with aspirin therapy and may require anticoagulation, depending on the degree of coronary dilation

Kawasaki Disease - Prognosis

- Timely treatment will reduce aneurysms
- Recurs in 1-3 % of cases

- Overall, 50% of coronary artery aneurysms regress to normal lumen diameter by 1 to 2 years after the illness. Smaller aneurysms being more likely to regress

- Giant aneurysms are unlikely to resolve and are most likely to lead to thrombosis or stenosis

Kawasaki Disease – Differential Diagnosis

- Viral infections – Measles, Adenovirus , CMV , EBV , Coxsackie
- Bacterial infections – Scarlet fever , Rocky mountain spotted fever , Leptospirosis , Bacterial cervical lymphadenitis
- Systemic Lupus Erythematousis
- Toxic shock syndrome
- Staphylococcal scalded skin syndrome
- Drug reactions
- Steven-Johnson syndrome