Cases in Acquired Pediatric Heart Disease Common Cases With a Twist

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9 year old Ali

- Ali is a previously healthy 9 yr old boy
- Referred to me via his pediatrician because of a heart murmur
- Although asymptomatic from a cardiac standpoint, he has a neurologist's appointment for newly started involuntary movements

Further Hx

- There is no Hx of sore throat or viral illness
- No skin rash or nodules, no joint pain or swelling
- No fever, chills or rigors
- No Hx of previous cardiac surgery
- No similar family Hx

Examination

- Obese child not in pain or distress
- There is a 3/6 pan-systolic murmur at the LMSB, radiating to the axilla
- You notice frequent non-purposeful movements of the arms and hands that the patient tries to suppress
- Rest of the exam is unremarkable

Investigations

- Blood count, renal and hepatic profiles were unremarkable
- ESR and CRP were normal
- ASO was negative
- ECG shows biphasic and prolonged p wave in V1
- Echocardiogram shows thickened mitral valve leaflets with moderate mitral regurgitation with LA dilation. Aortic valve leaflets are also thickened with mild regurgitation.

DIAGNOSIS?

Rheumatic fever, the modified Jones Criteria

- Major criteria

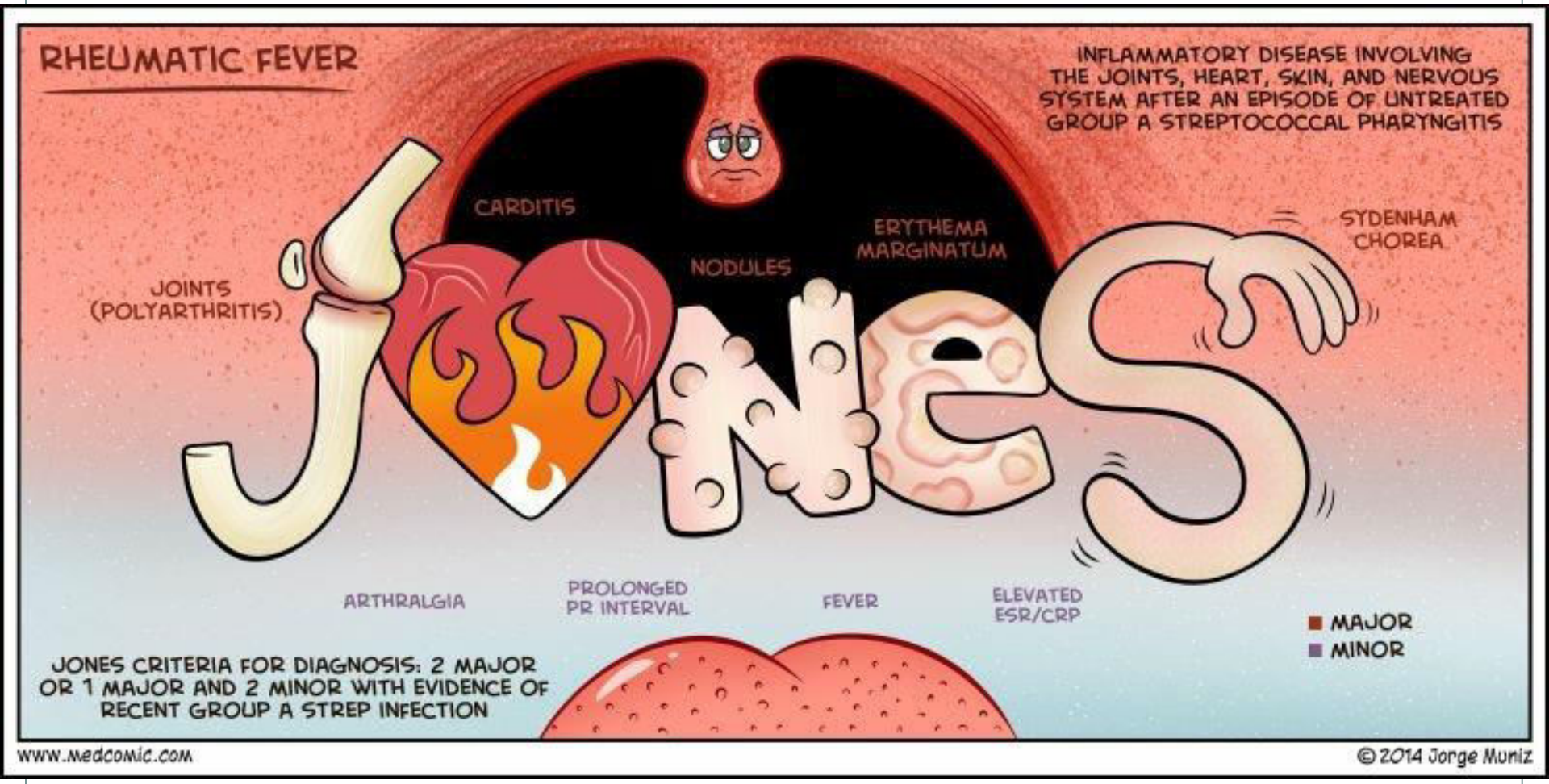
1. Carditis
2. Polyarthritis
3. Erythema marginatum
4. Subcutaneous nodules
5. Sydenham's chorea

- Minor criteria:

1. Fever
2. Arthralgia
3. Elevated acute phase reactants
4. Previous history of RF
5. Prolonged PR interval

*Have to have evidence of antecedent Strep infection: ASO / Strep antibodies / Strep group A throat culture / Recent scarlet fever / anti-deoxyribonuclease B / anti-hyaluronidase/ rapid test

** Two Major criteria or one major and two minor


RHEUMATIC FEVER
INFLAMMATORY DISEASE INVOLVING THE JOINTS, HEART, SKIN, AND NERVOUS SYSTEM AFTER AN EPISODE OF UNTREATED GROUP A STREPTOCOCCAL PHARYNGITIS
CARDITIS
ERYTHEMA MARGINATUM
SYDENHAM CHOREA
NODULES
JOINTS (POLYARTHRITIS)
JONES
ARTHRALGIA
PROLONGED PR INTERVAL
FEVER
ELEVATED ESR/CRP
MAJOR
MINOR
JONES CRITERIA FOR DIAGNOSIS: 2 MAJOR OR 1 MAJOR AND 2 MINOR WITH EVIDENCE OF RECENT GROUP A STREP INFECTION
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Other possible scenarios

- Chorea as the only manifestation
- Indolent carditis as the only manifestation in patients who come to medical attention months after GAS infection
- Recurrent ARF in patients with history of RF in the past

Rheumatic Fever

- Caused by Group A streptococcus pharyngitis
- Age group 5-15 years are most affected
- The body generates antibodies to fight the bug
- Because of similarity (mimicry) between some bacterial and human antigens, the antibodies might result in damage to native tissue (Joints, heart, skin and nervous system)

Epidemiology

- 470,000 new cases, 233,000 deaths a year
- Still endemic in our region
- Overcrowding, poor hygiene and limited access to health care are risk factors

Natural History

- Usually happens 2-4 weeks after throat infection with GAS but can happen with carrier patients with no history of pharyngitis.
- Affects the heart, CNS, joints and skin
- Heart disease results in most of the mortality and morbidity

Arthritis

- Usually migrates affecting large joints in quick succession
- Responds well to NSAIDs
- Does not leave damage to the joints

Differentiating ARF from PSRA

- Latent period shorter in PSRA (1-2 weeks)
- Arthritis responds to ASA better in ARF
- No carditis in PSRA
- Acute phase reactants usually higher in ARF
- Tenosynovitis or renal involvement more with PSRA

Carditis

- Causes pancarditis (pericardium, epicardium, myocardium and endocardium)
- Although significant damage can be caused by one episode, most of the damage is caused by recurrent episodes
- Valves most affected: Mitral alone, both mitral and aortic, aortic alone (left sided always affected)

Erythema marginatum or annulare

- Pink or faintly red non-pruritic rash on the trunk and sometimes the extremities but not the face
- Extends to the outside with return of normal skin in the center
- Not a frequent manifestation (5%)

Subcutaneous nodules

- The least common manifestation of ARF
- Firm, painless lesions.
- Usually located over bony surfaces or tendons
- Surface of the skin not inflamed, and is movable

Sydenham's Chorea (St. Vitus dance)

- Most common form of chorea in childhood
- Can be a very late manifestation (months after GAS)
- Non-rhythmic involuntary movements, muscular weakness and emotional disturbances
- Usually improves during sleep

The Lines of Management

- Eradication of GAS
- Symptomatic relief of acute disease manifestations
- Prophylaxis against future GAS infection to prevent recurrence

GAS Eradication

- Treat with antibiotics even if pharyngitis not present now
- Screen all family members, treat who test positive even if asymptomatic

Carditis and arthritis

- Anti-HF meds in severe cases
- High dose ASA (80-100 mg/kg/day) in children and 4-8 g/day in adults till symptoms are gone or inflammatory markers are normalized
- Use of steroids is controversial (unlikely useful except for resistant arthritis)
- Valve surgery

Chorea

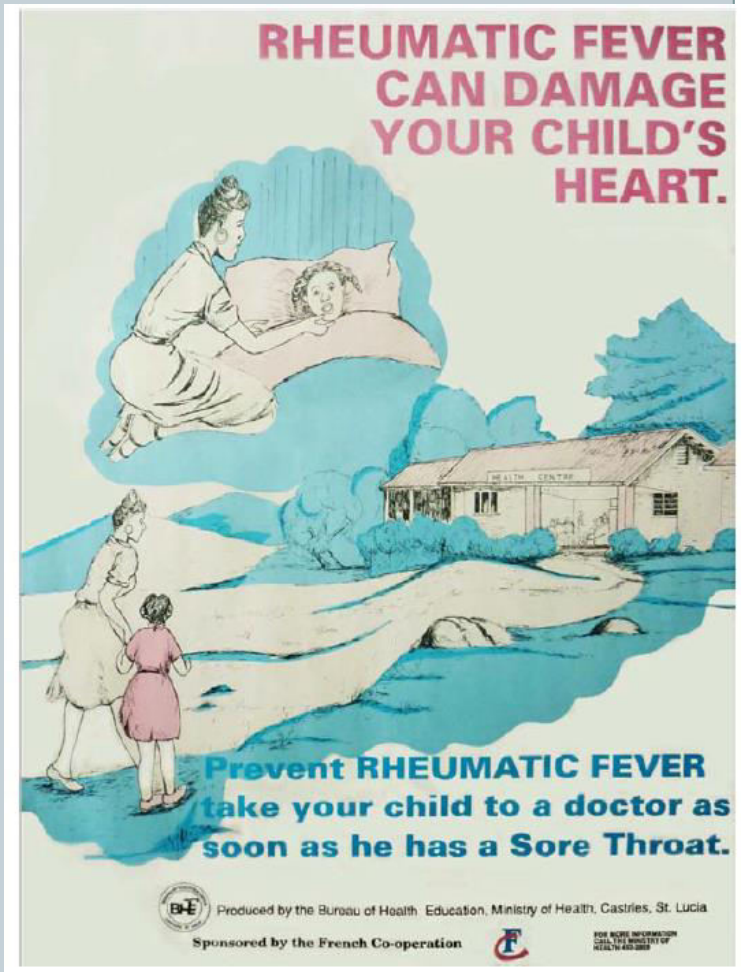
- Usually self-limited
- Responds to treatment with haloperidol
- Other modes of treatment: valproic acid, phenobarbital, diazepam, chlorpromazine, steroids, plasma exchange

Primary Prevention

- Primary prevention: by identifying patients with GAS pharyngitis and treating them promptly

- Can be difficult to achieve as one third of ARF patients do not have apparent infection

- Can use Oral penicillin V, Amoxicillin, single dose of IM penicillin G benzathine, Cephalexin, azithromycin, clarithromycin or clindamycin



Secondary prophylaxis

- Can use Penicillin G benzathine every 3-4 weeks (best results)
- Oral penicillin V twice daily
- Sulafadiazine once daily
- Azithromycin daily

Secondary prophylaxis duration

- Secondary prevention: duration is unclear but depends on number of previous episodes and the presence of risk factors

1. ARF with carditis and residual heart disease: 10 years or until 40 years of age or for life.
2. ARF with carditis but no residual disease: 10 years or until 21 years of age (whichever longer)
3. ARF without carditis: 5 years or until 21 years of age (whichever longer)

Late manifestations

- Rheumatic heart disease is the most severe manifestation of ARF
- Mitral valve is the most affected resulting in calcification, stenosis and +/- regurgitation
- Rarely recurrent arthritis can lead to Jaccoud arthropathy

Take Home Message

- A diagnosis of ARF can be made even with negative ASO and inflammatory markers

9 month old Juri

- Juri presented to our ER with fussiness, prolonged fever, pink eyes and swollen lips.
- She also has runny nose and there are multiple sick contacts at home

Examination

- Very difficult to examine
- Febrile with 39.6 C. No rash seen
- Teary red eyes without pus
- No cervical LAP
- Tongue is very red, lips are mildly swollen
- Hands and feet are normal
- She's tachycardic and a flow murmur is heard.

Work-up

- CBC showed elevated white cell (18,000) count with left shift. Hg is 9 g/dl
- CRP and ESR are very elevated
- Albumin 2.9 g/dl
- platelets are 110,000
- LFTs were mildly elevated (in the 100-200 range)
- WBCs seen in urine but no bacteria
- Chemistry shows hyponatremia (129 meq/L)

DIAGNOSIS?

Diagnostic criteria

- Fever > 5 days (must have)

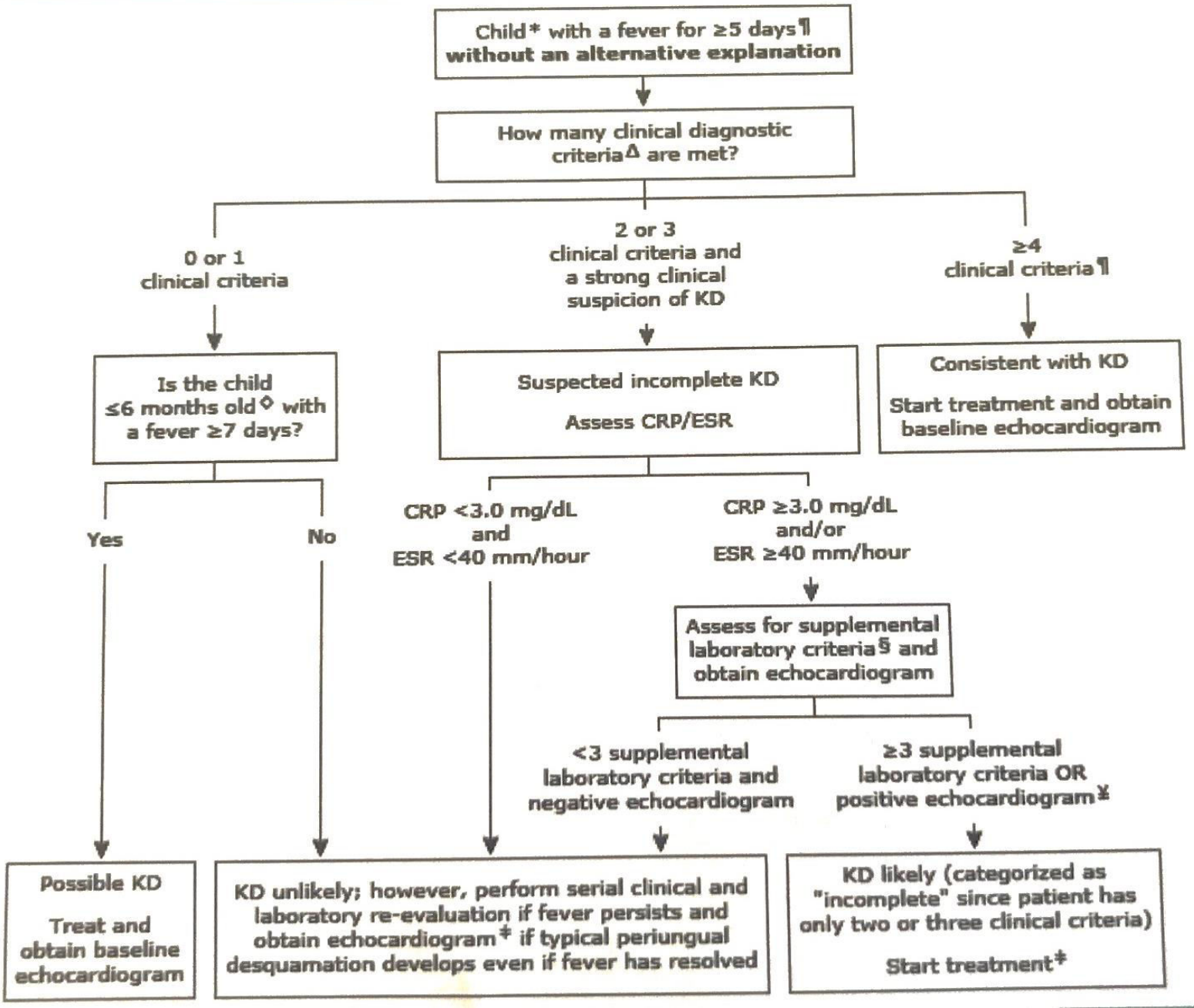
- And 4 out of 5:

1. Polymorphous rash
2. Cervical lymphadenitis (>1.5 cm)
3. Changes in the lips and mucus membranes
4. Extremity skin changes (redness, swelling, peeling of the skin)
5. Non-purulent bulbar conjunctivitis

Incomplete Kawasaki disease

- Following the strict criteria for Kawasaki disease resulted in missing 10-15% of patients who have Kawasaki disease.
- Not a small percentage of those patients ended up developing coronary artery aneurysms.
- So what to do in-order not to miss those patients?

Evaluation of suspected Kawasaki disease



Kawasaki Disease

- Also called mucocutaneous lymph node syndrome
- Likely caused by an abnormal immune system response to an infectious agent
- Most common between 1-2 years of life
- Cases below 3 months or more than 8 years are rare

Pathology

- During the acute phase of the illness, microvasculitis occurs with predilection to the coronary arteries
- Coronary artery aneurysms develop in 15-25% of untreated patients
- Diffuse pancarditis can happen leading to cardiac dysfunction, AV valve regurgitation, conduction abnormalities and pericarditis.
- During the late phases, fibrosis can result in narrowing of the coronary arteries leading to stenosis and possible myocardial infarctions
- Elevated platelet count increases the risk of MI

Other manifestations

- Sterile pyuria
- Elevation in liver enzymes
- Arthritis or arthralgia
- Gallbladder hydrops

Acute Phase (First 10 days)

- Abrupt onset of high fever and irritability. Conjunctivitis resolves quickly. Redness of mucus membranes, fissuring, ulcerations, strawberry tongue. Rash can be of different kinds. Cervical LAP happens in 50% of patients. Fever duration 11-12 days average, but resolves quickly with treatment.
- Leukocytosis, thrombocytosis, high inflammatory markers, sterile pyuria, elevated LFTs, lipid abnormalities.
- Coronaries can get affected during this stage.

Subacute Phase

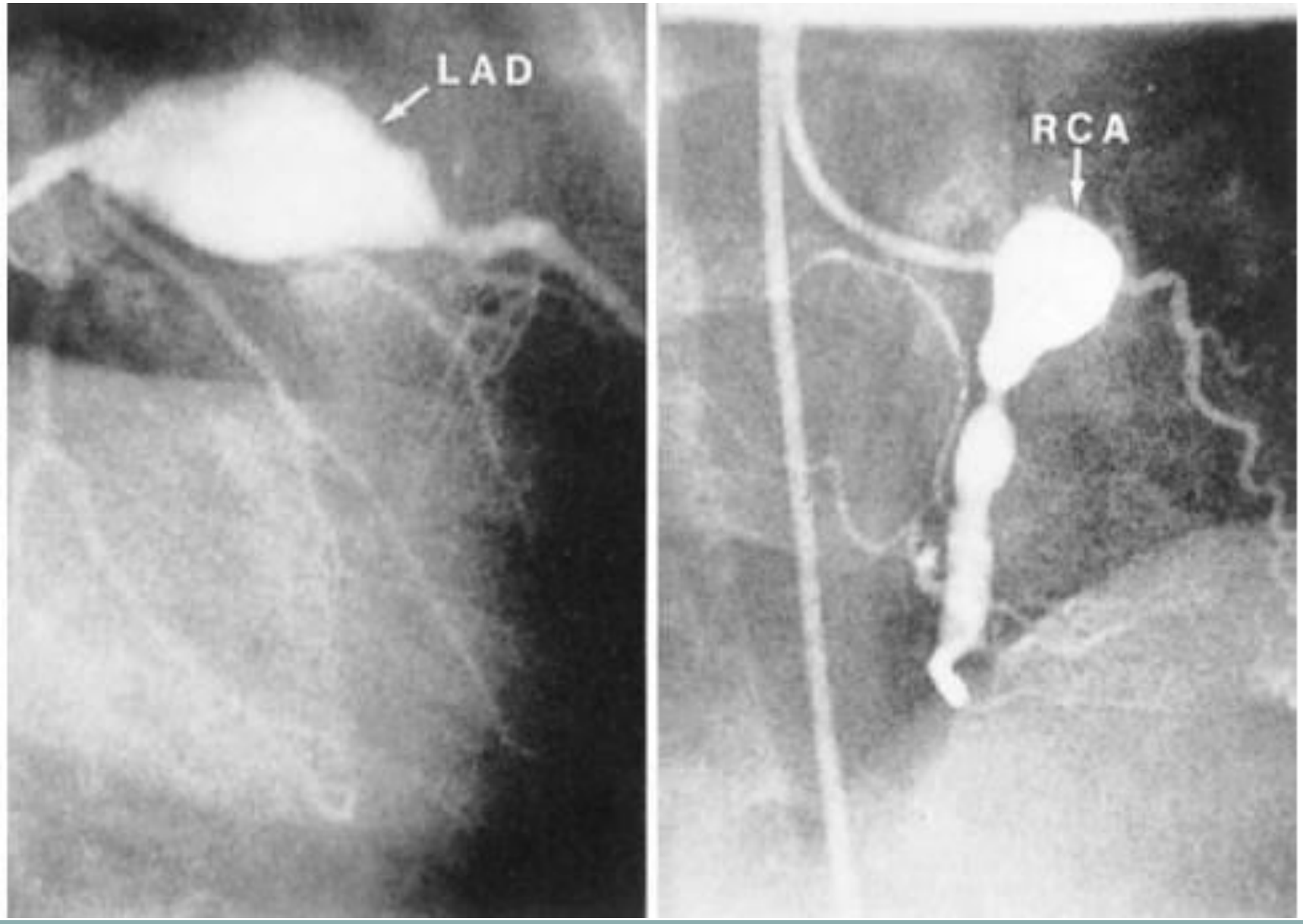
- Desquamation and peeling of the fingers and toes
- Rash, fever and LAP disappear
- Most of cardiovascular manifestations occur in this phase
- Worsening of thrombocytosis

Convalescent Phase

- This phase lasts till all inflammatory markers and platelets return to normal levels
- Beau's lines appear during this phase

Complications

- Cardiac manifestations dictate the prognosis
- Coronary aneurysms, thrombosis, stenosis
- Cardiac dysfunction, AV valve damage, dilation of the ascending aorta, effusion, heart block
- Peripheral artery aneurysms and stenosis


LAD
RCA

Management

- Admission
- High dose ASA (30-100 mg/kg/day)
- Anti-pyretics
- IVIG (2 g/kg IV infusion) +/- oral steroids. Repeat IVIG if no improvement
- Pulse steroids IV for non-responders
- Infliximab for resistant cases

Management

- Switch ASA to low-dose before discharge, some recommend not before at least 14 days of illness
- Stop ASA after platelets are normal, or 6-8 weeks after illness, whatever longer
- If coronary abnormalities persist, patient will be always on ASA
- In case of aneurysms, clopidogrel or even warfarin can be added

Question

- Would do you start our patient on steroids along with IVIG and ASA when the patient presented?

The Kobayashi score

- Sodium less than 133 mmol/L (2 points)
- AST >100 Units/L (2 points)
- CRP >10 mg/dl (1 point)
- Neutrophils >80% of WBC (2 points)
- Platelets less than 300,000/mm3 (1 point)
- Days of illness at initial treatment less than 5 (2 points)
- Age less than 12 months (1 point)

Follow up

- Depending on the level of coronary involvement

Risk Level	Pharmacological Therapy	Physical Activity	Follow-Up and Diagnostic Testing	Invasive Testing
I (no coronary artery changes at any stage of illness)	None beyond 1st 6–8 weeks	No restrictions beyond 1st 6–8 weeks	Cardiovascular risk assessment, counseling at 5-y intervals	None recommended
II (transient coronary artery ectasia disappears within 1st 6–8 weeks)	None beyond 1st 6–8 weeks	No restrictions beyond 1st 6–8 weeks	Cardiovascular risk assessment, counseling at 3- to 5-y intervals	None recommended
III (1 small-medium coronary artery aneurysm/major coronary artery)	Low-dose aspirin (3–5 mg/kg aspirin per day), at least until aneurysm regression documented	For patients <11 y old, no restriction beyond 1st 6–8 weeks; patients 11–20 y old, physical activity guided by biennial stress test, evaluation of myocardial perfusion scan; contact or high-impact sports discouraged for patients taking antiplatelet agents	Annual cardiology follow-up with echocardiogram + ECG, combined with cardiovascular risk assessment, counseling; biennial stress test/evaluation of myocardial perfusion scan	Angiography, if noninvasive test suggests ischemia
IV (≥1 large or giant coronary artery aneurysm, ormultiple or complex aneurysms in same coronary artery, without obstruction)	Long-term antiplatelet therapy and warfarin (target INR 2.0–2.5) or low-molecular-weight heparin (target: antifactor Xa level 0.5–1.0 U/mL) should be combined in giant aneurysms	Contact or high-impact sports should be avoided because of risk of bleeding; other physical activity recommendations guided by stress test/evaluation of myocardial perfusion scan outcome	Biannual follow-up with echocardiogram + ECG; annual stress test/evaluation of myocardial perfusion scan	1st angiography at 6–12 mo or sooner if clinically indicated; repeated angiography if noninvasive test, clinical, or laboratory findings suggest ischemia; elective repeat angiography under some circumstances (see text)
V (coronary artery obstruction)	Long-term low-dose aspirin; warfarin or low-molecular-weight heparin if giant aneurysm persists; consider use of β-blockers to reduce myocardial O_2 consumption	Contact or high-impact sports should be avoided because of risk of bleeding; other physical activity recommendations guided by stress test/myocardial perfusion scan outcome	Biannual follow-up with echocardiogram and ECG; annual stress test/evaluation of myocardial perfusion scan	Angiography recommended to address therapeutic options

Take home messages

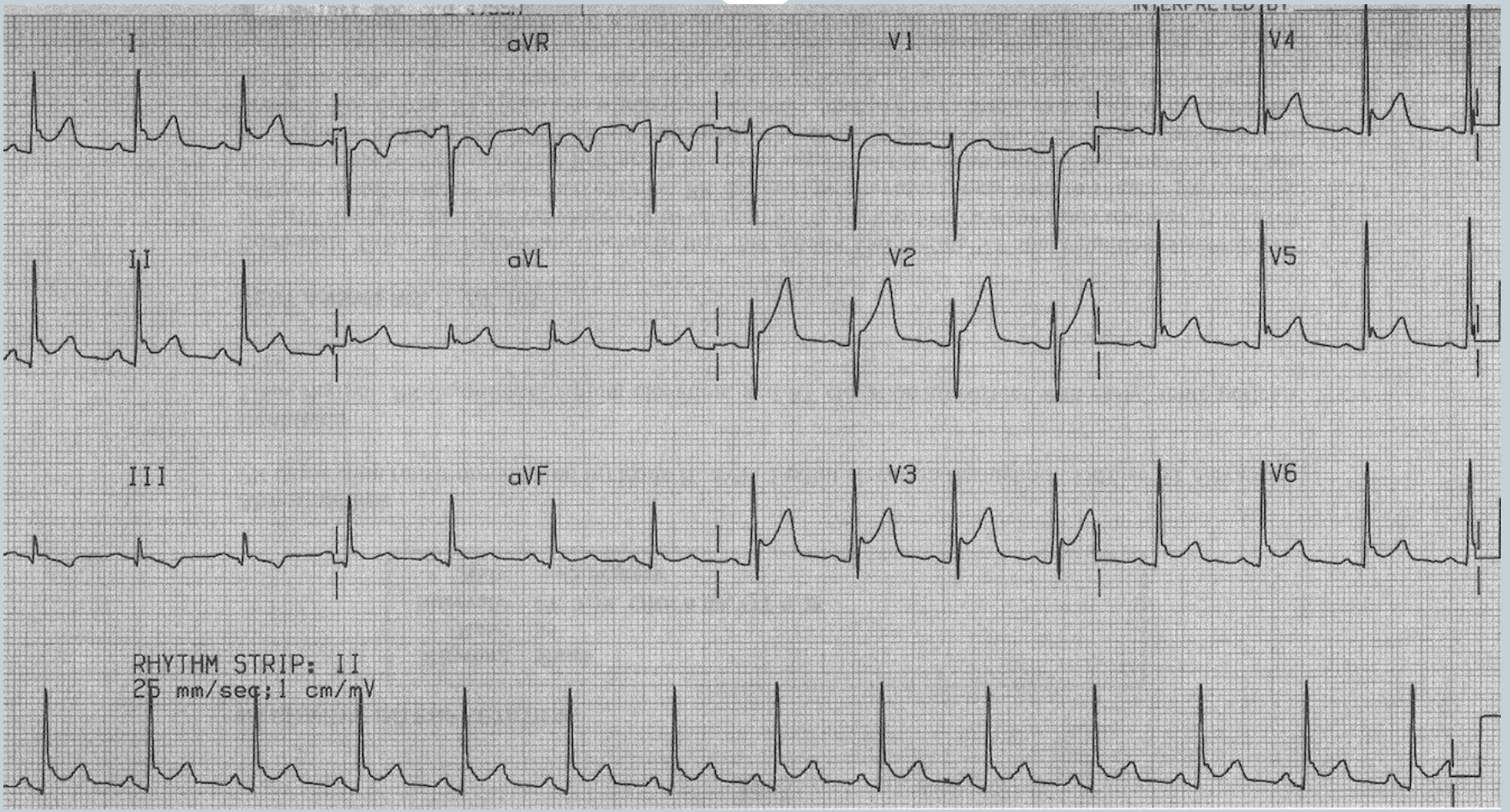
- You don’t have to fulfill 4 criteria to diagnose Kawasaki disease
- Infants less than 6 months with fever for more than 7 days, consider strongly treating as Kawasaki disease
- In patients with risk of IVIG treatment failure, start steroids at the same time of IVIG and ASA

Michelle

- 16 yr old female previously healthy
- Started having chest pain that is getting worse over the last couple of days
- Sharp, worse with breathing and leaning forward
- No SOB, palpitations or syncope

Examination

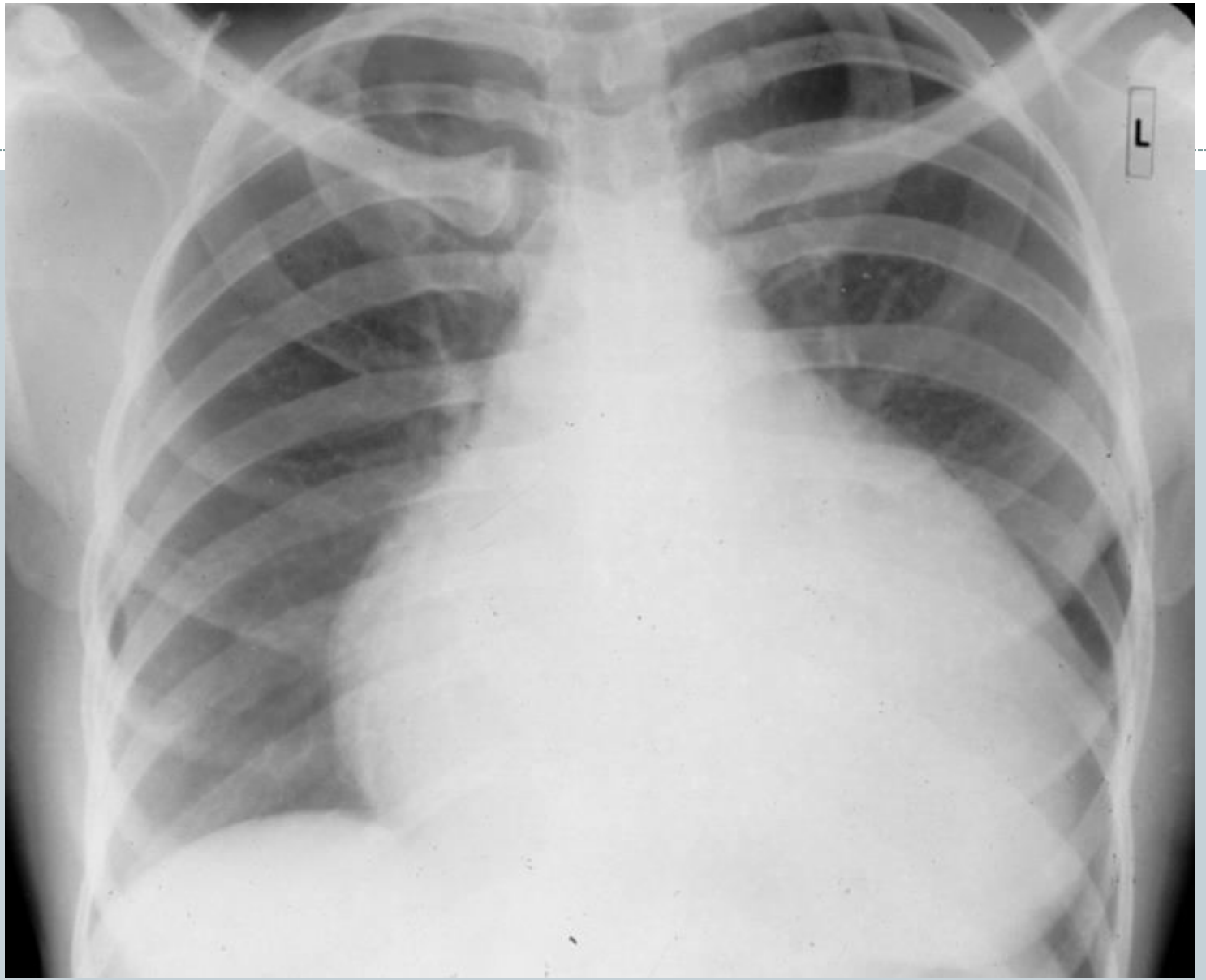
- There is a friction rub, otherwise exam is normal
- CXR showed mild cardiomegaly
- ECG showed elevated ST segment in anterior precordial leads.
- Echocardiogram showed small pericardial effusion.


I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
RHYTHM STRIP: II
25 mm/sec;1 cm/mV

DIAGNOSIS?

Follow up

- Patient was started on Ibuprofen and given a very close follow up
- After 1 week, the patient reported having SOB, fatigue and dizziness
- CXR showed enlarging heart shadow
- Echo showed large pericardial effusion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Follow up

- The patient was admitted and underwent pericardiocentesis
- Examination of the fluid showed elevated WBCs but cultures and viral studies were negative
- Patient was given colchicine in addition to the ibuprofen and on follow up, patient continued to do well.

Pericarditis

- The most common cause of CARDIAC chest pain in pediatrics
- Esp in pediatrics, pericarditis is most commonly caused by a viral infection, but in many cases, the cause won't be known.

Clinical manifestations

- Chest pain, Fatigue, SOB, syncope
- On exam, a friction rub might be heard
- Pulsus paradoxus if there is cardiac tamponade.

Work up

- Chest X-ray shows flask-shaped cardiomegaly in the presence of effusion
- ECG shows diffuse ST segment elevation with depression in V1 and aVR
- T-wave inversion can happen and usually persists with chronic pericarditis

Initial Work up

- Blood samples for markers of inflammation
- CBC, blood cultures (if febrile)
- Cardiac enzymes can be positive in 32% of patients
- CXR, ECG
- Echocardiogram shows effusion

Additional work up in atypical cases

- Tuberculin skin test
- HIV titers
- ANA
- CT scan
- MRI
- Pericardial fluid sample if pericardiocentesis done for diagnostic or therapeutic purposes

Treatment of acute pericarditis

- Activity restriction
- Anti-inflammatory medications like ibuprofen
- Colchicine to be initiated with NSAIDs
- Steroids for resistant cases
- Pericardiocentesis and drainage
- Pericardial window, pericardiotomy or pericardioectomy for resistant cases

Take Home message

- You don’t have to wait for refractory pericarditis to start colchicine. You can start it at the same time as NSAIDs

Thank you