Incomplete or Atypical KD

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Incomplete or Atypical Kawasaki Disease

1. Definition
2. Case Vignette
3. Clinical approach

Definition

Incomplete KD

- Fever lasting for more than 5 days with 2 or 3 of classical features
- The real incidence is difficult to define

Atypical KD

- Renal impairment
- Facial nerve palsy
- Testicular nodule
- Pleural effusion
- Diarrhea, abdominal pain
- Acute abdomen

Atypical KD/ Incomplete KD in children

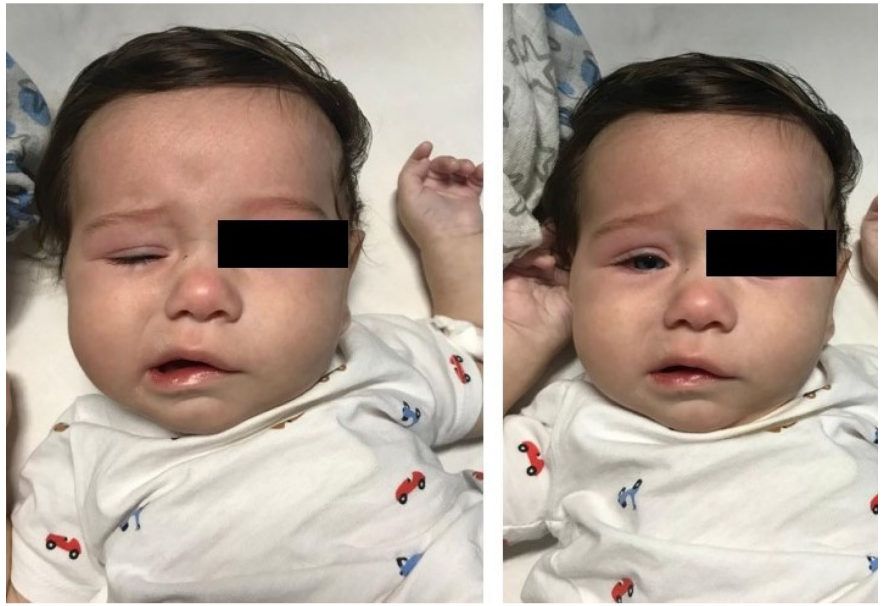
1. More common in young infants
2. Consider in any child fever > 5 days
3. Fewer than 4 classical criteria
4. High risk of Coronary arteries abnormalities
5. Compatible laboratory parameters
6. Coronary artery imaging
7. High index of suspicion is needed

Case vignette 1

- 7-month-old girl developmentally normal, was admitted to hospital with 18 days of fever
- She had a history of bilateral conjunctival injections
- Since 4-days developed drooping of the left side of her mouth

Differential diagnosis to be considered…

- Acute otitis media
- Chronic otitis media
- Meningitis



Would you consider the diagnosis of Kawasaki disease?

Physical examination revealed fever (38.5°C), lethargy, desquamation of the skin at fingertips, and LMN facial nerve palsy

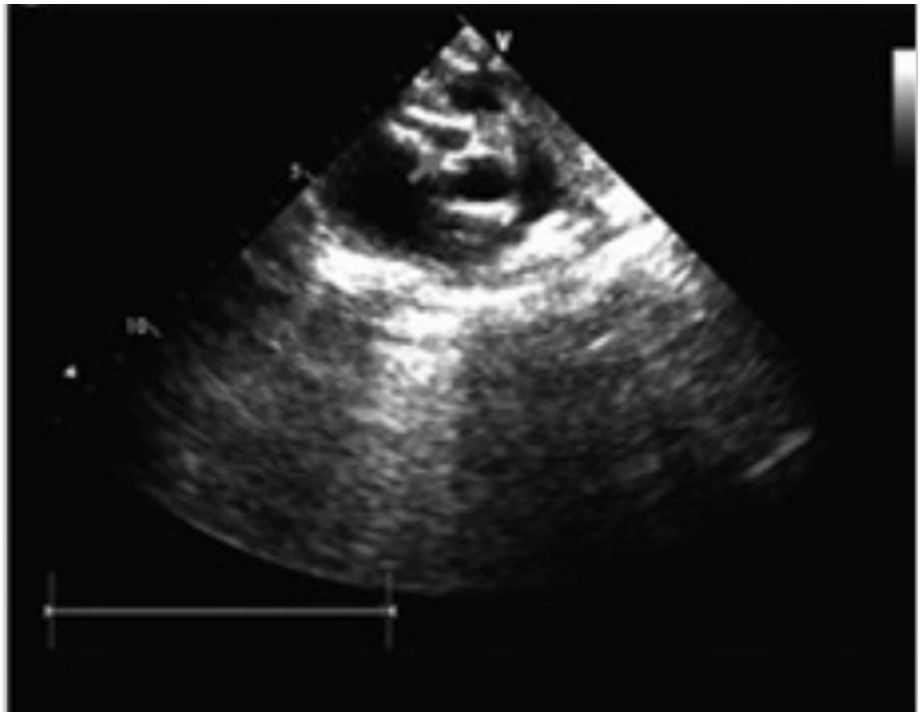
Investigations

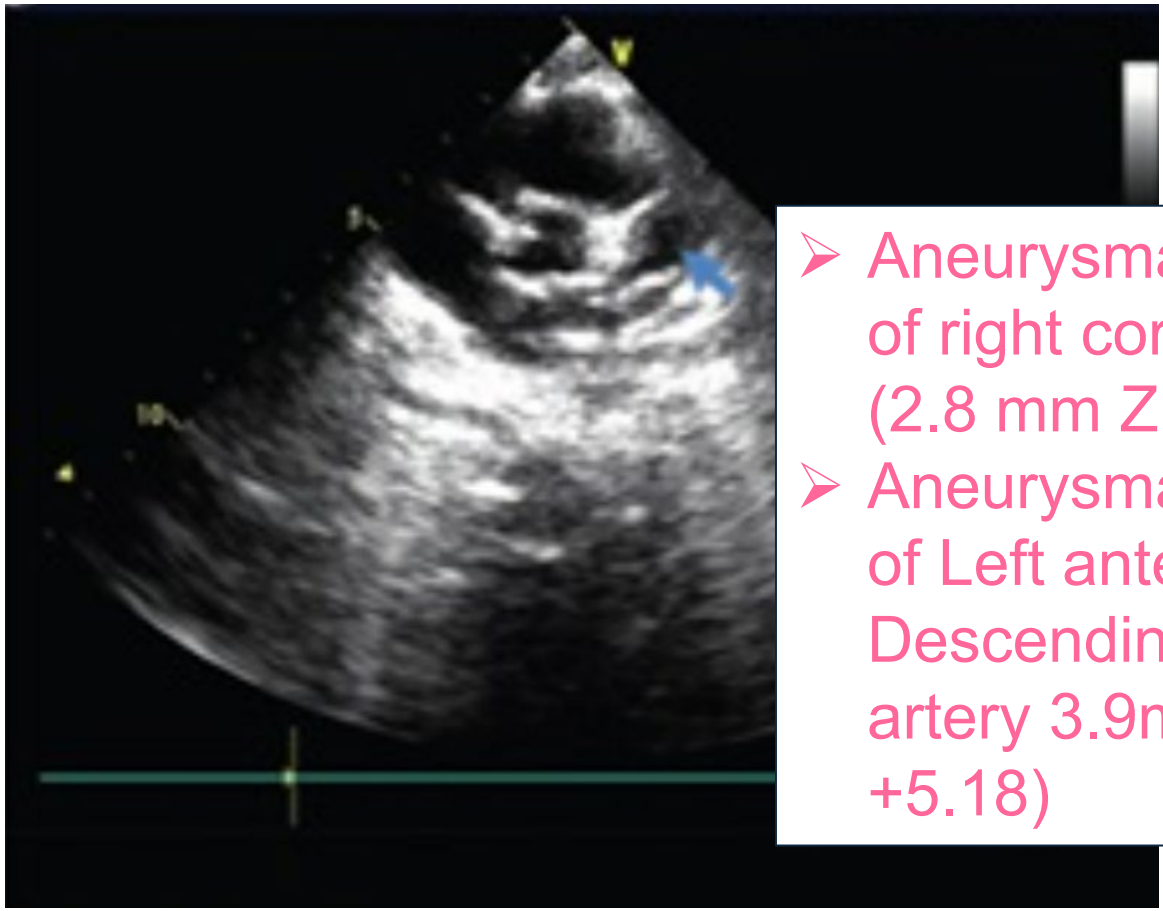
Laboratory parameters	
Hb	9.9g/dL
Total leucocyte count	18.11x10^9 /L
Platelet count	828 × 10^9 /L
ESR	120mm/h
CRP	70mg/L
CSF	36 cells
CSF Glucose & protein	Normal

- Anemia
- Leucocytosis
- Thrombocytosis
- Increased inflammatory parameters
- Aseptic meningitis

What will you do next?

2D-Echocardiography





- Aneurysmal dilatation of right coronary artery (2.8 mm Z Score +3.5)
- Aneurysmal dilatation of Left anterior Descending coronary artery 3.9mm (Z score +5.18)

Treated with IVIG 2/gm/kg and aspirin 30mg/kg/ in 4 divided doses

Key massage…

Facial palsy at times the manifestation of KD

Fever more than 5 days in any child KD should be a differential diagnosis

Increased inflammatory parameters are adjunct to the diagnosis of KD

2D Echocardiography crucial to identify coronary artery abnormalities

Case 2

- ✓ 4-year-old boy
- ✓ High grade fever x 7 days
- ✓ Unilateral neck swelling x5 days

On examination:

- ➢ Febrile with Temperature 104^0 F
- ➢ Extremely irritable
- ➢ Tender cervical lymphadenopathy over the left cervical region

What are the differential diagnosis ?

Case continued…

- There was no evidence of BCG site's reaction or perianal peeling
- Oropharyngeal examination – Normal

Differential diagnosis of bacterial lymphadenitis was considered

Received I.V. Antimicrobials x 5 days
However there was no improvement

What will you do next?

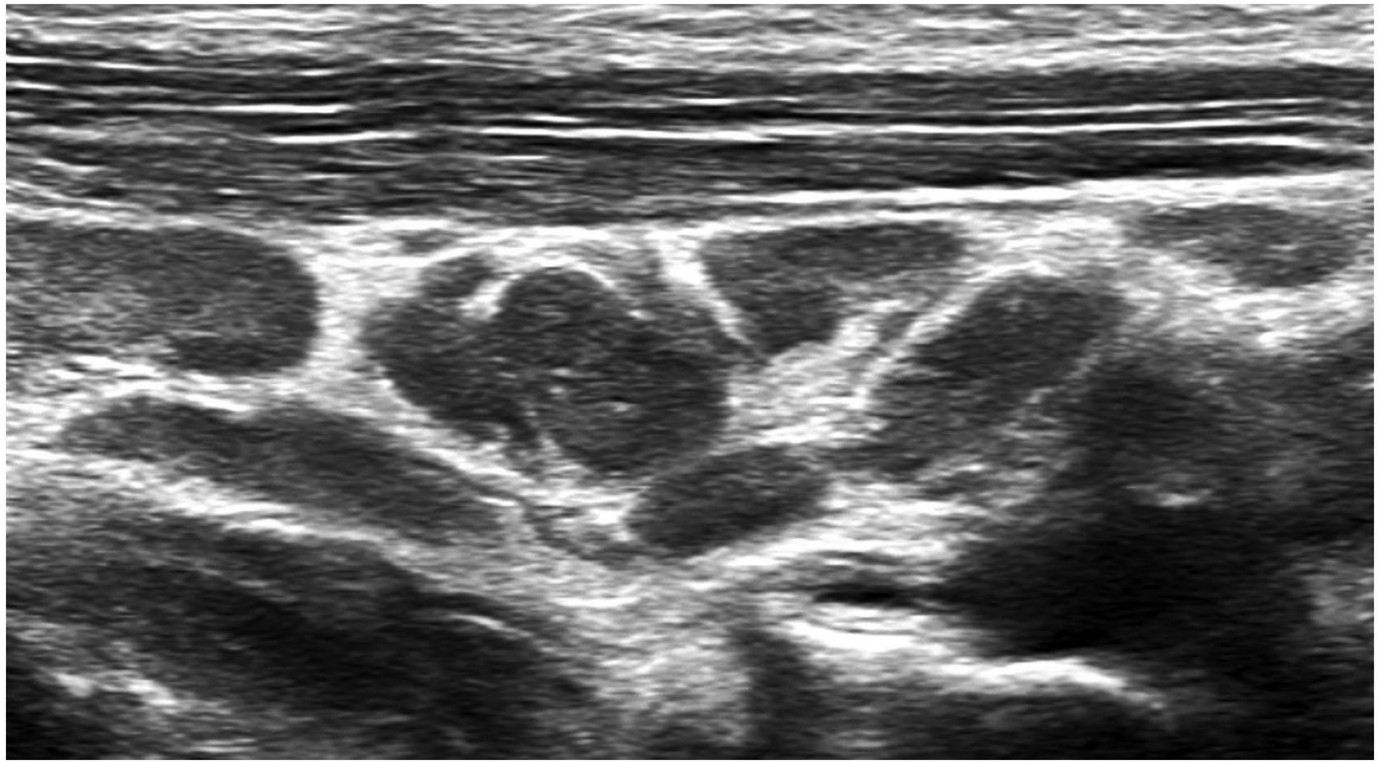
Investigations

- Haemoglobin - 8.5g/dL
- Total leucocyte count- $21.76x10^9/L$
- ESR- 56mm/1^{st} hour
- CRP –110mg/L

Case continued…

Neck Ultrasonography

Multiple enlarged uniformly hypoechoic well circumscribed lymph nodes without necrosis



"A cluster of grapes appearance"

Unique feature of KD lymphadenitis

2D Echo: Normal coronary arteries

Case continued…

- Final diagnosis- Kawasaki disease
- Received IVIg 2gm/Kg over 24 hours
- Aspirin 50 mg/kg till afebrile
- Lymph node swelling gradually subsides after 5 days admission

Key Message…

- Unilateral tender Lymph node enlargement at times the presenting features of KD
- Often termed as “Node First”
- Mistaken as infectious lymphadenitis
- Ultrasonography is an important tool demystifying the diagnosis bed side
- Lymph nodes are uniformly enlarged and hypoechoic without necrosis
- Well circumscribed margin and well visualised echogenic hilum

Case 3

18 months old boy

- Admitted with a 4-days of fever
- Associated with nasal congestions
- Cough and vomiting

- ✓ Had significant past history
- ✓ Urinary tract infection at 4 months of age
- ✓ Complex febrile seizure at 9 months

On examination:

Nasal congestion

Vitals- normal

Investigations:

- ✓ Total leucocyte counts: $14x10^9/L$
- ✓ ESR- 52mm 1^{st} hour
- ✓ CRP- 294mg/L
- ✓ Urine RE – 12 pus cells per HPF

Case continued…

What would be the next step?

He was started on Inj. Ceftriaxone as there was pyuria after sending urine culture

Urine culture: *Escherichia coli* (pan sensitive)

Continue to have fever despite antibiotics therapy

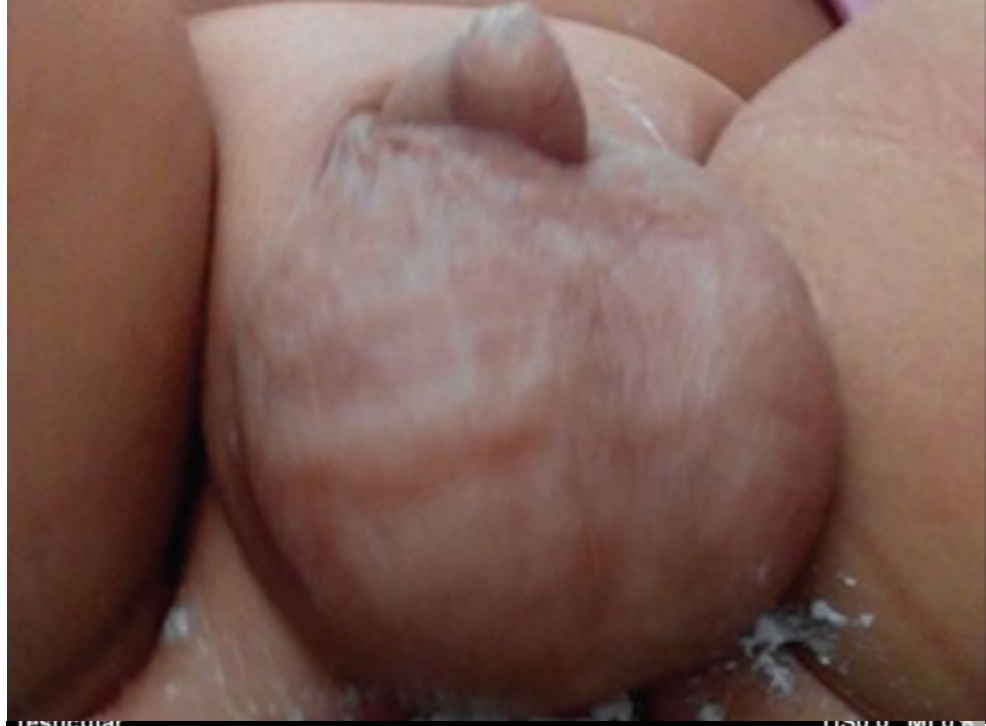
What should one do now?

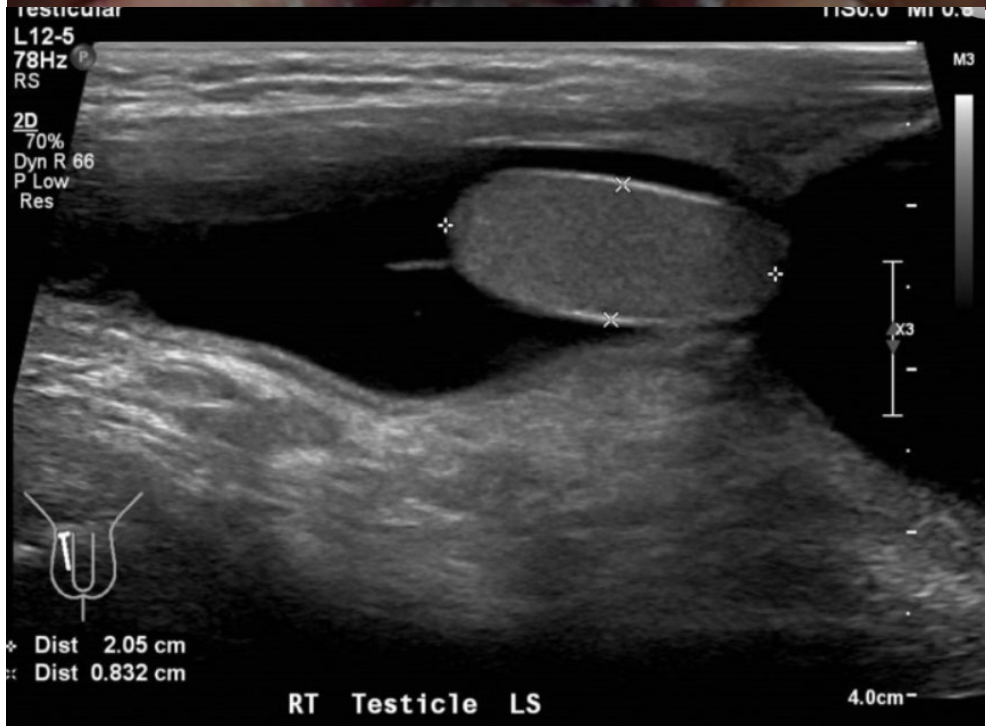
Ultrasonography of Abdomen: Left focal pyelonephritis without hydronephrosis

Case continued…

- Noted to have sudden onset scrotal swelling on day 8 of fever
- USG scrotum – Hydrocele
- Small right testis appendix
- Right hydrocele extended into the inguinal region, with patent processus vaginalis
- Also developed rash over the trunk on day 8 of fever

What should one do next?





Case continued…

Repeat investigations:

Haemoglobin: 10.5g/Dl

Total Leucocyte counts: 16.24x10^9/L

Erythrocyte sedimentation rate: 118mm

C- reactive protein - 220mg/L

Serum Na+ - 130meq/L

Serum albumin- 2.6mg/dL

SGOT – 20 IU/L

SGPT- 32 IU/L

Atypical Kawasaki Disease

Two of the classical features

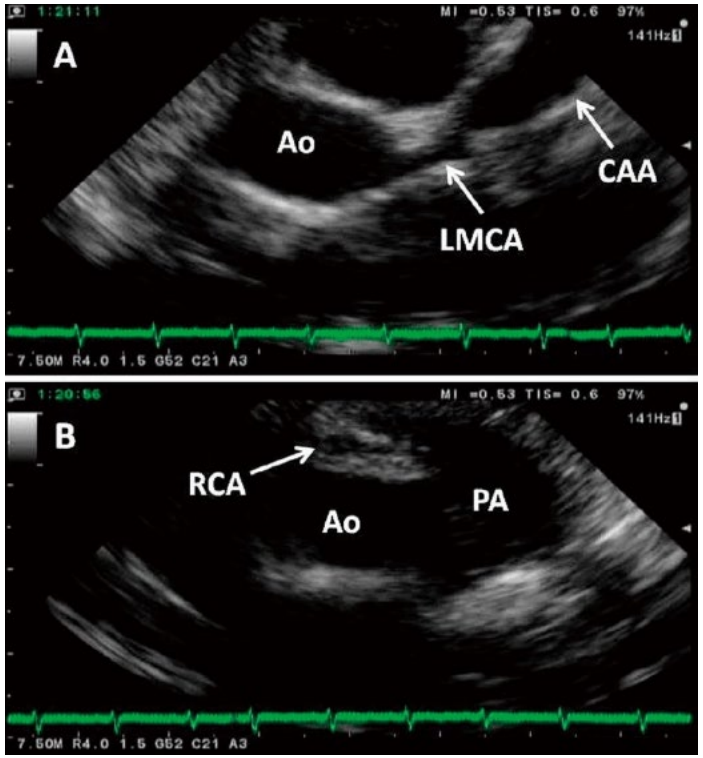
- Prolonged fever
- Rash
- Increased ESR, CRP
- Hypoalbuminemia
- Hyponatremia

Started on IVIg 2gm/Kg + aspirin 50mg/Kg and became afebrile

What would you do next?

Case continued…

- 2 D Echocardiography
- Aneurysmal dilatation of LMCA, RCA and left anterior descending artery



Key message…

- ❖ Hydrocele and scrotal swelling may be seen in Kawasaki disease
- ❖ Likely related to underlying inflammatory state leading to fluid extravasation
- ❖ Clinicians dealing with diagnostic dilemmas involving inflammatory processes should always consider Kawasaki Disease as a potential differential diagnosis

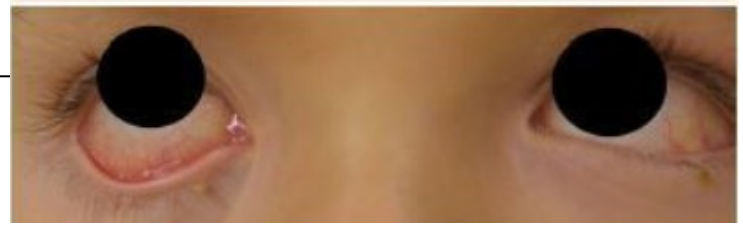
Case 4

- A 3-year-old boy
- Fever, Rash, icterus, and swollen and painful joints x5 days
- On examination lethargic
- Afebrile

What is first differential diagnosis?

Acute hepatitis?

On examination



- His conjunctivas were bilaterally hyperemic and icteric
- Distal and proximal interphalangeal joints in the lower and upper limbs and both knees were painful
- Developed fever after admission to hospital
- What will you do next?

Investigations

Hemoglobin 11.4g/dL

Platelet: 360000/ul

CRP: 105 mg/L

Bilirubin 5.4mg/dL

AST 149 U/L

ALT 150 U/L

Alkaline phosphatase 425 U/L

Albumin 3.1

Hospital course

- Ultrasound abdomen showed a normal gallbladder
- No intrahepatic or extrahepatic biliary ductal dilatation to suggest biliary obstruction
- Platelets 6.7 lacs ESR 113 mm/hr
- CRP 135mg/L
- Antistreptolysin O
- Hepatitis B surface antigen
- Hepatitis B core antibody
- Hepatitis immunoglobulin (Ig) M hepatitis C antibody,
- Cytolomegalo virus IgG/IgM,Toxoplasmosis IgG/IgM,
- Monospot, blood cultures, and leptospirosis testing were negative

Negative

Hospital course

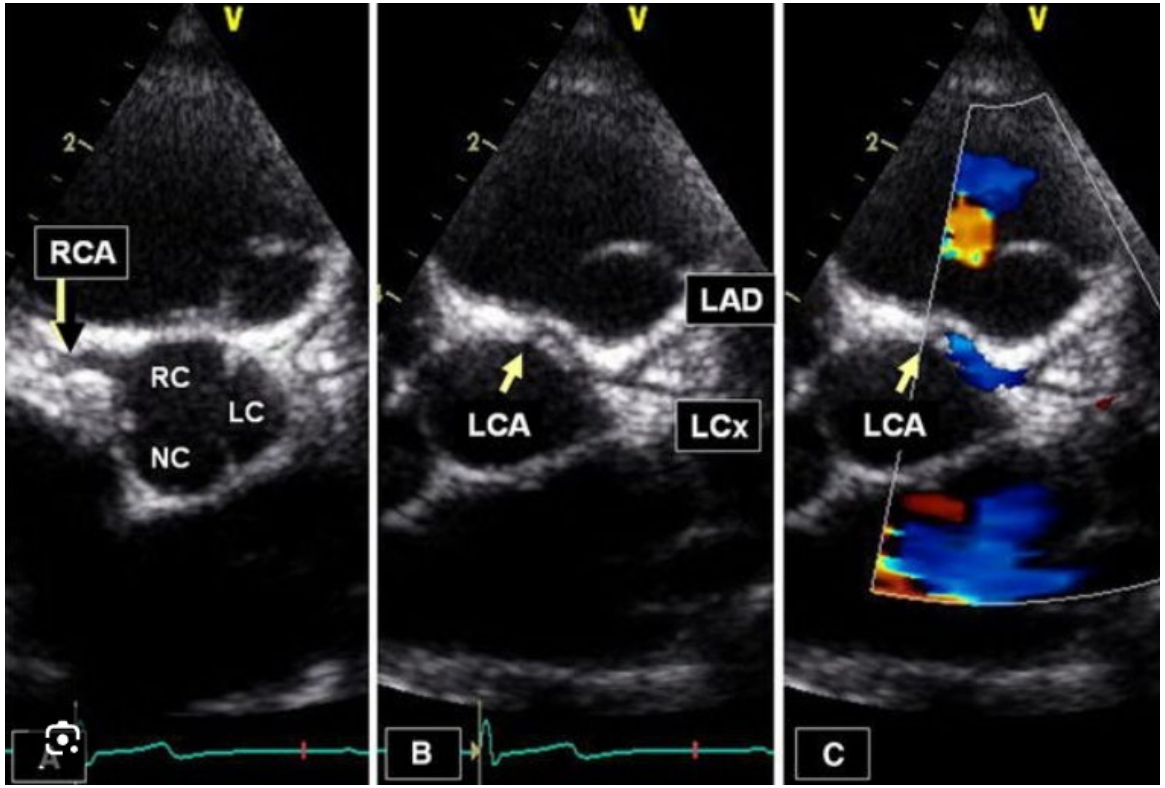
- On day seven of illness- B/L cervical lymphadenopathy
- He continued to spike fevers
- Poor response to antipyretics
- What would be next step?
- Repeat investigations

Elevated ALT, low albumin, thrombocytosis, and elevated WBC

What will you do next?

Diagnosed as atypical Kawasaki disease

Echocardiography



Normal

- IVIg infusion 2 gm/Kg
- Aspirin 50mg/Kg in divided doses
- Afebrile after 36 hours
- Continued on low doses aspirin
- Repeat 2D Echo normal

Key message

- Diagnosis incomplete KD was particularly challenging in absence of fever before hospitalization
- Unexplained jaundice with associated features should prompt physicians to consider KD
- High level of suspicion for KD when a child presents with febrile or afebrile obstructive jaundice

Case 5

- 11-year-old boy
- 4-day history of high fever
- Headache, generalized arthralgia, and rash, with nausea, vomiting, and diarrhea
- At the admission, he was febrile (40.4^{o}C)
- BP: 70/55 mmHg
- CFT – 5 Sec
- Clinical examination : bilateral non-purulent conjunctivitis, erythematous rash over the trunk and extremities

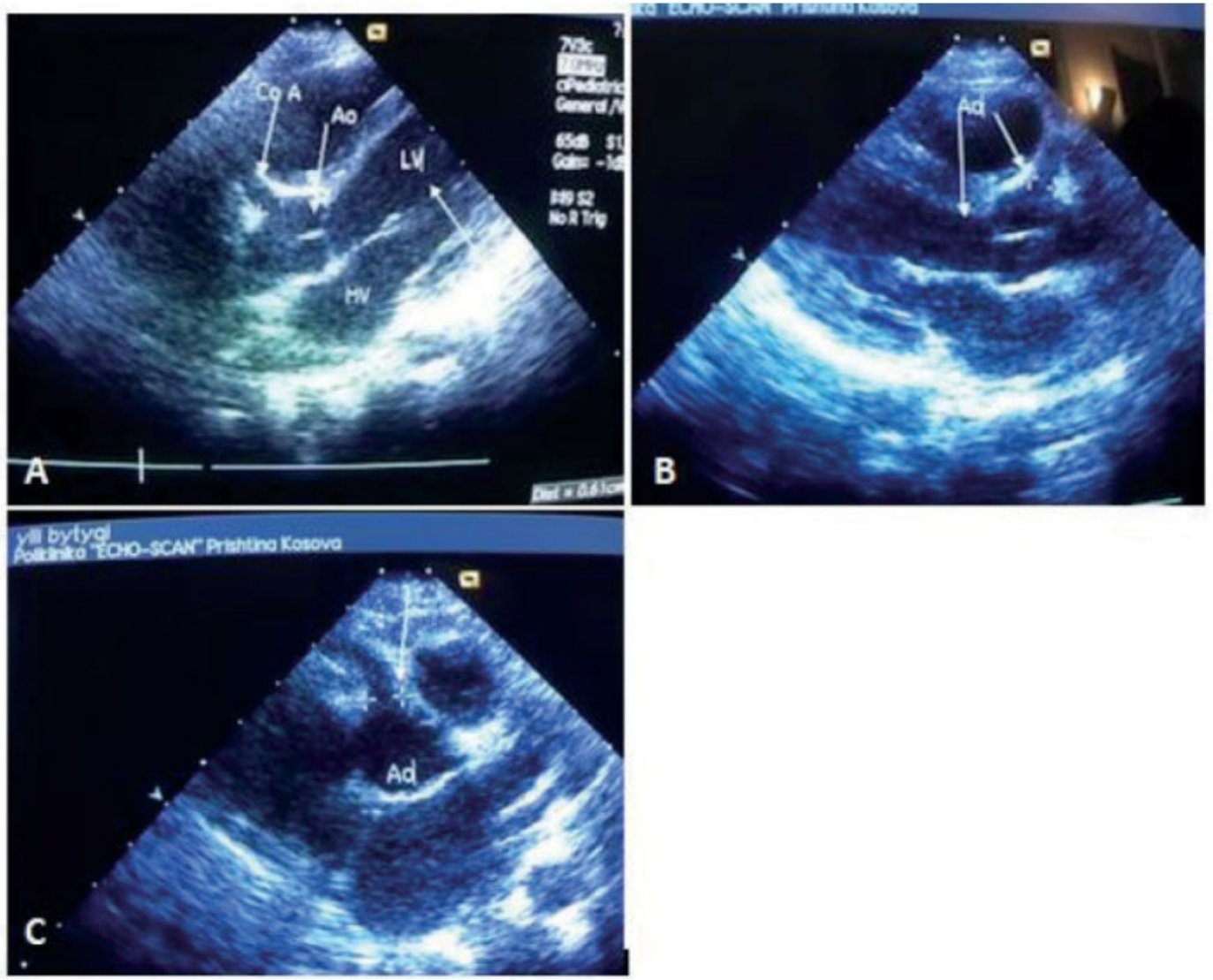
Laboratory investigations

- Hb – 10.g/dl
- TLC – 14,09X 10^9/L
- DC- N 72 L26 E 02 M 02
- Platelet count – 92,000
- Erythrocyte sedimentation rate of 122 mm/h
- CRP - 288 mg/L

- Na - 129 Meq/L
- K^+ - 3.6 Meq/L
- SGOT – 66 IU/L
- SGPT – 79 IU/L
- Serum Urea- 122 mg/dl
- Serum creatinine – 1.2mg/dl
- B-natriuretic peptide was 5560 ng/l (normal range < 100 ng/l)
- Blood culture - sterile

Hospital course

- His hypotension persisted despite adequate fluid resuscitation
- Inotropic support : Adrenalin maximum 0.4µg/kg/min
- ECG was normal
- 2 D Echocardiography
- Severe biventricular dysfunction
- Dilatation of right coronary artery



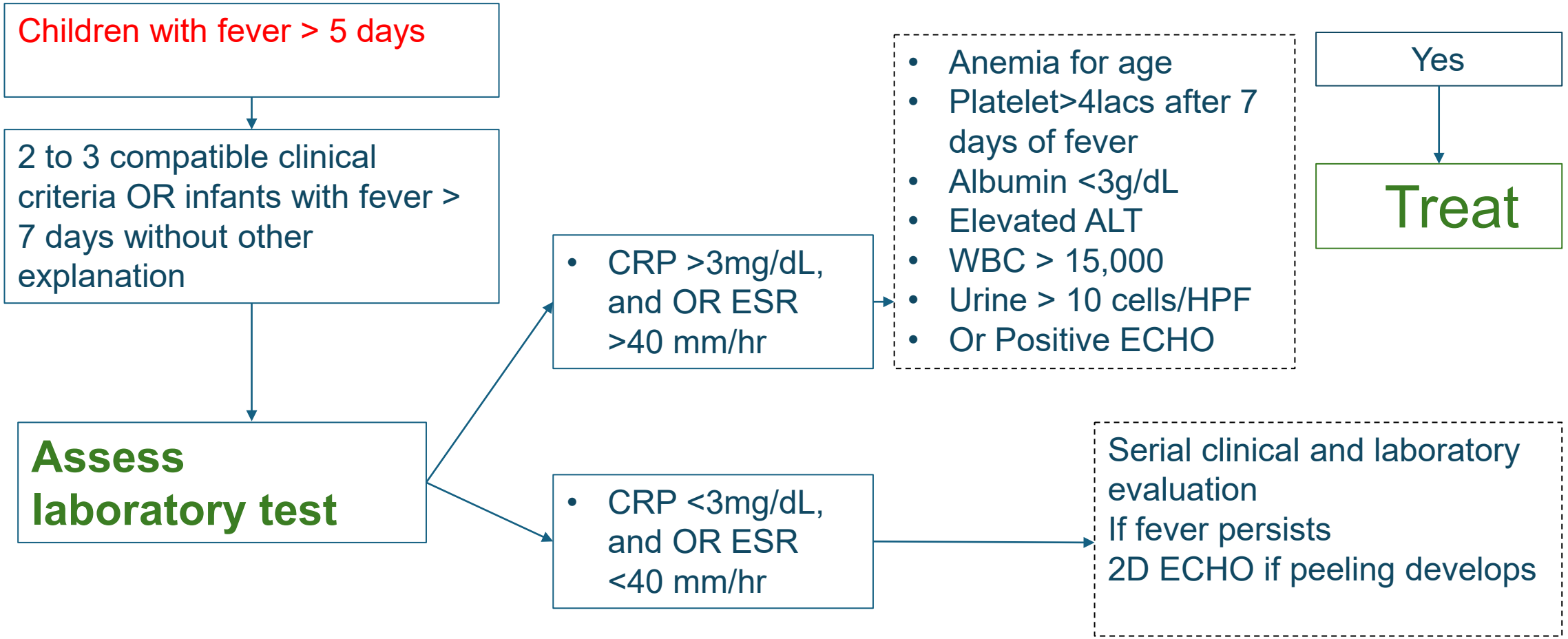
Treatment

- Treated with 2 g/kg IVIG
- Aspirin -50 mg/kg/d in four divided doses
- His general condition improved during the next 36 hours and subsequently became afebrile.
- After 2 weeks of admission a repeat transthoracic echocardiogram showed normal biventricular function
- Persistent RCA dilatation
- Discharged on low dose of aspirin

Key message

- Shock syndrome at times the manifestation of KD
- KDSS should be considered in all children with Hemodynamic instability , hypotension and myocardial dysfunction.
- Echocardiography should be performed frequently to look for coronary artery involvement and aggressive treatment is warranted

Atypical / Incomplete KD: Clinical Approach



RED FLAGS …

- Infants <6 months old with prolonged fever and irritability
- Infants with prolonged fever and unexplained aseptic meningitis
- Infants or children with prolonged fever and unexplained or culture-negative shock
- Infants or children with prolonged fever and cervical lymphadenitis unresponsive to antibiotic therapy
- Infants or children with prolonged fever and retropharyngeal or parapharyngeal phlegmon unresponsive to antibiotic therapy

Common Pitfalls in Diagnosis

- Infant <6 months of age, prolonged fever and irritability may be the only clinical manifestations of KD
- Delayed diagnosis is common in older children and adolescents with KD,

they appear to have a high prevalence of coronary artery abnormalities

- Presence of fever and pyuria in an infant or young child can be mistakenly attributed to a urinary tract infection
- Patients with cervical lymphadenitis as the primary clinical manifestation can be misdiagnosed as having bacterial adenitis
- KD shock may be misdiagnosed as having bacterial sepsis or staphylococcal or streptococcal toxic shock

A high index of suspicion required for the diagnosis