

Pediatric Rounds

# Miscellaneous cases

Iyad AL-Ammouri

Department of pediatrics, the University of Jordan

Please Use the Slide Show view to benefit from step wise animation of the case presentation and to have a chance to think in the case

# First Case

**A five year old previously healthy child presented to pediatric cardiology clinic because of incidental heart murmur heard by general practitioner.**

What are the important questions that you need to ask to differentiate between a pathological murmur and an innocent murmur?

# Approach to a heart murmur in children

## Murmur due to structural heart disease

Any abnormality in the cardiac structure resulting in turbulence of flow. Examples:

- Septal defect
- Abnormal valve (stenotic, regurgitant)
- Abnormal vessel (stenotic vessel)
- Dynamic obstruction (as HOCM)

## Functional murmur due to non-cardiac pathology

Any abnormality that results in increased flow across normal valves. Examples:

- Anemia
  - Fever
  - Sepsis
  - Hyperthyroidism
- (all are high output states)

## Innocent murmur with no pathology

Murmur caused by normal flow in certain situations, very common in children.

Examples:

- Still's murmur (musical vibratory sound)
- Pulmonary or aortic flow murmurs (increase in exercise or anxiety)
- Venous hum
- Subclavian/internal mammary souffle in pregnancy

So, we have to tailor the history towards ruling out cardiac pathology, and high output states.

### **Cardiac Pathology**

- Asking about symptoms of heart failure (exercise capacity, respiratory symptoms, Cyanotic heart disease symptoms)
- Asking about symptoms of HOCM (chest pain, syncope, palpitation)
- Family history of cardiomyopathies, sudden death, congenital malformations.. etc

### **High output states**

- Anemia (Pallor, nutritional history, growth, hemolysis, malabsorption, family history... etc.)
- Fever, sepsis, recurrent infections
- Hyperthyroidism symptoms

# The HPI Story

The child has no history of known cardiac disease, has normal exercise tolerance. Has no respiratory complaints, and never was noted to be cyanotic.

There was no history of palpitation, chest pain, syncope, or dizziness.

There is no family history of cardiac disease or sudden death.

There is no pallor, his diet is normal and balanced, no diarrhea, no jaundice, and no family history of anemias.

He had no fever at the time of examination by his doctor.

# How does the story help you in differential diagnosis?

The following is example of critical thinking

- **Structural heart disease?** Many structural heart disease produce symptoms early on during infancy and early childhood. Complex heart disease present in the neonatal period with either cyanosis or heart failure. However some MILD structural heart disease may not present with any symptoms. (examples: atrial septal defects, small ventricular septal defects, small ductus arteriosus, mild aortic or pulmonary valve stenosis)
- **Acquired heart disease and cardiomyopathy?** Acquired heart disease usually present with disease specific symptoms (endocarditis, myocarditic, cardiomyopathy). So it is unlikely to present as an incidental heart murmur. Keep in mind that HOCM may present with a heart murmur in an asymptomatic child, but it is still not a common disease at this age.
- **High output state?** Anemia is common in children, and sometimes it is asymptomatic if mild.
- **Innocent murmur?** These are very common in children, but the diagnosis should only be made by exclusion. So we cannot depend on the history only to diagnose innocent murmur. Physical exam is extremely important.

Next is physical exam

# What is the importance of the physical examination in this case?

Important points to do in the physical exam:

- 1) To rule out non-symptomatic structural heart disease
- 2) To look for evidence of high output states (such as anemia)
- 3) To describe the murmur to see if it is consistent with innocent murmur.

**Proceed for guidance**

# Physical examination findings

- Vital signs: RR 12, HR 92, BP 100/65 in right arm, Temp 37.2, Oxygen saturation 95%.
- Wt and height are normal on the 25<sup>th</sup> percentile
- Well appearing boy, not anxious, cooperative.
- No pallor, normal respiratory exam, normal abdominal exam with no hepatosplenomegaly, normal neurological exam
- Cardiac exam: normal pulses, normal perfusion, normal precordial palpation

**• Question: How do you describe a heart murmur?**

# Evaluation of a heart murmur

- There are four objective points in evaluation of a heart murmur that **must** be described:
  - Location (where is the murmur best heard)
  - Radiation if present (neck, back, or axilla)
  - Grade (i.e loudness) → (2/6: soft, 3/6 loud with no thrill, 4-6/6 loud with thrill)
  - Systolic, diastolic, continuous (systolic corresponds with pulse)
- Other features of the murmur can also be described, and may require some experience
  - Specific timing (early or late systolic, holosystolic, early or mid-diastolic)
  - Pitch (high pitch vs low pitch)
  - Shape (crescendo de-crescendo..)
  - Quality of sound (blowing, musical, harsh)
- Evaluate the presence of associated additional cardiac sounds (S3, S4, Clicks, snaps, Splitting)
- Maneuvers: murmurs can be manipulated through several maneuvers that can help in the diagnosis

• **Question: Describe maneuvers and their effect on physiology and murmurs.**

# Maneuvers for murmurs: Can be divided in two

## Anatomical

(maneuvers that make the pathology closer to your stethoscope)

Lean to the left

Lean forward

(think of how these maneuvers help you determine the lesion)

## Physiological

(maneuvers that manipulate flow, resistance, or preload status)

Supine, sitting, or standing

Inspiration vs expiration

Valsalva

Squatting

Sustained hand grip

(think of the effect of these maneuvers on flow, resistance or preload, think of situations in which these maneuvers help you determine the lesion)

# Rest of physical examination findings, four examples

Note: all examples may apply to our patient who is asymptomatic

- There is a soft 2/6 systolic murmur at the left upper sternal border, with no radiation. Murmur is midsystolic.
- The second heart sound is split, and it does not vary with inspiration or expiration.
- Rest of exam is normal

- There is a grade 4/6 systolic murmur at the apex, with no radiation. Holosystolic murmur.
- Thrill at the apex palpated
- Rest of the physical exam is normal

- There is a 3/6 systolic murmur at the right upper sternal border, radiates to the carotid artery. Murmur is midsystolic.
- There is ejection systolic click.
- Rest of exam is normal

- There is a soft 2/6 systolic murmur at the left sternal border, with no radiation. Murmur is musical, vibratory in nature
- The murmur becomes not audible in standing position
- Rest of physical exam in normal

**Diagnosis?**

**ASD**

**Diagnosis?**

**Small VSD**

**Diagnosis?**

**Bicuspid aortic valve with aortic stenosis**

**Diagnosis?**

**Innocent murmur**

# Second Case

A 10 year old previously healthy child presented with joint pain and swelling for the last 8 days. Initially this involved left ankle, was mild, and lasted for 3 days then resolved spontaneously, the parents thought it was a simple ankle sprain. But then his right knee became painful. The pain is severe causing limping, today he is not able to bear weight (progressive). His knee is noted to be swollen and red in color. He had no similar episodes in the past.

There is also associated fever of 38.2 axillary for the last 4 days, without chills or rigors.

There was no current history of sore throat, or signs of viral upper respiratory infection. But the mother recalls having sore throat and fever 2 months ago, for which he received paracetamol, and amoxicillin for 2 days then he stopped it because he was better.

There has been no trauma, no insect bites. No skin rash.

No history of abdominal pain, diarrhea or blood in stool. No history of hematuria or change in urine color. No history of chest pain or fatigue. No history of headache, blurred vision, or weakness.

No family history of joint problems. Past medical and surgical history is not remarkable. Nutritional, developmental, vaccination, and social histories were all not remarkable.

**Think of the scenario and analyze it critically in view of differential diagnosis of arthritis.**

## The following is example of critical thinking

- **In a child with joint pain and swelling, I think of common causes including but not limited to:**
  - Infectious process such as septic arthritis
  - Rheumatic fever
  - Reactive arthritis due to viral infection
  - Severe trauma either causing joint pain and swelling in a normal patient, or mild trauma causing hemarthrosis in patients with coagulopathy
  - Rheumatoid arthritis and other connective tissue disorder
  - Arthritis in other disorders, malignancy, inflammatory bowel disease, Henoch- schönlein purpura.
- **In this story, the following are quick considerations:**
  - Septic arthritis is less likely due to more than one joint involvement
  - Reactive arthritis is a possibility, usually they are due to concomitant viral infection
  - Trauma is ruled out by history, hemarthrosis usually occur with previous history of recurrent manifestation of bleeding disorder, and with positive family history of hemophilia
  - Connective tissue disorders are not diagnosed with acute presentations. In addition, there are usually associated with extra-articular manifestations.
  - HSP is a disease in younger children and has to have skin rash in the lower extremities.
  - Rheumatic fever is a strong possibility based on the story, particularly with the past history of pharyngitis.

**How do you make the diagnosis of Rheumatic fever?**

# Modified Jones Criteria (Major)

- **Arthritis**: very painful, migratory, involving large joints, non-destructive
- **Carditis**: presents as congestive heart failure, new murmur, friction rub of pericarditis
- **Subcutaneous nodule**: non-tender nodules on the extensor surfaces of joints (very infrequent)
- **Erythema marginatum**: Long lasting skin rash on the trunk or limbs
- **Chorea**: can be very late in the disease

# Minor Criteria

- Fever
- Arthralgia (in the absence of arthritis)
- Lab: Elevated ESR or CRP or WBC count
- Prolonged PR interval
- Previous rheumatic fever

For the diagnosis, there must be 2 major, or 1 major and 2 minor criteria

**Plus evidence of group A strept. Infection  
(either positive throat culture or positive antibody titer-  
antistreptolysin ASO)**

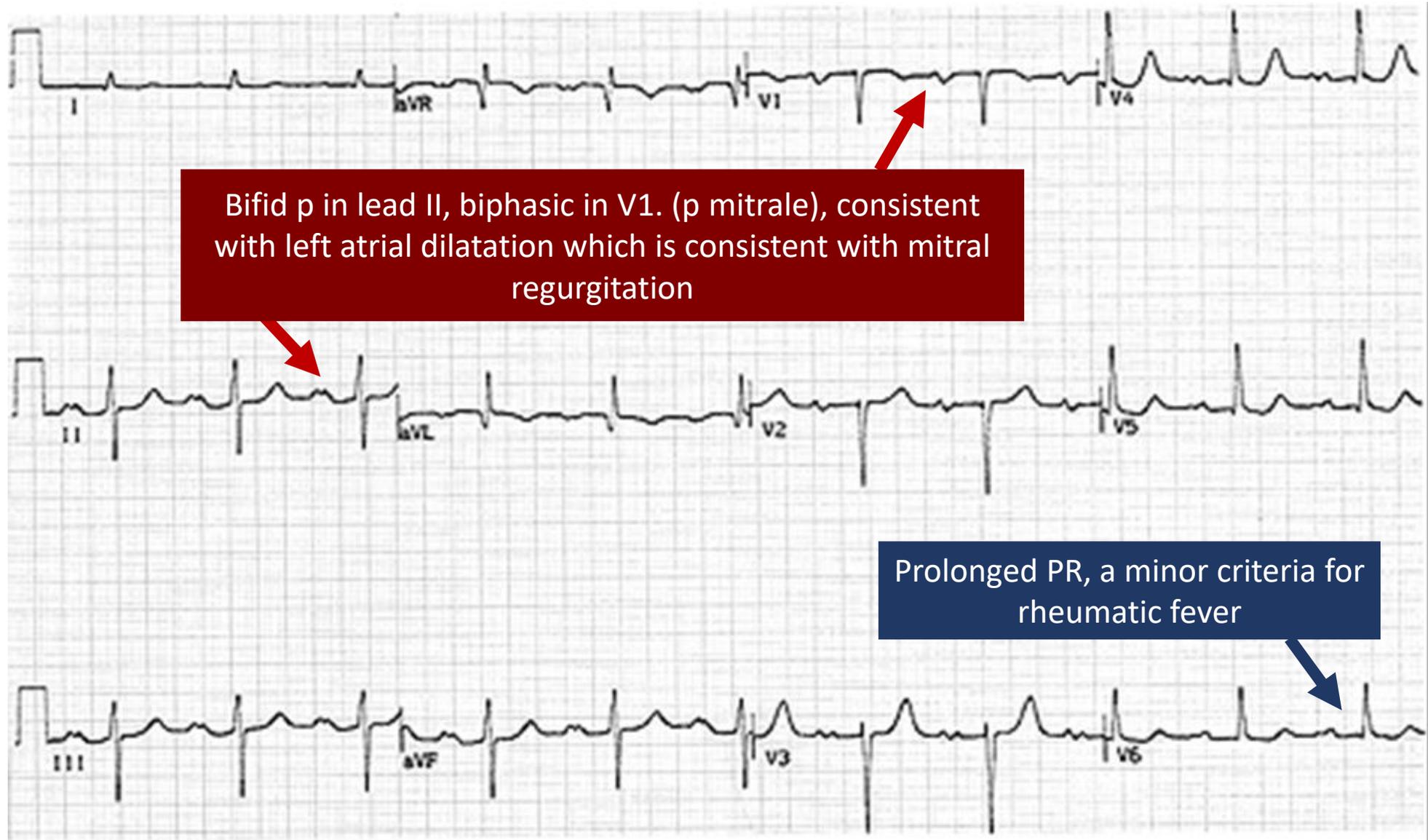
# Physical examination findings

- Vital signs: RR 14, HR 90, BP 120/65 in right arm, Temp 37.5, Oxygen saturation 95%.
- Wt and height are normal on the 25<sup>th</sup> percentile
- Well appearing boy, not anxious, cooperative, but appears in pain.
- The right knee is swollen, hot and tender.
- No skin rash, no subcutaneous nodules seen, no abnormal movements
- Cardiac exam: normal pulses, normal perfusion, normal precordial palpation. There is an apical holosystolic murmur at the apex with radiation to the axilla.

• **Question: What investigations you would ask for?**

**Examples of important testing: ECG, CBC, ESR, ASO titer, Echocardiography**

What are the two findings in this ECG, and how do you explain it in view of Rheumatic fever?



# • What is carditis in acute rheumatic fever?

- Inflammation can involve all cardiac layers, PANCARDITIS (endocardium, myocardium, and pericardium)
- Most likely affected valves: Mitral valve followed by aortic valve (usually mitral and aortic regurgitation)
- Mitral valve and aortic regurgitation may improve after treatment with anti-inflammatory medications
- With time mitral stenosis can develop many years after the acute rheumatic fever due to fibrosis. Follow up should be done on regular basis



**Exercise: Think of the findings on physical exam in a patient with mitral valve stenosis..**

# Cause and Pathophysiology of rheumatic fever

- Group A Beta hemolytic streptococcal pharyngitis
  - ARF follows untreated pharyngitis by 2-6 weeks
  - Most common age group 5-15 yrs
  - No gender differences except for Sydenham's chorea → more females
- Antibody cross-reactivity (molecular mimicry)
  - Cell wall of group A strept. Contains a highly antigenic protein (M protein)
  - Anti- M proteins may cross react with cardiac and smooth muscle myosin inducing cytokine release and tissue destruction

# Treatment

- Treat infection if active pharyngitis
- Anti-inflammatory ( Aspirin (ASA), consider steroids if intolerant to aspirin)
- Treat heart failure if present
- For chorea, the use of valproic acid or haloperidol is usually effective

# Prophylaxis

- Patients who had rheumatic fever should receive prophylaxis long acting penicillin injection every 3-4 weeks
  - If there is cardiac involvement → prophylaxis should be given well into adulthood
  - If there is no cardiac involvement → prophylaxis should be given for 5 years or till the age of 18 years

END