



# 41<sup>st</sup> National Conference on Pediatric Health Care

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## RHEUMATOLOGY & IMMUNOLOGY

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# Disclosures

Albrey Berber DNP, CPNP-PC

- Has no financial relationship with commercial interests
- This presentation contains no reference to unlabeled/unapproved uses of drugs or products

# Learning Objectives

Upon completion of this review, the course attendee should be able to:

- Describe the process of history taking and physical assessment of Rheumatology/Immunology concerns
- Summarize common diagnostic tests (laboratory and radiology) utilized when evaluating a Rheumatology/Immunology concern
- Compare and contrast the pathophysiology, clinical presentation, management, and follow-up of the most common Rheumatology/Immunology diagnoses seen in primary care
- Describe education needs related to the most common Rheumatology/Immunology diagnoses



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## Rheumatologic Disorders

# Juvenile Idiopathic Arthritis (JIA)

- **Key Characteristics:**

- Encompasses several disorders that have a common feature of arthritis
- Diagnosis requires a persistent arthritis for more than 6 weeks in a patient < 16
- Onset type defined by type of disease in first 6 month:
  - Polyarthritis: >5 inflamed joints
  - Oligoarthritis : <5 inflamed joints
  - Systemic-onset: arthritis with characteristic fever
- Higher incidence in girls

- **S/S:**

- **Key physical findings:**

- Pain (mild-mod) aching
- Joint stiffness (worse in the morning & after rest)
- Swelling of the joint w/effusion
- Heat over inflamed joint
- Loss of ROM of affected joint(s) –may keep joint slight flexed, limp

- **Systematic manifestations:**

- Fever
- Salmon-colored rashes
- Leukocytosis
- Lymphadenopathy
- Rheumatoid nodules

# Juvenile Idiopathic Arthritis (JIA)

## • Evaluation:

- Diagnosis of exclusion
- No diagnostic lab tests for JIA
- MRI can help in managing joint pathologies
- Useful lab tests include:
  - CBC (to exclude leukemia)
  - ESR, CRP
  - Lyme titers
  - LFTs
  - ANA (if + needs ophthalmology referral)

## • Management:

- Pediatric Rheumatologist
- NSAIDs
- Corticosteroids
- Antirheumatic drugs (Methotrexate)
- Physical Therapy



# Polyarticular JIA

- **Key Characteristics & S/S:**

- About 30% of patients affected
- **5 joints or more, symmetrical**
- F > M
- Two age groups:
  - **Young onset 1-2 y: most RF-**
  - **Teens 12-16y: RF+ (12%)**
- Uveitis is rare. Fever & rash at onset are possible.
- May have subcutaneous Rheumatoid Nodules

- **Evaluation & Management:**

- Prognosis: potential for more chronic and destructive disease
- Patients can be anemic due to chronic inflammation. ESR, CRP & platelet count are elevated when disease is active
- **Treatment:** First line agents include NSAIDs and DMARDs such as Methotrexate

# Oligoarticular JIA

- **Key Characteristics & S/S:**

Most common of JIA: 60%

- **4 or less joints**
- Peak age of onset 1-2 years, F > M
- Most patients are ANA+ (65-85%)
- Joint disease has a good prognosis

- **Evaluation & Management:**

- **Uveitis in 5-15%**, initially silent but tends to become chronic, w/poor prognosis: **Must have a Slit Lamp eye exam every 3 months!**
- **Treatment:** Initial approach as been NSAIDs; Naproxen has been shown effective at a dosage of 15 to 20mg/kg given w/food twice daily
- Methotrexate is considered the 2<sup>nd</sup> line for patients w/more chronic arthritis. Dosing is once weekly & can be given orally or by subcutaneous injection



# Systemic JIA

- 10% of patients, F: M 1:1
- Onset at any time before age 16
- **Fever, rash and arthritis** (any number of joints)
- **Inflammation in & around internal organs:** heart (carditis/ pericarditis), lungs (pneumonitis/ pleuritis), gut, brain
- **Swollen lymph nodes, enlarged liver & spleen**
- **Anemia of chronic inflammation, elevated inflammatory lab markers: ESR, CRP, ferritin & WBC**
- **Evidence of Macrophage Activation Syndrome: prolonged clotting time, low fibrinogen & elevated fibrin split products (D –Dimers)**

# Uveitis

- **Key Characteristics:**

- Inflammation of the anterior, intermediate, or posterior chamber of the eye
- It is predominately an insidious onset, anterior, nongranulomatous inflammation affecting the iris & ciliary body

- **S/S:**

- Ocular pain & redness
- Change in vision
- Photophobia
- Headache
- Asymptomatic

- **Diseases associated w/uveitis:**

- Rheumatic Diseases
  - JIA
  - Reactive Arthritis
- Vasculitis
  - Kawasaki disease
  - Behcet's
  - HSP (rare)
  - Wegener's Granulomatosis
- Other diseases
  - Pars planitis
  - HIV, EBV
  - Cat scratch
  - Herpes
  - Lyme disease

- **Management:**

- Refer to ophthalmologist to manage
- 1<sup>st</sup> line (topical corticosteroids w/mydriatics)

# Systemic Lupus Erythematosus (SLE)

- **Key Characteristics:**

- Chronic, systemic disease characterized by altered immune regulation that can involve inflammation in multi-organ systems
- Childhood onset is rare; however, it is more acute & severe in children than adults
- Higher incidence in Asians, AA, Hispanics & Native Americans
- Median age onset 11-12 years of age
- Females > Males
- Hallmark is butterfly or malar erythematous facial rash which increases in intensity in sunlight

# Systemic Lupus Erythematosus (SLE)

- **History may include the following:**
  - Joint pain (most common)
  - Low grade fevers (sustained or intermittent)
  - Weight loss
  - Painless mouth ulcerations
  - Skin rashes
  - Sun sensitivity
  - Fatigue
  - Renal disorders (proteinuria)
  - Hematologic disorders
- **Physical exam:**
  - Pallor
  - Petechiae
  - Purpura
  - Malar (Butterfly rash)
  - Mouth ulcerations
  - Gingivitis
  - Joint tenderness
  - Serositis
  - Cardiac friction rub (pericarditis)
  - Pleural friction rub (pleuritic)
  - Hepatosplenomegaly
  - Lymphadenopathy

# Systemic Lupus Erythematosus (SLE)

## • Evaluation:

- CBC
- ANA (is + in more than 97% of children who have active untreated SLE, and a – ANA excludes SLE)
- ESR
- CRP
- UA

## • Management:

- Rheumatology referral
- Sunscreen
- NSAIDs, oral steroids, antimalarial drugs for cutaneous & MSK manifestations
- Vitamin D & calcium supplement to reduce osteoporosis risk related to corticosteroid use

# Fibromyalgia Syndrome

- **Key Characteristics:**

- Chronic, idiopathic pain syndrome
- Age of presentation is typically during the adolescent years with mean onset of 12
- Females > Males
- Pathophysiology is unclear but there is evidence that the CNS has increase pain/sensory feelings

- **S/S:**

- Pain at multiple sites, including muscles & soft tissues around joints
- Pain may awaken from sleep & interfere with w/activities
- Fatigue & malaise
- Paresthesia & headache
- Insomnia or prolonged night wakening
- Depression
- Anxiety
- School absence due to pain

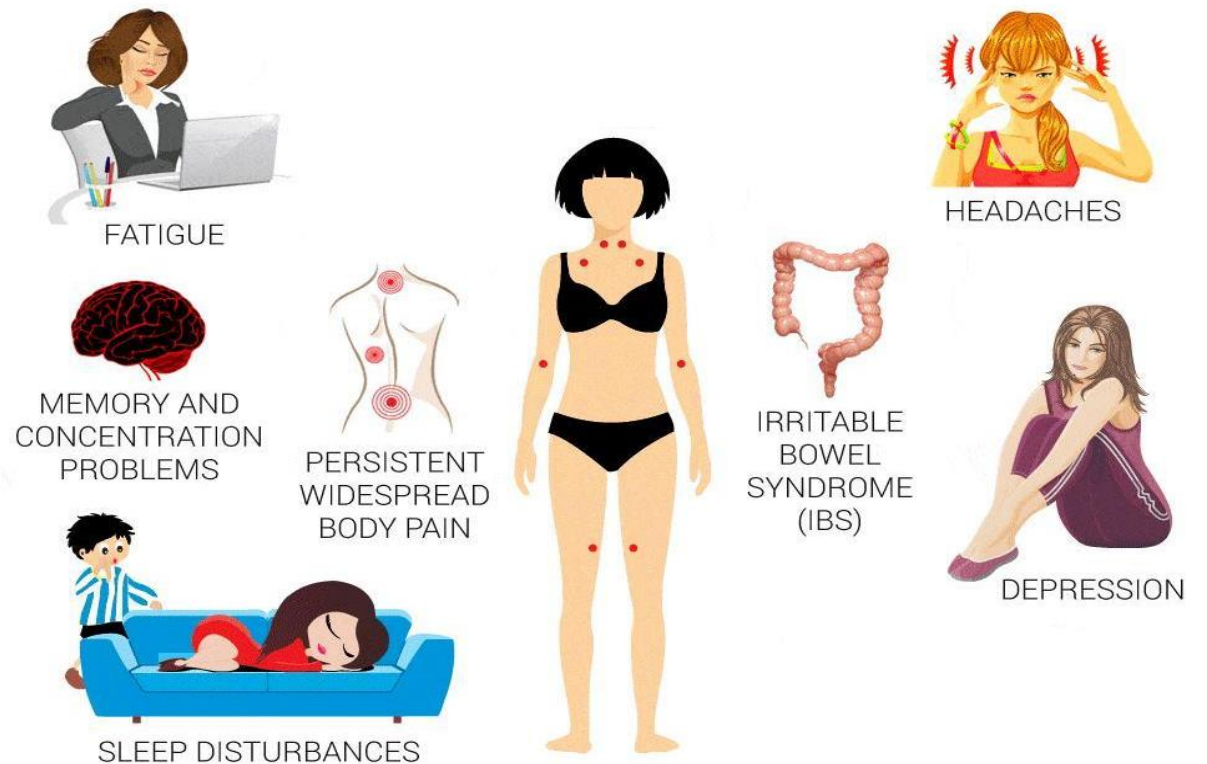
# Fibromyalgia Syndrome

## • Evaluation:

- Pain in at least 3 areas for more than 3 months (w/normal labs)
- More than 5 of 18 tender points
- 3 of 10 major criteria:
  - Fatigue
  - Poor sleep
  - IBS
  - Chronic tension or anxiety
  - Soft tissue swelling
  - Pain affect by weather
  - Paresthesia
  - Pain affected by activity
  - Headache
  - Pain affected by anxiety and/or stress

## • Management:

- Physical therapy
- Psychotherapy & relaxation
- NSAIDs
- Gabapentin to reduce pain sensitivity





# Acute Rheumatic Fever

Acute rheumatic fever is a non-suppurative sequela that occurs 2-4 weeks following Group A *Streptococcus* pharyngitis & may consist of arthritis, carditis, chorea, erythema marginatum, & subcutaneous nodules

Acute febrile illness	Neurologic illness (25-30%)
<ul style="list-style-type: none"><li>• Onset two to four weeks after GAS infection</li><li>• Fever is common</li><li>• Acute joint signs/symptoms</li><li>• Carditis<ul style="list-style-type: none"><li>• Clinical &amp; subclinical</li></ul></li><li>• Skin manifestations &amp; subcutaneous nodules (both are rare)</li><li>• Raised inflammatory markers (ESR)</li><li>• Evidence of preceding GAS infection (elevated ASO &amp; anti-DNase B titers)</li><li>• Dramatic symptomatic response to aspirin &amp; NSAIDS</li><li>• Duration usually &lt;6 weeks</li><li>• Followed by RHD in approximately 75%</li></ul>	<ul style="list-style-type: none"><li>• Later onset<ul style="list-style-type: none"><li>• 2-6 months after GAS infection</li></ul></li><li>• No fever</li><li>• Joint manifestations are not a feature</li><li>• Behavioral disorder &amp; distinctive chorea</li><li>• Carditis &gt;30%<ul style="list-style-type: none"><li>• Often subclinical</li></ul></li><li>• Often normal inflammatory markers</li><li>• ASO often unhelpful</li></ul>

# Acute Rheumatic Fever: Prevention & Treatment

Prevention of RF is accomplished by prompt diagnosis & treatment of GAS

However if ARF develops the treatment includes the following:

- Eradication of group A beta-hemolytic *Streptococcus* (GAS)
- Symptomatic relief of acute disease manifestations (arthritis)
- Chest X-ray, ECG, & Echo are indicated: carditis usually develops w/in the first 3 weeks of symptoms (Referral if indicated)
- Education for the patient and patient's caregivers

# Severe Combined Immunodeficiency (SCID)

- **Key Characteristics:**

- Genetic disorder in which both “arms (B and T cells) of the adaptive immune system are impaired
- Also known as “bubble boy disease” because of the extreme vulnerability to infectious diseases
- Risk for life threatening infection. From the 1<sup>st</sup> months of life they have infection that may be frequent, severe, long-lasting or hard to treat
- Infections in lungs (pneumonia) around the brain or spinal cord (meningitis) or the blood stream
- There is a delay in detection because newborns carry mother’s antibodies for the few weeks of life SCID babies look normal/healthy

- **S/S:**

- If a baby exhibits any of the following persistent symptoms w/in the 1<sup>st</sup> year of life, evaluation for SCID or other types of immune deficiency syndromes is indicated:
  - 8 or more ear infections
  - 2 or more cases of pneumonia
  - Infections that do not resolve w/antibiotic treatment for 2 + months
  - Failure to gain weight or grow normally
  - Infections that require IV antibiotic treatment
  - Deep-seated infections, such as pneumonia that affects an entire lung or an abscess in the liver
  - Persistent thrush
  - A family history of immune deficiency or infant deaths due to infections

# Severe Combined Immunodeficiency (SCID)

- **Evaluation:**

- The age average age at which babies are diagnosed is just over 6 months, usually because of recurrent infections & FTT
- Blood tests typically reveal significantly lower-than-normal levels of T cells & a lack of germ-fighting antibodies
- Even if B cells are present, they do a poor job of producing antibodies
- Low antibody levels & lack of specific antibodies after vaccination or a natural infection are characteristic features

- **Management:**

- Prevent infections
- Enzyme therapy
- Gene therapy
- Most common therapy is bone marrow transplant (matched sibling is the most successful)

# Question

An 8-year-old child is diagnosed with systemic lupus erythematosus (SLE), and the child's parent asks if there is a cure. What will the primary care pediatric nurse practitioner tell the parent?

1. Complete remission occurs in some children at the age of puberty.
2. Periods of remission may occur but there is no permanent cure.
3. SLE can be cured with effective medication and treatment.
4. The disease is always progressive with no cure and no remissions.

An 8-year-old child is diagnosed with systemic lupus erythematosus (SLE), and the child's parent asks if there is a cure. What will the primary care pediatric nurse practitioner tell the parent?

Answer: Periods of remission may occur but there is no permanent cure.

# Question

The primary care pediatric nurse practitioner examines a child who has had stiffness and warmth in the right knee and left ankle for 7 or 8 months but no back pain. The nurse practitioner will refer the child to a rheumatology specialist to evaluate for

1. Enthesitis-related JIA.
2. Oligoarticular JIA.
3. Polyarticular JIA.
4. Systemic JIA.



The primary care pediatric nurse practitioner examines a child who has had stiffness and warmth in the right knee and left ankle for 7 or 8 months but no back pain. The nurse practitioner will refer the child to a rheumatology specialist to evaluate for

Answer: Oligoarticular JIA.

# Question

An 8-year-old boy has a recent history of an upper respiratory infection and comes to the clinic with a maculopapular rash on his lower extremities and swelling and tenderness in both ankles. The pediatric nurse practitioner performs a UA, which shows proteinuria and hematuria and diagnoses HSP. What ongoing evaluation will the nurse practitioner perform during the course of this disease?

1. ANA titers
2. Blood pressure measurement
3. Chest radiographs
4. Liver function studies

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Answer: Blood pressure measurement

# Question

The primary care pediatric nurse practitioner is reviewing the rheumatology plan of care for a child who is diagnosed with SLE. Besides reinforcing information about prescribed medications, what will the nurse practitioner teach the family to help minimize flaring of episodes?

1. Have the child rest between activities.
2. Obtain regular ophthalmology exams.
3. Participate in low-impact exercises.
4. Use UVA and UVB sunscreen daily.

The primary care pediatric nurse practitioner is reviewing the rheumatology plan of care for a child who is diagnosed with SLE. Besides reinforcing information about prescribed medications, what will the nurse practitioner teach the family to help minimize flaring of episodes?

Answer: Use UVA and UVB sunscreen daily.

# Question

An adolescent female reports poor sleep, fatigue, muscle and joint pain, and anxiety lasting for several months. The primary care pediatric nurse practitioner notes point tenderness at several sites. What will the nurse practitioner do next?

1. Evaluate the adolescent's pain using a numeric pain scale.
2. Obtain ANA, CBC, liver function, and muscle enzymes tests.
3. Refer the adolescent to a rheumatologist for further evaluation.
4. Reassure the adolescent that this condition is not life-threatening.

An adolescent female reports poor sleep, fatigue, muscle and joint pain, and anxiety lasting for several months. The primary care pediatric nurse practitioner notes point tenderness at several sites. What will the nurse practitioner do next?

Answer: Refer the adolescent to a rheumatologist for further evaluation