1 **RHEUMATOLOGY**

1 Yonit Sterba, MD
2 Attending, Pediatric Rheumatology
Nicklaus Children's Pediatric Specialists
Nicklaus Children's Hospital
Miami, Florida

2 **Disclosure of Relevant Relationship**

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Topics You Need to Know

1) Systemic Lupus Erythematosus*
2) Juvenile Idiopathic Arthritis*
3) Vasculitis Syndromes (focus on Kawasaki and HSP)*
4) Dermatomyositis
5) Scleroderma
6) Ankylosing Spondylitis
7) Reactive Arthritis and Post-infectious arthritis
8) Sarcoidosis
9) Hypermobility Syndrome
10)Functional Joint Complaints
11)Marfan's
12)Ehlers-Danlos

4 **Question 1**

A 16-year-old girl comes to your office for a follow-up visit from the emergency department, where she went for the acute onset of knee pain and swelling. The emergency department physician had ordered an antinuclear antibody test, which was positive at 1:320. Further history reveals that she has had intermittent joint pains for several weeks and dark-colored urine. Findings on her physical examination are normal except for an effusion in her right knee. You decide that further evaluation for systemic lupus erythematosus (SLE) is warranted.
Of the following, the MOST specific test in helping you make the diagnosis of SLE is:

a) Lupus anticoagulant
b) Anti double stranded DNA antibody
c) Anti-Ro measurement
d) Complement measurement
e) VDRL (venereal disease research laboratory)

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A 14-year-old girl presents with a 2-month history of joint pain that is responding poorly to over-the-counter anti-inflammatory medications. She reports some sores in her mouth and mild swelling around her eyes and ankles. On physical examination, her temperature is 37.0°C, heart rate is 76 beats/min, respiratory rate is 14 breaths/min, and blood pressure is 130/86 mm Hg. She has oral ulcers, mild periorbital and pretibial edema, and mild swelling of her wrists and knee joints. Laboratory findings include:

ulcers, mild periorbital and pretibial edema, and mild swelling of her wrists and knee joints. Laboratory findings include:

- Sodium, 136 mEq/L (136 mmol/L)
- Potassium, 4.8 mEq/L (4.8 mmol/L)
- Chloride, 100 mEq/L (100 mmol/L)
- Bicarbonate, 22 mEq/L (22 mmol/L)
- Blood urea nitrogen, 24.0 mg/dL (8.6 mmol/L)
- Creatinine, 1.3 mg/dL (114.9 mcmol/L)
- Albumin, 2.5 g/dL (25.0 g/L)
- Hemoglobin, 10.1 g/dL (101.0 g/L)
- White blood cell count, 3.0x103/mcL (3.0x109/L)
- Platelet count, 190x103/mcL (190x109/L)
- Urinalysis: 3+ blood, 3+ protein, with 20 to 50 red blood cells/high-power field
- Antinuclear antibody titer: 1:1,280
- Anti-double-stranded DNA titer: 1:640

Of the following, the next BEST step in management is to:

A. Admit the patient for intravenous cyclophosphamide treatment
B. Initiate treatment with ibuprofen
C. order a 24-hour urine for protein collection
D. refer the patient for a renal biopsy
E refer the patient for bone marrow aspiration

6 **Systemic Lupus Erythematosus**

7 **SYSTEMIC LUPUS ERYTHEMATOSUS**

Clinical Manifestations

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- Oral ulcers
- Immunologic disorder
  - (dsDNA, Anti-Sm, Antiphospholipid Ab)
- Neurologic symptoms
- Malar rash
- Discoid rash

2 ACR Criteria (4/11 to meet criteria)
"A RASH POINTs MD"

- Arthritis
- Renal disease
- ANA positive
- Serositis
- Hematologic disorder
- Photosensitivity

8

- Hematologic disorder
- Photosensitivity

8 **SYSTEMIC LUPUS ERYTHEMATOSUS**

Clinical Manifestations: Updated Criteria 2019

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10 **SYSTEMIC LUPUS ERYTHEMATOSUS**

Antinuclear Antibody (ANA) and SLE

- ANA test is almost 100% SENSITIVE, but NOT SPECIFIC
- Approximately 20-30% of healthy children can have positive ANA
- ANA's occur in other autoimmune conditions as well (JIA, scleroderma, Sjogren's, autoimmune thyroid disease)
- Anti dsDNA and Anti Sm tests are SPECIFIC for SLE
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11 **SYSTEMIC LUPUS ERYTHEMATOSUS**

Course and Complications

- Wide spectrum of severity based on organs involved
- Lupus nephritis is a common complication
  - Spectrum of severity
  - Multiple Classes of lupus nephritis (I-VI)
- Renal and Neurologic involvement tend to lead to the most morbidity
- Immunosuppression occurs from disease and medications
- Following anti-dsDNA and complement levels is helpful in disease management
  - C3 and C4 levels drop in active disease
- Treatment (immunosuppression) is based on systems involved and severity of disease
  - Hydroxychloroquine for essentially all patients
  - Prednisone +/- additional immunosuppressive meds based on severity
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12 **NEONATAL LUPUS**

Maternal Anti-Ro (SSA), Anti- La (SSB)

Cross placenta (IgG)

Lead to inflammation/scarring

Clinical Manifestations:
Rash
Thrombocytopenia
Heart Block

Heart Block

Treatment:
Rash and Thrombocytopenia self resolve
Cardiac pacing for complete heart block

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You treat a 15-year-old girl in your practice who has juvenile idiopathic arthritis (JIA). She is brought in by her mother today with complaints of a low-grade fever and diffuse pain. On physical examination, she has a temperature of 38.0°C and a heart rate of 100 beats/minute. As she sits on the examination table, she leans forward. During auscultation of her lungs, she complains of pain with deep inspiration.

Of the following, the MOST likely explanation for her symptoms is:

a) Costochondritis
b) Gastritis
c) Pericarditis
d) Pneumonia
e) Pulmonary Embolism

14 **Juvenile Idiopathic Arthritis**

15 **Juvenile Idiopathic Arthritis**

Definition
- Arthritis of UNKNOWN ETIOLOGY
- Lasting 6 weeks or more
- Occurring in child <16
- Other causes investigated and ruled out
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16 **Juvenile Idiopathic Arthritis**

17 **Juvenile Idiopathic Arthritis**

18 **Other important causes of arthritis**

Lyme Arthritis
- ALWAYS on the differential of monoarthritis
- Most commonly affects the knee
- Late manifestation of lyme disease (think months later)
- Most likely will never have noticed any tick bite or ECM rash
- Understand early vs. late manifestations

Diagnosis based on serology with confirmatory western blot

Treatment: 4 weeks antibiotics (Doxy or Amox based on age)

19

20

Treatment: 4 weeks antibiotics (Doxy or Amox based on age)

19 **Other important causes of arthritis**

Acute Rheumatic Fever vs. Post Strep Reactive Arthritis

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- An 18-month-old boy is brought to the clinic with fever and irritability. His mother explains that he has had a fever for the past week and a red rash on his extremities. On physical examination, he has a temperature of 39.2°C; he is irritable; his eyes are injected without discharge (Item Q197A); and his lips are dry, red, and cracked (Item Q197B). All other findings are within normal limits.

  Of the following, the MOST appropriate next step in this patient's care is to

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A) administer intravenous antibiotics
B) administer intravenous gamma globulin
C) obtain blood cultures
D) obtain electrocardiography
E) perform a lumbar puncture and culture the cerebrospinal fluid

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21 **Vasculitis**

22 **Vasculitis**

23 **Kawasaki Disease**

Clinical Manifestations:
Most commonly young child (toddler age)
Must have at least 5 days of fever + 4/5:

- Cervical adenopathy (≥1.5cm)
- Mucous membrane changes (dry, cracked lips or "strawberry tongue")
- Conjunctivitis ("limbic sparing")
- Rash (basically any type)
- Extremity changes (edema, desquamation)

Lab findings:

- sterile pyuria
- Elevated acute phase reactants (can remain high for weeks)
- Thrombocytosis
- Leukocytsosis

24 **Kawasaki Disease**

25 **Kawasaki Disease**

Treatment:

Goals: avoid cardiac complications and calm ongoing inflammation
Don't need definitive diagnosis to start treatment
Ideally IVIG + Aspirin before within first 10 days of illness

Don't need definitive diagnosis to start treatment
Ideally IVIG + Aspirin before within first 10 days of illness

- IVIG: 2mg/kg (can give repeat dose)
- Aspirin 80-100mg/kg/day for 24-48 hours
- Low dose aspirin (3-5mg/kg/day until ESR&CRP normalize)

Lifetime aspirin if coronary aneurysm present

ECHO: Baseline
Follow up at 2-3 weeks
Follow up at 6-8 weeks

26 **Henoch-Schonlein Pupura (HSP)**

27 **Henoch-Schonlein Pupura (HSP)**

Diagnosis

- Usually clinical
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- Skin biopsy
  - usually reserved for atypical presentation
  - IgA deposition in post-capillary venules
  - Pathology described as "leukocytoclastic vasculitis"

28 **Dermatomyositis**

29 **Dermatomyosits**

Clinical Manifestations:

- Typically school age child
- Skin findings:
  - Heliotrope rash "violacious rash to eyelids or face"
  - Gottron's papules "erythematous papules on extensor surfaces"
  - Nailfold changes
  - Lipodystrophy
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- Proximal muscle weakness
  - Difficulty with stairs, brushing hair, getting on the bus
- Voice changes or dysphagia***
  - These are worrisome signs of pharyngeal muscle involvement
  - Risk of aspiration
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- Risk of aspiration
-
- Calcinosis
  - Deposition of calcium substances in soft tissues

30 **Dermatomyositis**

31 **Dermatomyositis**

Treatment:

- High dose steroids
- IVIG
- Methotrexate

Polymyositis vs Dermatomyositis : NO RASH in PM
Muscle sx very similar
PM super rare in kids

No increased risk of malignancy in juvenile dermatomyositis (like there is in adults)

32 **Scleroderma**

33 **Scleroderma**

Localized Scleroderma

Linear morphea
Circumscribed morphea
Generalized morphea
Mixed
En Coup de Sabre

34

A previously healthy 11-year-old girl has had a dry hacking cough for 3 months associated with fatigue, occasional fevers (temperature of 38.4°C), and a 4-kg weight loss. On physical examination, the tired-appearing child has multiple firm, non-tender posterior cervical, axillary, and inguinal nodes; her respiratory rate is slightly elevated; and she has occasional wheezes. Small nodules are visible along the iris-pupil margin, and an ophthalmologist recently diagnosed anterior uveitis. Laboratory findings of note include:

Hemoglobin, 10.9 g/dL (109 g/L)
White blood cell count, 16.0x103/mcL (16.0x109/L)
Erythrocyte sedimentation rate, 32 mm/hr
Calcium, 12.3 mg/dL (3.1 mmol/L)
Serum angiotensin converting enzyme, 110 units/L (normal, 5 to 89 units/L)

Calcium, 12.3 mg/dL (3.1 mmol/L)
Serum angiotensin converting enzyme, 110 units/L (normal, 5 to 89 units/L)
A purified protein derivative test is negative. Chest radiography shows bilateral hilar adenopathy but no obvious parenchymal disease (Item Q227).

Of the following, the MOST useful test(s) for establishing the diagnosis is(are):

A) ANA and Rheumatoid Factor
B) Bone marrow biopsy
C) EBV serology
D) Immunofluorescence and ELISA for mycoplasma pneumonia
E) Lymph node biopsy

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35 **Sarcoidosis**

36 **Sarcoidosis**

Systemic granulomatous disease
- Non-caseating granulomas

More common in African Americans
Can affect just about any system: Most commonly
- General: weight loss, fatigue, malaise
- Skin: E. nodosum, rash, nodules
- Eyes: uveitis, scleritis, nodules
- Lungs: cough, chest pain, dyspnea, abnormal PFT's
- Lymphadenopathy

Also can have cardiac, neuro, endocrine invlolvement
In kids more often: rash, arthritis, uveitis

Labs: Elevated ACE, elevated Ig's, hypercalciuria, hypercalcemia

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A 3-year-old boy is brought to the clinic by his parents due to concerns about how easily he bruises. They say that since he began walking at 18 months, he frequently has large, purple bruises that appear with no known history of trauma. They do not believe that he falls more frequently than other children his age, and they deny a family history of easy bruising. On physical examination, the normally grown child has prominent eyes, a delicate and narrow nose, and numerous bruises in various stages of healing, primarily overlying his shins but also scattered elsewhere on his body. He has translucent skin over the chest, with prominent vascular markings, and his fingers are slender and hypermobile (Item Q39B).

Of the following, the condition that is MOST consistent with this boy's features is

A) Ehlers Danlos
B) Hemophilia A
C) Von Wilibrand disease
D) Stickler's

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C) Von Wilibrand disease
D) Stickler's
E) Marfan's

38 **Ehlers Danlos**

39 **Marfan Syndrome**

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A 6-year-old boy complains of achy pain in his lower legs about 1-2 times per night per week that is relieved with massage and heat. He has had no fever, rash, fatigue, joint swelling, weight loss, or other systemic symptoms. The pain is always better in the morning, and he remains very active. He has had no unusual or compulsive leg movements associated with the pain. Findings on physical examination, including thorough joint, muscle, and neurologic evaluation, are normal.

Of the following, the MOST appropriate next step in the care of this child is to:

A) Obtain a bone scan
B) Obtain CBC,ESR,RF
C) Prescribe Calcium and Vit D
D) Prescribe muscle stretching, analgesia and warmth
E) Refer to orthopedist

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What if the question said he gets pain in one leg, worse at night that sometimes wakes him from sleep and is always relieved with an NSAID?

Think osteoid osteoma

42 **Benign Causes of Joint Pain**

- Joint complaints are very common in childhood
- 85% of children asked at a well visit will say they have joint pain
- Always look for red flags: night waking, unilateral pain, no relief with simple measure, growth disturbance

Think of:

- Hypermobility syndrome
- "Benign joint pains"
- Growing pains
-

Usually reassurance to child and parent that it will improve with time if sufficient treatment. PRN NSAID ok too.

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