Objectives
• Compare epidemiologic and clinical manifestations of the various types of chronic arthritis in children, mainly juvenile idiopathic arthritis (JIA).
• Identify the main clinical and laboratory findings in pediatric systemic lupus erythematosus.
• Recognize the major clinical features of juvenile dermatomyositis and differential diagnosis.

JIA Nomenclature
• Juvenile Idiopathic Arthritis (JIA)
  – different subgroups and definitions
JIA is the most common chronic rheumatic disease in childhood

- Incidence: 1/10,000 children/year
- Prevalence: ~1:1,500
- Estimated 300,000 children in US with some form of chronic arthritis

Classification of JIA

- Systemic
- Pauciarticular (oligoarthritis)
- Polyarticular RF-
- Polyarticular RF+
- Other types of chronic arthritis
  - Psoriatic arthritis
  - Enthesitis related arthritis - spondyloarthropathies

JIA Definition

- Age at onset < 16 years
- Arthritis
  - Swelling or limitation of motion, warmth, tenderness (two of three)
- Duration > 6 weeks
- Exclusion of other causes
- Onset type determined in first 6 months
Question 1
Which type of arthritis is NOT associated with uveitis?
1. Systemic onset
2. Pauciarticular
3. Polyarticular
4. Spondyloarthropathy (enthesitis-related)
5. Psoriatic

Systemic JIA
- Arthritis with (or preceded) by >2 weeks of fever
  - Intermittent,
  - Well appearing when afebrile

Systemic JIA
- Rash: evanescent, salmon or erythematous, migratory
  - Increased during fever
  - Skin folds
  - Koebner phenomena
Systemic JIA
- Hepatosplenomegaly, lymphadenopathy
- Serositis: pleuritis, pericarditis
- Arthritis can appear up to 6 months after fever
  - Polyarticular – destructive
  - Pauciarticular – usually nondestructive

Systemic JIA – Epidemiology and Lab
- 15% of JRA cases
- Male = female
- All ages
- No uveitis
- ANA, rheumatoid factor negative
- Ferritin often very elevated
Pauciarticular JIA: 1-4 Joints

- 40-50% of JRA
- Peak age: 1-3 yrs
- Large joints
  - Hip disease rare as isolated joint
- Female: Male = 3:1
- ANA+: 50-75%
- Rheumatoid factor negative

Local Complications of Pauciarticular JIA

- Joint contractures
- Leg-length discrepancy
- Atrophy of muscles

Uveitis

- Chronic, insidious, anterior, bilateral, usually not painful or red
  - 30% in oligoarthritis;
  - 5-10% in polyarthritis
- Need to screen for uveitis
  - Mainly young girls, ANA+, early in disease

Image courtesy of Careen Lowder, MD, PhD Cole Eye Institute, Cleveland Clinic
Uveitis Complications

- Cataract
- Glaucoma
- Band keratopathy
- Visual loss 15-20%

Image courtesy of Careen Lowder, MD, PhD
Cole Eye Institute, Cleveland Clinic

Prognosis, Complications of Pauciarticular JIA

- Eyes determine prognosis
- 60% of persistent disease resolves
- 30% of extended disease resolves

Polyarticular JIA

- >4 joints in first 6 months
  - Rheumatoid factor negative
  - Rheumatoid factor positive
Special Joint, Tendon Conditions in Polyarticular JIA

- C-spine and TMJ involvement
- Baker's cyst
- Tenosynovial cyst

Polyarticular JIA (RF-)

- 25-30% of JRA cases
- Younger patients (2-4 years)
- Female: Male = 2.5:1
- Constitutional signs common
- Large joints, symmetric
  - Later small joints
  - TMJ, c-spine involvement common

Polyarticular JIA (RF-)

- ANA+: 30%
- Uveitis: 5-10%
- Prognosis
  - 75% disease persists to adulthood
  - Usually good functional outcome, mild to moderate joint damage
Polyarticular JIA (RF+)

- Similar to rheumatoid arthritis; 10% of JRA
- Adolescent females (75%)
- Symmetrical, small joint involvement
- Early erosive disease, poor prognosis
- Cyclic citrullinated peptide (CCP) antibodies usually positive

Polyarticular JIA (RF+): Extraarticular Manifestations

- Rheumatoid nodules
- Vasculitis
- Felty’s syndrome (splenomegaly, neutropenia)

Enthesitis-Related Arthritis

- Formerly called spondyloarthropathy or pauciarticular type II JRA
- HLA B27 related arthritis
- Includes
  - Juvenile ankylosing spondylitis
  - Inflammatory bowel disease related arthritis
  - Reactive arthritis (Reiter’s syndrome)
Features of ERA

• Arthritis and enthesitis

• Arthritis or enthesitis with at least 2:
  – Sacroiliac tenderness and/or inflammatory spinal pain
  – Presence of HLA B27
  – Family history of confirmed HLA B27 associated disease
  – Acute anterior uveitis (15-25%) with pain, redness, photophobia
  – Arthritis onset in >8 year old male (80% males)

Reactive Arthritis (Reiter’s Syndrome)

• Post dysenteric or genitourinary infections
  – Post dysenteric more common in younger children
• Asymmetric arthritis, lower extremities, large joints
• Conjunctivitis
• Urethritis
Reactive Arthritis (Reiter’s Syndrome)

- Rash
  - Keratoderma blennorrhagicum
  - Circinate balanitis

Psoriatic Arthritis

- Arthritis and psoriasis
  or
- Arthritis and at least 2 of following:
  - Dactylitis
  - Nail abnormalities (pitting or onycholysis)
  - Family history of psoriasis in 1st relative
- Course similar to other forms of JIA

Dactylitis – “Sausage Digits”
Nail Changes: Pitting, Onycolysis

Treatment Philosophy Has Changed

- Inverted pyramid
- Aggressive therapy
  - Window of opportunity
  - Complications, surgery
- Disease continues to adulthood
Drug Treatment

- NSAIDs (pain control)
- Disease modifying antirheumatic drugs (DMARDs)
  - Mostly Methotrexate, also Sulfasalazine, Leflunomide
- Steroids
  - Intra-articular
  - Systemic
- Anti-tumor-necrosis factor for polyarticular JIA
  - Etanercept, Adalimumab, Infliximab
- T-cell co-stimulator signal inhibitor for polyarticular JIA - Abatacept
- Antagonists to IL-1 for systemic JIA: Anakinra, Rilonacept
- Antagonist to IL-6 for systemic JIA: Tocilizumab

Other Facets of JIA

- Multidisciplinary treatment, consultants
  - Ophthalmologist, orthopedist, radiologist
- Physical and occupational therapy
- Surgery: synovectomy, joint replacement
- Psychological, education, life changes
  - Compliance issues
- Nutrition

Systemic Lupus Erythematosus (SLE)

Prototypic Autoimmune Disease
**SLE Definition**

Episodic, multisystemic, autoimmune disease characterized by widespread inflammation of blood vessels and other tissues and the presence of auto antibodies, especially antinuclear antibodies (ANA).

ANA is not sufficient to diagnose SLE. Few systems may be involved for years prior to full outbreak.

**1997 ACR Classification Criteria for SLE**

- Malar rash
- Discoid lesions
- Photosensitivity
- Oral/nasal mucosal lesions
- Non erosive arthritis
- Encephalopathy
- Nephritis
  - Casts or proteinuria >0.5 g/day
- Pleuritis or pericarditis
- Cytopenias
- Positive ANA
- Positive immunoserology
  - Anti-double stranded DNA
  - Anti-Smith
  - Antiphospholipid antibodies

Must satisfy 4/11 criteria

**Epidemiology**

- Incidence in children <15: 0.5/100,000/yr
  - 20-25% of cases develop <20 years
- Prevalence: 1-6/100,000
- Increases with age; rare < 5 years old
- Female: Male 4.5:1
  - Under 10 equal incidence
- Black > Oriental > Hispanic > Caucasian
Presentation of pediatric SLE

- Variable; acute to insidious
- Arthralgia>arthritis, mild fever, myalgia, alopecia, skin rash
  - Malaise, weight loss, fatigue, adenopathy
- Thrombocytopenia with bruising or bleeding
- Nephritis, nephrosis (hematuria/proteinuria)
- Seizures, psychosis or unusual neurologic problems

Mucocutaneous – Malar Rash

Mucocutaneous – Discoid LE

- Hyperkeratosis, follicular plugs, atrophic, annular lesions
- Dermal Band Test
Mucocutaneous – Photosensitivity Rash

Mucocutaneous – Raynaud Phenomena

Mucocutaneous SLE - Other
- Oral ulcers or erythema
- Vasculitis
- Alopecia
Other Important Clinical Features

- Pleuritis, pneumonitis, “shrunken lung”
- Cardiac
  - Pericarditis
  - Myocarditis
  - Libman Sachs endocarditis
  - Coronary artery disease (late)
- GI: autoimmune hepatitis, pancreatitis

Question 2

Which of the following findings is NOT characteristic of SLE?

1. Anemia
2. Thrombocytosis
3. Anti-nuclear antibodies
4. Low complement levels
5. Thyroid antibodies

Hematologic Abnormalities

- Anemia
  - Hemolytic - Coombs positive
- Leukopenia < 4,000
  - Mainly lymphocytopenia
- Thrombocytopenia
  - Immune, may precede disease by years
- Thrombosis
  - Antiphospholipid antibodies, prolonged PTT
CNS Involvement

- Seizures
- Psychosis
- Intractable migraine headaches
- Cognitive disturbances
- Chorea
- Stroke
- Aseptic meningitis
- Need to rule out infection or metabolic cause

Nephritis

- Determines prognosis
- More frequent in children
- Develops early
- Mostly presents as asymptomatic hematuria, proteinuria, nephrotic syndrome
- Associated with low complement levels and anti double stranded DNA antibodies

WHO Classification for Lupus Nephritis

<table>
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<th>WHO Class</th>
<th>Light microscopy</th>
<th>Hematuria</th>
<th>Proteinuria</th>
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<td>VI</td>
<td>Global sclerosis</td>
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Major Laboratory Findings

- Inflammatory response – ESR
  - CRP normal unless infection
- Hematologic
  - Anemia, leukopenia, thrombocytopenia
- Hematuria, proteinuria, casts (RBC, WBC)
- Low C3, C4 levels
  - CNS, pulmonary, renal, hematologic disease

Antinuclear antibodies

- Present in virtually 100% of SLE patients
- If negative essentially rules out SLE
- Titer does not correlate with clinical course
- 30% of general pediatric population has +ANA
ANA Patterns: Peripheral (rim), homogenous, speckled, nucleolar

Lupus-associated Autoantibodies

- Anti-Ds DNA antibodies
  - Renal disease
- Extractable nuclear antigens (ENA)
  - SS-A (Ro), SS-B (La)
  - Prognostically, related to lupus (congenital heart block), Sjogrens
  - Sm: high specificity for SLE
  - RNP: more frequently found in mixed connective tissue disease
  - Histone: drug-induced lupus
  - Ribo: renal, CNS disease
- Thyroid antibodies
  - Antithyroprotein = TSH-Receptor
- Lupus Anticoagulant
  - Prolonged aPTT
  - Anticardiolipin and Anti-Beta-2-glycoprotein Ia

Treatment of SLE

- Multidisciplinary
- Steroids
  - IV pulse for serious events
- Cyclophosphamide – IV monthly pulse
  - Severe renal involvement, CNS involvement
- Mycophenolate mofetil
- Azathioprine
Treatment
- Hydroxychloroquine
  - Skin, arthritis, prevent flares (may decrease mortality)
- NSAID
  - Arthritis, pleuritis
- Rituximab – anti CD 20 (mature B cell) antibodies

Supportive Therapy
- Sun protection
  - SPF 30 or greater
- Hypertension
- Hyperlipidemia
- Preventive cardiology
- Osteoporosis prevention
- Immunize: influenza, pneumococcus

Course, Prognosis, Mortality
- Exacerbations and remissions
- Mortality <10% at 10 years
- Early deaths from active disease (renal, CNS, vasculitis) and infection
- Late deaths from coronary artery disease
- Worse in blacks, males
Neonatal Lupus

- Transmitted from mother
  - SS-A (Ro), SS-B (La) IgG antibodies
  - Mother may not have symptoms
- 3rd degree heart block - nonreversible
  - In utero, needs pacemaker

Neonatal Lupus

- Skin rash
  - Subacute cutaneous lesions
- Hepatitis
- Thrombocytopenia
- Transient, usually doesn’t need treatment

Juvenile Dermatomyositis (JDM)

Autoimmune inflammatory disease of muscle and skin
JDM Definition
Multisystem disease characterized by acute and chronic inflammation of skin and striated muscle, resulting from small blood vessel vasculopathy.

Diagnostic Criteria
- Symmetrical proximal muscle weakness
- Characteristic cutaneous changes
- Elevation of at least one muscle enzyme
- Electromyography myopathic changes
- Muscle biopsy showing inflammation, necrosis and regeneration
- Rash + 3 other criteria

JDMS: Epidemiology
- Incidence: 2-4/100,000/yr
- Age: mean 7 years
- Race: Black > Caucasians
- Geography: North America, Europe
- Gender: female 2:1 male in US, Europe
Clinical Onset

- Usually insidious
  - Acute, subacute in 33%
  - Rare rhabdomyolysis
- Frequent constitutional signs
  - Fever, malaise, weight loss, fatigue

Muscle Disease

- Symmetrical proximal muscle weakness
  - Limb girdle, neck flexors, abdomen
  - Affects climbing stairs, walking, combing hair etc.
  - Regression of milestones
  - Distal weakness late
  - Gower sign
- Muscle pain (30-80%)
- Dysphagia, drooling, dyspnea, aspiration, dysphonia, pulmonary disease (5-45%)

Question 4

Which of the following skin findings is specific for JDMS?

1. Calcinosis
2. Photosensitivity
3. Gottron’s papules
4. Raynaud’s phenomena
5. Periungual capillary findings
Cutaneous Disease (75-100%)

- Heliotrope: violaceous discoloration, edema and telangiectasia of eyelids

Cutaneous Disease

- Gottron’s papules: pathognomonic
  - Shiny, scaly erythematos papules
  - Finger knuckles (MCP, PIP)
  - Elbows, knees

Cutaneous Disease

- Photosensitivity
- Ulcers
- Lipodystrophy
Cutaneous Disease

- Periungual capillary abnormalities

Other Clinical Features

- Nonerosive arthritis (30%)
- Raynaud’s phenomena
- Abdominal pain – visceral vasculopathy
  - Poor prognosis
- Myocarditis, heart block

Calcinosi

- Prevalence 25-50%
- Late complication
- Usually poorly treated disease
Differential Diagnosis

• Infectious
  – Postviral - influenza, coxsackie B
  – Severe calf myalgia, CPK increase, transient
  – Streptococcal, staphylococcus
  – Trichinosis
  – Toxoplasma
  – Polio

• Primary myopathies
  – Dystrophy (Duschenne, etc.)
  – Congenital
  – Metabolic myopathies
    – Glycogen storage (V, VII), others
    – Endocrinopathies: Addison, thyroid (hyper and hypo), Cushing

• Drugs and toxins
  – Steroids, statins, colchicine, diuretics, hydroxychloroquine, amphotericin B
  – Alcohol, snake venom
  – Trauma, heat exhaustion - exertion
  – Neurogenic atrophy
Laboratory Evaluation

- Inflammatory indices
  - Often normal, even in active disease
- Muscle enzymes
  - CPK, AST, ALT, LDH, aldolase
- Immunoglobulin deficiency
- Autoantibodies
  - ANA, muscle specific (Jo-1, Mi-2)

MRI

Muscle Biopsy

- Perifascicular inflammation
- Atrophy
- Regeneration
- Vasculopathy
**Treatment**
- Steroids, steroids, and more steroids
- Methotrexate
- Hydroxychloroquine for skin
- IVIG
- Cyclosporin/tacrolimus
- Cyclophosphamide
- Others (Rituximab?, Infliximab?)

**Treatment - Supportive**
- Respiratory support
- Thickened feedings, elevate bed, nasogastric feeding
- Oral suctioning
- Skin care
  - Ulcers, fissures, calcinosis, secondary infection
- Sun protection

**Prognosis and Course**
- Greater than 90% survival
  - Malignancy rare
  - Worst in black females
- Monocyclic (40%)
  - Polycyclic (20%)
  - Chronic (30%)
  - Ulcerative (10%)
Answers to multiple choice questions

- Question 1: 1
- Question 2: 2
- Question 3: 1
- Question 4: 3

Objectives

- Review presentation, evaluation, and management of pediatric rheumatologic conditions including:
  - Familial Mediterranean Fever
  - Henoch-Schönlein Purpura
  - Juvenile Fibromyalgia
  - Periodic Fever, Aphthous Stomatitis, Pharyngitis, and Adenopathy Syndrome
  - Raynaud’s phenomenon
  - Scleroderma
  - Sarcoidosis
  - Sjögren’s Syndrome
  - Takayasu arteritis
  - Wegener’s Granulomatosis
Question 1

- You are seeing a 3 yr white male for fever who has recently moved to your city. Going through his chart, you notice that he has been seen about every 4 weeks for fevers for the last 6 months. During the history, his mother reports that the fevers come every 26-27 days ‘like clockwork’ and are associated with sore throat, swollen glands, and abdominal pain. His fevers remain in the 102-103ºF range for 4-5 days and are minimally responsive to Tylenol and Motrin. Between episodes he is completely healthy. On examination, his growth parameters are normal. He is febrile at 39.6ºC and is ill-appearing. There is oropharyngeal erythema, tonsillar hypertrophy, posterior pharyngeal ulcers, anterior cervical lymphadenopathy, and abdominal tenderness to palpation. What is the most likely laboratory finding in this patient?
  1. Tumor Necrosis Factor Receptor Super Family 1A gene mutation
  2. Elevated erythrocyte sedimentation rate and C-reactive protein
  3. Elevated ferritin
  4. Mevalonate kinase gene mutation
  5. Mediterranean fever gene mutation

Question 2

- Which therapeutic option has shown the greatest efficacy in treatment of this patient’s condition?
  1. Adenotonsillectomy
  2. Prednisone 1mg/kg by mouth at onset of symptoms
  3. Cimetidine 10 mg/kg by mouth four times a day
  4. Infliximab 5 mg/kg every 4 weeks
  5. Methotrexate 15 mg/m2 subcutaneously every week

Answers: Question 1 – B; Question 2 – A

- Periodic Fever, Aphthous Stomatitis, Pharyngitis, and Adenopathy (PFAPA) Syndrome
  - Most common cause of recurent fevers
  - Etiology unknown
  - Periodic, stereotypic; attacks of fever lasting 5 days and constellation of above symptoms every 4-5 weeks
  - Abdominal pain very common
  - Consider cyclic neutropenia
  - Single dose of prednisone at 1 mg/kg po effective in 90% of patients but may increase frequency of attacks
  - Adenotonsillectomy is ‘curative’ – complete cessation of attacks within 6-12 months
Question 3

A 16 yo white female presents for evaluation of fatigue. For the last 4 months, she has been experiencing intermittent muscle and joint pain, in addition to chronic daily headaches. The pain wakes her from sleep and she often feels tired upon waking. She has not been to school in 2 weeks and takes a 3-hour nap every day. Her examination is normal except for tenderness over her trapezius, 2nd ribs, medial tibial tuberosities, and gluteus medius. What treatment is most likely to result in long-term improvement in this patient?

1. Pregabalin 50 mg three times a day
2. Referral to pain medicine
3. Physical therapy with cognitive behavioral therapy
4. Oral prednisone at 1 mg/kg per day for 5 days followed by a 2-week taper
5. Oral naproxen at 10 mg/kg twice a day

Answer: Question 3 - C

Juvenile Fibromyalgia
- Constellation of chronic headaches, abdominal pain, fatigue, disordered sleep, musculoskeletal pain, and anxiety
- Tender points
- Adolescent female
- No data supporting use of pregabalin or duloxetine in children
- Physical therapy + counseling offer best opportunity to restore function
- Low dose amitriptyline may also be beneficial

Question 4

A 15 yo female presents to your office for evaluation of fatigue, low-grade fevers, headaches, abdominal pain after eating, and pain in her left arm with exercise. On examination she is obese and mildly hypertensive (132/89) but your nurse also notes a 15 mm Hg difference in systolic blood pressure between arms. The remainder of your exam is normal. What is the next most appropriate step in your evaluation?

1. Magnetic resonance angiography of aorta and great vessels
2. Referral to physical therapy
3. Referral to neurology
4. Initiate amitriptyline at 25 mg by mouth before bed
5. Initiate enalapril 10 mg once a day
Answer: Question 4 - A

- Takayasu Arteritis
  - Large vessel vasculitis affecting aorta and branches
  - Constitutional symptoms are common
  - Claudication, bruits, and asymmetric blood pressures reflect arterial stenoses
  - Stenoses >> aneurysms
  - Children more likely to present with hypertension
  - Magnetic resonance angiography very sensitive

Question 5

- An 8 yo white female is being seen for a rash. Her mother noted a linear band of violet skin on the left outer thigh which has become larger and 'thicker' over the last several months. On examination, there is a 11 cm by 3 cm plaque with violaceous borders extending from the groin to the knee on the left medial thigh. What information can you offer to this mother about the prognosis of this condition?

1. There is an 80% chance of interstitial lung disease
2. Many children with this condition develop Raynaud’s phenomenon
3. Over 20% of children have extra-cutaneous manifestations
4. The rash will get smaller and fade away completely
5. About 10% develop pulmonary hypertension

Answer: Question 5 – C

- Localized scleroderma
  - Progressive fibrotic condition affecting skin and subcutaneous tissue
  - Localized >> Systemic (10:1)
  - 2 Most common subtype
    - Linear
    - Circumscribed morphea
  - Almost 25% have extra-cutaneous manifestations
    - Arthritis
    - CNS
Question 6

- You are seeing a 16 year-old white female for an annual check-up who had been cared for by your partner until his recent retirement. Going through her chart, you notice that she has been treated 4 times in the last year for sinusitis and 3 times for culture-negative urinary tract infections. On examination, her blood pressure is mildly elevated at 139/88 mm Hg. Inspection of her nasal mucosa demonstrates very edematous and friable mucosa. Her lungs are clear and she has no pitting edema. Her urinalysis today demonstrated 2+ blood, 2+ protein, and 1+ leukocyte esterase. Which of following are her labs most likely to demonstrate?
  1. (+) Antineutrophil cytoplasmic antibody, cytoplasmic pattern, anti-myeloperoxidase(+)
  2. (+) Antineutrophil cytoplasmic antibody, cytoplasmic pattern, anti-Proteinase 3(+)
  3. Antineutrophil cytoplasmic antibody, perinuclear pattern, anti-myeloperoxidase(+)
  4. Antinuclear antibody(+), anti-double-stranded DNA(+)
  5. Antinuclear antibody(+), anti-Smith(+)

Answer: Question 6 – B

- Wegener’s Granulomatosis
  - Vasculitis of predominantly medium-sized vessels
  - M:F = 1:4; mean age 14.5 yrs
  - Triad: Sinusitis, pulmonary hemorrhage, glomerulonephritis
  - Consider in recurrent/refractory asthma, otitis media, sinusitis, epistaxis or UTI
  - Constitutional symptoms common
  - CT scan much more sensitive than plain radiography of chest
  - Subglottic stenosis present in 50%
  - (+) ANCA, cytoplasmic pattern, + anti-Proteinase-3 antibodies in >90% patients
Question 7

- Which of the following findings indicate an underlying rheumatologic condition as a cause of Raynaud’s phenomenon?
  1. Positive antinuclear antibody testing and fatigue
  2. Positive antinuclear antibody testing and joint pain with activity
  3. Positive antinuclear antibody testing and abnormal nailfold capillaries
  4. Positive antinuclear antibody testing and family history of lupus
  5. Positive antinuclear antibody testing and hypermobility

Answer: Question 7 – C

- Raynaud’s
  - Episodic color changes in extremities
    - White → Blue → Red; biphasic more common
    - Sharply demarcated at joint
    - Paresthesias
    - Non-healing ulcers
  - Risk factors: female, thin, stimulant usage
  - ANA and presence of abnormal nailfold capillaries are indicative of underlying rheumatologic process
Question 8

A 15 yo Caucasian male presents for evaluation of cough. He has also reported that his vision has become ‘blurry’ recently and that he has painful ‘dark red bumps’ over his shins. On examination, you note keratic precipitates, tender, erythematous nodules over the shins, and soft crackles in the lower lung fields bilaterally. PFTs in your office demonstrate his FVC and FEV1 are 60% of predicted and a chest xray demonstrates hilar lymphadenopathy. Which of the following tests demonstrates the highest specificity in this condition?

1. Angiotensin converting enzyme
2. Antinuclear antibody
3. Anti-neutrophil cytoplasmic antibody
4. Inflammatory bowel disease series-7 panel
5. Lymph node biopsy demonstrating noncaseating granulomas

Answer: Question 8 - E

• Sarcoidosis
  – Rare multisystem granulomatous disease
  – Noncaseating epithelioid granulomas
  – Present with constitutional complaints
  – Respiratory, cutaneous, and musculoskeletal complaints also common
  – Erythema nodosum, peripheral lymphadenopathy, and uveitis are most common physical exam findings
  – Most children with have hilar adenopathy
  – ACE: low-specificity
  – Diagnosis should be based on tissue*

*Children's Hospital Boston
A 4 yo white male is brought to your office by his mother for an urgent appointment. Two days ago, he developed a rash on his legs and feet and today he refused to bear any weight. He has also vomited several times today and his mother thinks that she may have seen some blood in his stool. His vital signs are normal. On examination, his abdomen is tender to palpation and he has a purpuric rash on his feet and lower legs associated with diffuse non-pitting edema. What is the next most appropriate step in his management?

1. Education regarding his condition and reassurance
2. Abdominal ultrasound
3. Prednisone 2 mg/kg by mouth once a day for 5 days
4. Urinalysis
5. 25 cc/kg normal saline bolus
**Answer: Question 9 – B**

- **Henoch-Schönlein Purpura**
  - Most common childhood vasculitis
  - Medium-to-small vessel, IgA-mediated, vasculitis
  - Palpable purpura in 100%
  - GI complaints in 65-70%; intussusception (ileoileal) 1-5%
  - Arthritis: 50-80%
  - Nephritis: 20-35%
  - Role of steroids:
    - May reduce duration and severity of arthritis and abdominal pain
    - No effect on renal disease

**Question 10**

- An 8 yo white male presents for evaluation of recurrent fevers. For the last 9 months, he has had fevers every 5-6 weeks which last 3 days and are associated with intense abdominal pain. Several relatives in Italy have had similar symptoms and passed away from renal failure. Which medication will prevent fevers and improve this patient’s prognosis?
  1. Daily oral prednisone
  2. Daily oral colchicine
  3. Daily oral aspirin
  4. Weekly subcutaneous etanercept
  5. Monthly intravenous infliximab

**Answer: Question 10 – B**

- **Familial Mediterranean Fever**
  - Autosomal recessive autoinflammatory syndrome
  - Mutation MEFV gene → pyrin → IL-1 regulation
  - Sephardic Jews, Ashkenazi Jews, Turks, Arabs, Armenians
  - 80% <10 yo; 90% < 20 yo
  - 3 days of fever associated with abdominal pain and arthritis
  - Increased risk of amyloidosis & renal failure
  - Colchicine reduces incidence of amyloidosis from 35% to 5%
• Question 1 & 2

• Question 3

• Question 4

• Question 5

• Question 6

• Question 7

• Question 8

• Question 9

• Question 10
GOOD LUCK!!!