

# PEDIATRIC RHEUMATOLOGY: A PRIMARY CARE PERSPECTIVE

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## PEDIATRIC RHEUMATOLOGY I

I have no personal or financial conflicts of interest involving this presentation.  
I WILL BE discussing some treatments that are not FDA approved.

## OBJECTIVES

- ▶ INTRODUCE THE LEARNER TO COMMON PEDIATRIC RHEUMATOLOGICAL PROCESSES.
- ▶ DISCUSS COMMON CLINICAL PRESENTATIONS AND APPROPRIATE EVALUATION
- ▶ UNDERSTAND GENERALIZED TREATMENT APPROACHES FOR TREATMENT OF ABOVE.

## JUVENILE IDIOPATHIC ARTHRITIS

- ▶ Confusing because of various nomenclatures and confusion with adult specific disease processes.
- ▶ Some overlap and imperfect classification.
- ▶ New classification is based on the 1997 International League of Associations for the Rheumatology (ILAR)
- ▶ Terms "Juvenile Rheumatoid Arthritis" and "Juvenile Chronic Arthritis" were abandoned.
- ▶ The new term Juvenile Idiopathic Arthritis (JIA) was adopted.

## JUVENILE IDIOPATHIC ARTHRITIS

- ▶ Occurring before the age of 16 y/o.
- ▶ Involving Persistent Synovitis in one or more joints.
- ▶ Synovitis for at least 6 weeks.

## JUVENILE IDIOPATHIC ARTHRITIS

- ▶ OLIGOARTICULAR
- ▶ POLYARTICULAR- RHEUMATOID FACTOR POSITIVE
- ▶ POLYARTICULAR –RHEUMATOID FACTOR NEGATIVE
- ▶ JUVENILE PSORIATIC ARTHRITIS
- ▶ ENTHESITIS-RELATED ARTHROPATHIES
- ▶ UNDIFFERENTIATEDJIA
- ▶ SYSTEMIC JIA

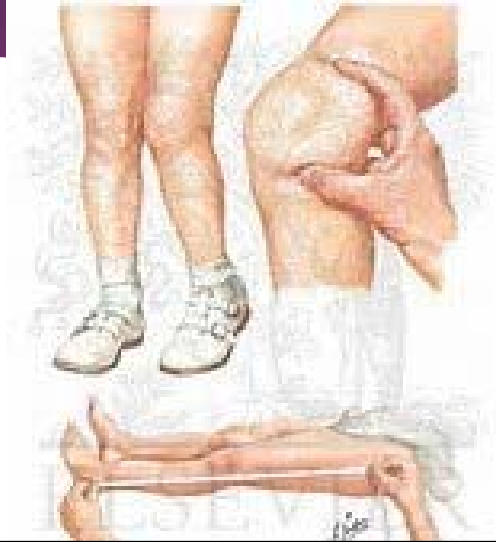
## OLIGOARTICULAR JIA

- ▶ Four or fewer joints during the first 6 months.
- ▶ 1-7 years of age
- ▶ F:M 3:1
- ▶ Uveitis F:M 6.5:1
- ▶ Typically involves incidental joint swelling or limp
- ▶ Knee>ankle>elbow>wrists
- ▶ Fever, rash, and night pain typically do not occur

## OLIGOARTICULAR JIA

- ▶ Persistent- only 4 or fewer joints throughout the course of the disease
- ▶ Extended- Initially develops 4 or fewer joints but other joint involvement after 6 months

## OLIGOARTICULAR JIA



## OLIGOARTICULAR JIA

- ▶ UVEITIS is most commonly associated with this form of JIA
- ▶ Typically asymptomatic but if not diagnosed and treated appropriately can result in cataracts, glaucoma and blindness
- ▶ ANA status is determined strictly to determine risk of Uveitis and determine frequency of slit lamp examination
- ▶ High risk factors include positive ANA, female sex, age at onset less than 6 years old, and duration of disease less than 4 years

## OLIGOARTICULAR JIA



## OLIGOARTICULAR JIA

- ▶ If ANA positive, less than 6y/o at diagnosis and duration less than 4 years then needs eye exam every 3 months.
- ▶ If ANA positive, age at onset less than 6 y/o and duration of disease greater than 4 years then check every 6 months.
- ▶ If ANA positive, age at onset less than 6 y/o and duration of disease >7 years then check every 12 months.
- ▶ If ANA positive, age at onset greater than 6y/o and duration less than 4 years then check every 6 months.
- ▶ If ANA positive, age at onset greater than 6 y/o and duration more than 4 years then check every 12 months.

## OLIGOARTICULAR JIA

- ▶ Treatment usually involves joint injections and NSAIDs.
- ▶ Occasionally requires Disease Modifying Rheumatological Diseases (DMRDs) or biologics.
- ▶ Pts will often out grow this disease.
- ▶ Continue eye exams even if arthritis is quiet.

## POLYARTICULAR JIA

- ▶ Rheumatoid Factor Negative- peaks at 1-3 y/o and at 10 y/o.
- ▶ Rheumatoid Factor Positive- More common in teens. Probably early onset of Adult type Rheumatoid Arthritis.
- ▶ Uveitis only in 10-15% of ANA positive. Needs eye exams every 6 months.
- ▶ Typically do require DMRD and often times biologic.

## POLYARTICULAR JIA



## PSORIATIC ARTHRITIS

- ▶ Arthritis and psoriasis
- ▶ Arthritis and at least two of the following:
  - Dactylitis
  - Nail pitting or oil spots or onycholysis
  - Family history of psoriasis in at least one 1<sup>st</sup> degree relative



# PSORIATIC ARTHRITIS



Figure 2 - Dactylitis, or "sausage digit," is seen in the toes of a child with psoriatic juvenile idiopathic arthritis.

# PSORIATIC ARTHRITIS



© ACR

## PSORIATIC ARTHRITIS



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## PSORIATIC ARTHRITIS

- ▶ About 20% of Pts with Psoriasis develop arthritis.
- ▶ Arthritis can proceed Psoriasis by months to years.
- ▶ Uveitis and positive ANA in 30-50% of Pts.
- ▶ DIP involvement can occur.

## PSORIATIC ARTHRITIS

- ▶ Almost all these Pts will require Methotrexate.
- ▶ Many will require biologics.
- ▶ Topical Steroids are safe but be careful with systemic steroids.
- ▶ Systemic Steroids can induce erythroderma.

## PSORIATIC ARTHRITIS



## ENTHESITIS-RELATED ARTHRITIS

- ▶ Presence or history of SI joint tenderness and /or inflammatory lumbosacral pain
- ▶ HLA -B27 Antigen Positive
- ▶ Male >6 years of age
- ▶ Associated Uveitis
- ▶ 1<sup>st</sup> Degree relative with Ankylosing Spondylitis, Enthesitis-Related Arthropathy, SI with IBD, or Reactive Arthritis

## ENTHESITIS-RELATED ARTHRITIS

- ▶ 25% of Enthesitis -Related Arthritis Pts are HLA-B27 Negative
- ▶ Most People with HAL-B27 Antigen do NOT have Enthesitis-Related Arthritis
- ▶ Older, male children
- ▶ Familial 10-20% of the time
- ▶ Worse after rest
- ▶ Peripheral Arthritis
- ▶ Sacroiliac Pain

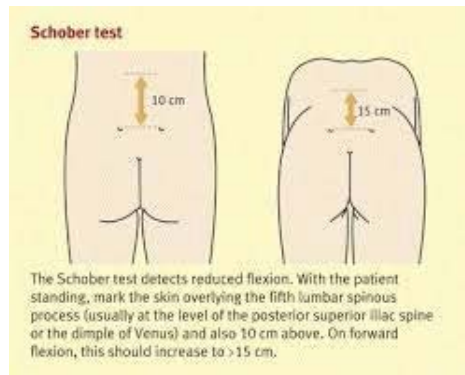
# ENTHESITIS-RELATED ARTHRITIS



# ENTHESITIS-RELATED ARTHROPATHIES



## ENTHESITIS-RELATED ARTHROPATHIES



## ENTHESITIS-RELATED ARTHROPATHIES

- ▶ Peripheral arthritis will respond to NSAIDs, Steroids, DMRDs.
- ▶ Axial Symptoms typically axial symptoms require biologic.
- ▶ Increased activity and PT is critical

## UNDIFFERENTIATED JIA

- ▶ ARTHRITIS ASSOCIATED WITH INFLAMMATORY BOWEL DISEASE
- ▶ REACTIVE ARTHRITIS

## ARTHRITIS ASSOCIATED WITH INFLAMMATORY BOWEL DISEASE

- ▶ 25% of Pts with IBD
- ▶ PERIPHERAL ARTHRITIS
- ▶ AXIAL ARTHRITIS

## PERIPHERAL IBD ARTHRITIS

- ▶ Arthritis flares are correlated with GI flares
- ▶ M=F
- ▶ NOT ASSOCIATED WITH HLA-B27

## AXIAL IBD ARTHRITIS

- ▶ Spine, hips, SI joints
- ▶ M>>F
- ▶ ASSOCIATED WITH HLA-B27
- ▶ Independent of GI flares



## REACTIVE ARTHRITIS

- ▶ Associated with GI or GU infections
- ▶ Yersinia, Shigella, Salmonella, Campylobacter
- ▶ Chlamydia or Mycoplasma
- ▶ May be associated with oral and genital ulcers, and papular skinlesions.
- ▶ Enthesitis and dactylitis
- ▶ Large weight bearing joints
- ▶ Treat active infection
- ▶ Start with NSAIDs, may require Methotrexate, sulfasalazine or biologics

## SYSTEMIC JIA (STILL'S DISEASE)

- ▶ Not truly a JIA but more closely related to recurrent fever syndromes
- ▶ Autoinflammatory (Intrinsic Immune System) vs Autoimmune (Adaptive Immune System)
- ▶ M=F
- ▶ Peak Age 5-10 years old
- ▶ Fever is always present and usually quotidian
- ▶ Toxic appearing during fever but can appear normal when afebrile

## SYSTEMIC JIA (STILL'S DISEASE)

- ▶ Migratory rash, often with the fever
- ▶ Macular, pink to salmon color
- ▶ May be pruritic
- ▶ Koebner's Phenomenon

## SYSTEMIC JIA (STILL'S DISEASE)



## SYSTEMIC JIA (STILL'S DISEASE)

- ▶ Synovitis-may be delayed for weeks or months
- ▶ Myalgia-CPK usually normal but aldolase can be elevated
- ▶ Pericarditis and myocarditis
- ▶ Serositis
- ▶ Lymphadenopathy
- ▶ Hepatosplenomegaly
- ▶ Abdominal Pain
- ▶ Weight loss and fatigue

## SYSTEMIC JIA (STILL'S DISEASE)

- ▶ WBC typically high 10s to 40 or 50K
- ▶ PLTs typically 400K or higher
- ▶ Normocytic, normochromic anemia
- ▶ Elevated Ferritin
- ▶ Elevated CRP and ESR

## SYSTEMIC JIA (STILL'S DISEASE)

- ▶ Diagnosis of exclusion
- ▶ R/O Infectious Processes
- ▶ R/O Leukemia/Lymphoma (Peripheral Smear, LDH, UA)
- ▶ R/O Kawasaki's Dz

## SYSTEMIC JIA (STILL'S DISEASE)

- ▶ Treatment includes NSAIDs, Steroids, Methotrexate
- ▶ Does not respond well to TNF antagonists
- ▶ Responds well to IL-1 (Anakinra, Ilaris) and IL-6 Antagonist (Actemra)
- ▶ Risk of progressing to Macrophage Activation Syndrome which can be fatal

## MACROPHAGE ACTIVATION SYNDROME (MAS)

- ▶ A progressive inflammatory response in which the inhibitors of inflammation are not adequately controlling the inflammatory system.
- ▶ Leads to depletion to NK Cell and a "Cytokine Storm"
- ▶ Very similar to Hemophagocytic Lymphocytic Histiocytosis (HLH)
- ▶ HLH is a genetic defect in counter inflammatory regulators-typically occurs in younger Pts and usually requires Bone Marrow Transplant for cure

## MACROPHAGE ACTIVATION SYNDROME (MAS)

- ▶ Can be caused by infections process'- EBV, Rickettsial Dz, Gram negative sepsis
- ▶ Leukemia/Lymphoma
- ▶ Immunodeficiency
- ▶ Rheumatologic Processes-Typically Still's Disease but also Systemic Lupus Erythematosus

## MACROPHAGE ACTIVATION SYNDROME (MAS)

- ▶ Results in high fever
- ▶ Rash
- ▶ HSM
- ▶ LAD
- ▶ Elevated LFTs
- ▶ DIC
- ▶ ARDS
- ▶ AKI

## MACROPHAGE ACTIVATION SYNDROME (MAS)

- ▶ Ferritin is typically high, often above 10,000
- ▶ CRP is high but ESR can be low (secondary to depletion of fibrinogen)
- ▶ Cytopenia of WBCs and PLTs (be wary of a sudden drop of either)

## MACROPHAGE ACTIVATION SYNDROME (MAS)

- ▶ Treat the underlying cause
- ▶ Steroids
- ▶ Methotrexate
- ▶ IL-1 Inhibitor Anakinra

## SYSTEMIC LUPUS ERYTHEMATOSIS



## SYSTEMIC LUPUS ERYTHEMATOSIS

- ▶ A chronic inflammatory disease affecting the skin, kidneys, lungs, nervous system, serous membranes and other organs.
- ▶ Involves excessive activation of the Innate Immune System.
- ▶ Involves excessive activation of the adaptive immune system including both the humoral and cell mediated pathways.

## SYSTEMIC LUPUS ERYTHEMATOSIS

- ▶ Clear Genetic Component- concordance rate of 14-57% in monozygotic twins.
- ▶ Hormonal Factors Prepubertal F: M ratio 3.1 while post puberty that Ratio increases to 7-15:1
- ▶ X Chromosome Factors-  
XXY (Klinefelter's) 14 fold increase  
XO (Turner's) significantly lower rate



## SYSTEMIC LUPUS ERYTHEMATOSIS

- ▶ Infectious Triggers EBV and CMV most notable
- ▶ UV light exposure

## SYSTEMIC LUPUS ERYTHEMATOSIS

- ▶ Much more common in Americans of Asian, African, or Hispanic Descent
- ▶ Overall Prevalence is 1-2/1,000
- ▶ 18-20% Diagnosed before the age of 18

## SYSTEMIC LUPUS ERYTHEMATOSIS

- ▶ HEMATOLOGIC
  - Anemia Chronic Inflammation
  - Hemolytic Anemia
  - Thrombocytopenia
  - Leukopenia-especially lymphopenia
  - Coagulopathies

## SYSTEMIC LUPUS ERYTHEMATOSIS

- ▶ MUCOCUTANEOUS CHANGES
  - Malar Rash
  - Oral Ulcers
  - Nasal Ulcers
  - Photosensitivity
  - Alopecia

SYSTEMIC LUPUS ERYTHEMATOSIS



SYSTEMIC LUPUS ERYTHEMATOSIS



# SYSTEMIC LUPUS ERYTHEMATOSIS



# SYSTEMIC LUPUS ERYTHEMATOSIS



## SYSTEMIC LUPUS ERYTHEMATOSIS

- ▶ MUSCULOSKELETAL INVOLVEMENT

- Arthritis

- Arthralgia

- Osteopenia

## SYSTEMIC LUPUS ERYTHEMATOSIS

- ▶ FEVER-

- If Greater than 38.6 or associated with chills  
consider infectious until proven otherwise

## SYSTEMIC LUPUS ERYTHEMATOSIS

- ▶ NEUROLOGIC
  - Depression
  - Headaches
  - Seizures
  - Peripheral Neuropathy

## SYSTEMIC LUPUS ERYTHEMATOSIS

- ▶ RENAL DISEASE
  - 27 percent of children with SLE
  - Class of Disease based on renal biopsy
  - Class I- VI
  - Class III and IV require aggressive immunosuppression
  - Class V requires Mycophenolate

## SYSTEMIC LUPUS ERYTHEMATOSIS

- ▶ PULMONARY
  - Pneumonitis
  - Plevritis
  - Pulmonary Hemorrhage
  - Pulmonary HTN
  - "Shrinking Lung Syndrome"

## SYSTEMIC LUPUS ERYTHEMATOSIS

- ▶ CARDIAC
  - Pericarditis
  - Valvular Disease
  - Myocarditis
  - Increased Risk for future CAD

## SYSTEMIC LUPUS ERYTHEMATOSIS

- ▶ EVALUATION:
  - CBC/Peripheral Smear
  - CMP- elevated liver enzymes, Increased BUN/Cr
  - C3 and C4- both decreased in active Dz
  - UA- can see proteinuria, pyuria, hematuria
  - Urine Pr: Cr Ratio
  - Blood Cultures if Febrile

## SYSTEMIC LUPUS ERYTHEMATOSIS

- ▶ What about an ANA?
- ▶ If ANA positive will need further check with Extractable Nuclear Antigens (ENA) and Anti DS DNA



## SYSTEMIC LUPUS ERYTHEMATOSIS

- ▶ Anti-DS ANA >97% Specific for SLE
- ▶ Anti-Smith >95 % Specific for SLE
- ▶ Anti-Ro and Anti-La associated with Neonatal Lupus as well as congenital heart block

## SYSTEMIC LUPUS ERYTHEMATOSIS

- ▶ Chest Xray
- ▶ +/- Echo
- ▶ Amylase/Lipase, abdominal imaging if abdominal pain

## SYSTEMIC LUPUS ERYTHEMATOSIS

- ▶ Steroids
- ▶ Hydroxychloroquine
- ▶ Mycophenolate
- ▶ Methotrexate
- ▶ Tacrolimus
- ▶ Rituximab
- ▶ Cyclophosphamide

## KAWASAKI DISEASE

- ▶ A vasculitis involving medium sized arteries which if untreated can result in aneurysms of the coronary arteries and early onset myocardial infarction in children or young adults.
- ▶ Associated with fever, nonexudative conjunctivitis, mucositis, rash, edema of the hands and feet and lymphadenopathy

## KAWASAKI DISEASE

- ▶ Bilateral bulbar conjunctival injection (dry) can also involve anterior uveitis
- ▶ Oral mucous membrane changes including cracked lips, injected pharynx or strawberry tongue
- ▶ Peripheral erythema and/or edema of the hands and feet

## KAWASAKI DISEASE

- ▶ Polymorphous rash
- ▶ Cervical lymphadenopathy-at least 1.5cm in diameter, usually unilateral

KAWASAKI DISEASE



KAWASAKI DISEASE



## KAWASAKI DISEASE



## KAWASAKI DISEASE

- ▶ CBC-Leukocytosis and thrombocytosis
- ▶ Elevated AST/ALT
- ▶ Elevated CRP and ESR
- ▶ Urinalysis- sterile pyuria. Remember that the source of this is urethral so may miss with catheterized sample. Inflammatory Cells are not neutrophils so leukocyte esterase will be negative

## KAWASAKI DISEASE

- ▶ Echo
- ▶ Ultrasound

## KAWASAKI DISEASE

- ▶ IVIG and Aspirin
- ▶ High Dose Solumedrol
- ▶ Remicade

## JUVENILE DERMATOMYOSITIS

- ▶ An autoimmune disease associated with T-cell infiltration of skeletal muscles and vasculitis
- ▶ Similar to the adult form of Dermatomyositis but JDM does not have an association with increased risk of occult neoplasia
- ▶ Variants include Amyopathic JDM and Polymyositis

## JUVENILE DERMATOMYOSITIS

- ▶ Typical presentation includes a history of increasing proximal weakness
- ▶ Typical Rash including:
  - Heliotropic Rash
  - Shawl Sign
  - V Sign
  - Gottron's Papules
  - Capillary Drop Out

# JUVENILE DERMATOMYOSITIS



# JUVENILE DERMATOMYOSITIS





## JUVENILE DERMATOMYOSITIS



Figure 2. Nailfold capillary abnormalities in a child with juvenile dermatomyositis. Note the abnormally large, dilated capillary loops that are abnormally few in number because neighboring loops have been obliterated ("capillary dropout"). There is also mild cuticular hypertrophy.

## JUVENILE DERMATOMYOSITIS

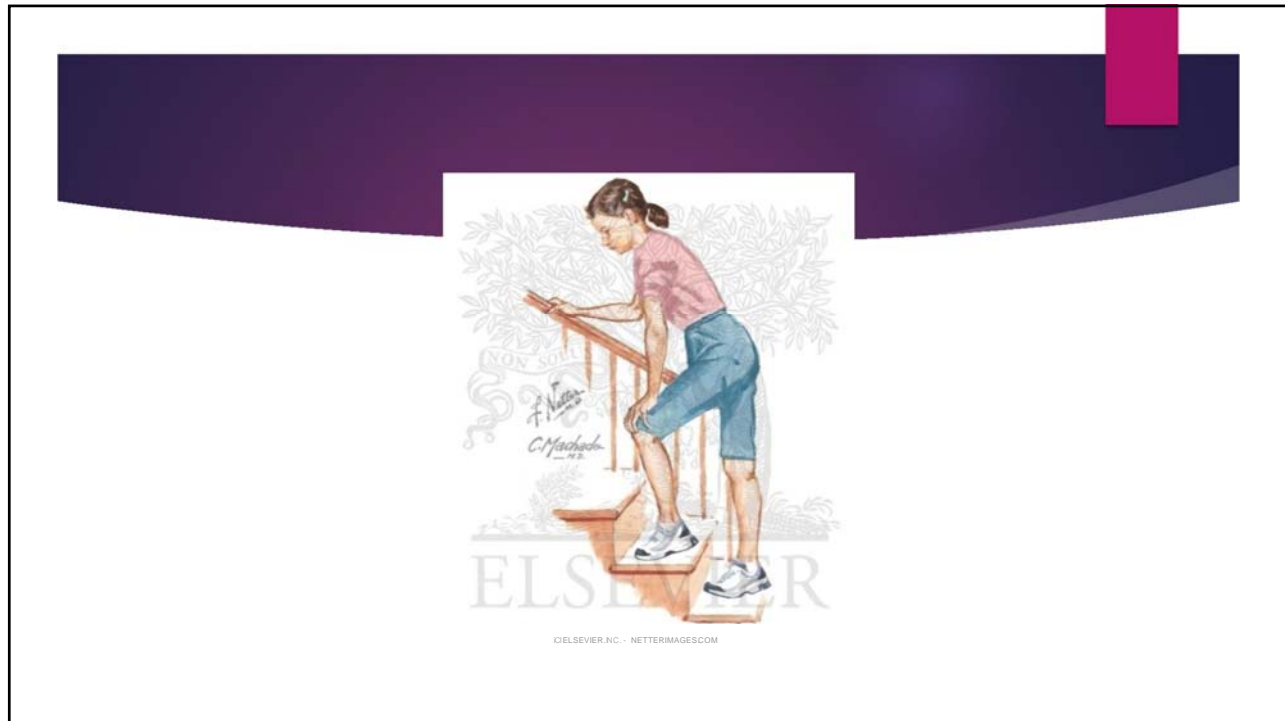
- ▶ Climbing Stairs
- ▶ Getting up from the chair or toilet
- ▶ Combing or fixing their hair
- ▶ Problems swallowing
- ▶ Voice change

## JUVENILE DERMATOMYOSITIS

- ▶ CPK
- ▶ Aldolase
- ▶ LDH
- ▶ Von Willebrand's level
- ▶ Remember that AST and ALT are also muscle enzymes

## JUVENILE DERMATOMYOSITIS

- ▶ Treatment includes steroids, methotrexate, IVIG, Mycophenolate
- ▶ Rule of 1/3rds
  - 1/3 Remission without relapse
  - 1/3 with Remission but subsequently relapse
  - 1/3 Improvement in inflammation but never go into remission



## HENOCH-SCHONLEIN PURPURA

- ▶ Also known as Ig A vasculitis
- ▶ Most common vasculitis in childhood
- ▶ Typically occurs in children between 3 and 15 years old
- ▶ Disease in older patients associated with more severe disease and an increased risk of renal disease

## HENOCH-SCHONLEIN PURPURA

- ▶ Skin
- ▶ Joints
- ▶ GI Tract
- ▶ Kidney
- ▶ Brain

## HENOCH-SCHONLEIN PURPURA



## HENOCH-SCHONLEIN PURPURA

- ▶ CBC
- ▶ PT/PTT/INR
- ▶ UA including Urine Pr:Cr Ratio

## HENOCH-SCHONLEIN PURPURA

DDX:  
Sepsis including meningococcus and Strep pneumo  
Rickettsial Disease  
Coagulopathy  
Non accidental Trauma  
Other vasculitis

## HENOCH-SCHONLEIN PURPURA

### Treatment:

NSAIDs-Consider COX-2 specific or adding PPI

Steroids-Risk of relapse if course is too short

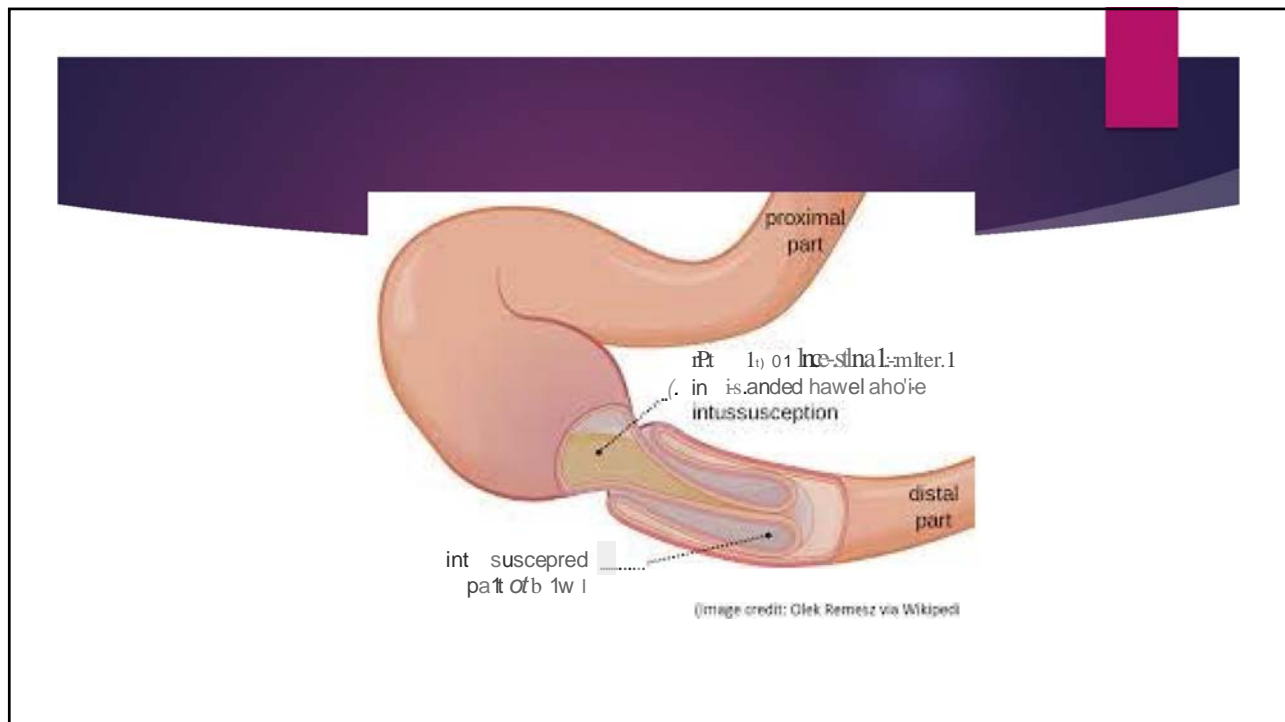
## HENOCH-SCHONLEIN PURPURA

### Complications:

Intussusception

Chronic Renal Disease

Relapse



## HENOCH-SCHONLEIN PURPURA

- ▶ Uncertain of the diagnosis
- ▶ Severe abdominal pain or significant GI bleeding
- ▶ Debilitating Pain

## FIBROMYALGIA

- ▶ A syndrome of chronic pain associated with poor sleep, cognitive dysfunction, and deconditioning

## FIBROMYALGIA-ETIOLOGY

- ▶ Not an inflammatory lesion
- ▶ Thought to be related to a central processing disorder
- ▶ Related to sleep disturbances
- ▶ Some evidence of small fiber disorder



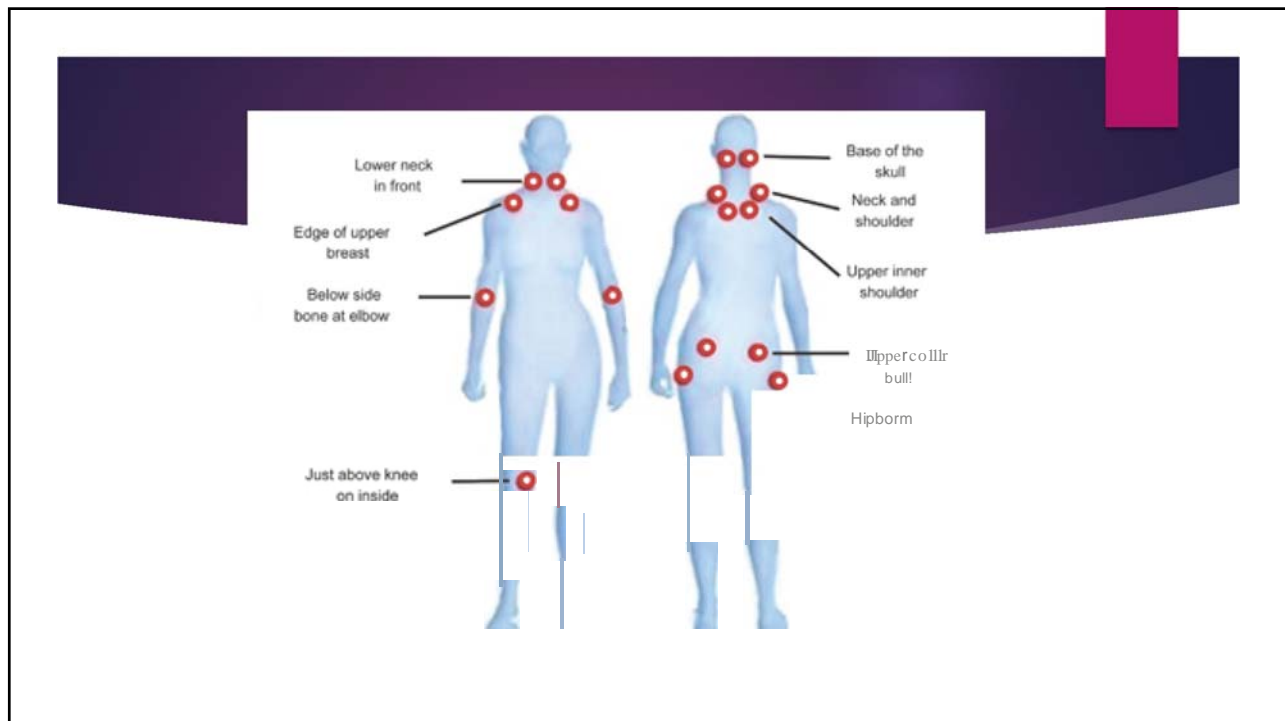
## FIBROMYALGIA-ETIOLOGY

- ▶ Functional scans show completely different brain activity response in those with FM and those without in response to both painful and non painful stimuli
- ▶ Increased sensitivity to "non painful stimuli"
- ▶ Increased Substance P in CSF
- ▶ Evidence of derangement in endogenous opioid response

## FIBROMYALGIA-ETIOLOGY

- ▶ Skin biopsies show decreased numbers of small nerve fibers

Table 1. Criteria for Diagnosing Fibromyalgia	
1990 ACR Criteria <sup>a</sup>	2010 ACR Criteria <sup>a</sup>
<p>1. Widespread Pain for at least three months</p> <p>Above and below the waist</p> <p>Both sides of the body</p> <p>2. Pain on palpation of 11 of 18 tender points</p>	<p>1. Widespread Pain Index (WPI) score ≥ 7 (0-19)</p> <p>19 anatomical locations</p> <ul style="list-style-type: none"> <li>Shoulder: left, right</li> <li>Upper arm: left, right</li> <li>Lower arm: left, right</li> <li>Jaw: left, right</li> <li>Neck</li> <li>Buttock, hip trochanter: left, right</li> <li>Upper leg: left, right</li> <li>Lower leg: left, right</li> <li>Upper back</li> <li>Lower back</li> <li>Chest</li> <li>Abdomen</li> </ul> <p>2. Symptom Severity Scale (SS) (0-12)</p> <p>A. Presence and severity (0-3) of fatigue, cognitive symptoms, waking unrefreshed</p> <p>B. General presence of somatic symptoms</p> <ul style="list-style-type: none"> <li>0: no symptoms</li> <li>1: few symptoms</li> <li>2: a moderate number of symptoms</li> <li>3: a great deal of symptoms</li> </ul> <p>3. Exclusion of other medical conditions that could account for pain</p> <p><sup>a</sup>Symptom severity scale ≥ 9 and WPI ≥ 3 to 6 are also acceptable</p>



Widespread Pain Index  
(1 point per checked box; total = 19 points)

Q) Beside each area on the diagram, indicate if you have had pain or tenderness during the past 7 days in the areas shown below. Check the boxes in the diagram for each area in which you still have had pain or tenderness.

Q) For each symptom listed below, use the following scale to indicate the severity of the symptom during the past 7 days.

- No problem
- Slight or mild problem: generally mild or intermittent
- Moderate problem: considerable problems, often present and/or at a moderate level
- Severe problem: continuous, life-disturbing problems

Point	No problem	Slight or mild problem	Moderate problem	Severe problem
A. Fatigue	0	1	2	3
B. Trouble thinking (forgetting)	0	0	0	0
C. Waking up tired (unrefreshed)	0	0	0	0

Q) During the past 6 months have you had any of the following symptoms?

Points	0	1
A. Pain or cramps in abdomen	<input type="radio"/> No	<input type="radio"/> Yes
B. Irritability	<input type="radio"/> No	<input type="radio"/> Yes
C. Headache	<input type="radio"/> No	<input type="radio"/> Yes

Additional questions (no score):

C) Have the symptoms (questions 2 and 3 and widespread pain) been present at a minimum level for at least 3 months?  
0 No 0 Yes

D) Do you have a disorder that would otherwise explain the pain?  
0 No 0 Yes

## FIBROMYALGIA

- ▶ Not an inflammatory disease
- ▶ Most likely a neuropathic disease involving both CNS and PNS
- ▶ Genetic Predisposition
- ▶ Clearly associated with environmental factors

## EPIDEMIOLOGY

- ▶ Worldwide the prevalence is felt to be between 1.5-2.5%
- ▶ Increases in frequency in age-approx 4% in older patients
- ▶ Prevalence 1% In Pediatric Patients
- ▶ Prevalence is consistent regardless of economic status of the country
- ▶ Prevalence can range between 20-40% of primary care patients
- ▶ Can be seen in 20-40% of Pts with RA, SLE "Secondary Fibromyalgia"
- ▶ Rate also increased with other chronic diseases -CKD, CHF, COPD

## HX

- ▶ Length of time with symptom
- ▶ Disturbed sleep pattern
- ▶ Altered mental state-"FIBRO FOG"
- ▶ Fatigue
- ▶ Absence of joint swelling- sometimes subjective swelling
- ▶ Absence of Fever

## PE

- ▶ Absence of joint effusions
- ▶ May have some enthesitis
- ▶ Poor posture
- ▶ Lack of dermatological findings
- ▶ Normal neurological exam except for subtle sensory findings

## LAB

- ▶ No set of universal labs
- ▶ A reasonable approach
  - CBC
  - CMP
  - TSH
  - ESR/CRP
  - CPK
  - Celiac Disease

## FIBROMYALGIA TX

- ▶ EDUCATION
- ▶ RECONDITIONING/EXERCISE
- ▶ PHARMACOLOGICAL

## EDUCATION

- ▶ "Call it like you see it"
  - Diagnosis is often relieving
  - May be cost effective
  - You have to have buy in to be successful

## EXERCISE/RECONDITIONING

- ▶ Aerobic exercise most effective although strength training may help
- ▶ Start graduated and progressive program

## SLEEP HYGIENE

- ▶ Same sleep/wake time
- ▶ Use an hour wind down time
- ▶ Repeat 20 minutes of wind down time if unable to go sleep
- ▶ If Pt has a bad night go ahead and get up at regular time
- ▶ May nap- no longer than 1 hour
- ▶ Avoid non sex or sleep activities in the bedroom

## MEDICATIONS-What doesn't work

- ▶ Steroids
- ▶ Opioids\*\*
- ▶ NSAIDs

## MEDICATIONS –what does work

- ▶ Nortriptyline/Amitriptyline-helps the pain, also can improve sleep
- ▶ Duloxetine\*
- ▶ Pregabalin\*
- ▶ Milnacipran\*
- ▶ Cyclobenzaprine
- ▶ SSRIs +/- TCAs
- ▶ Tramadol

\* Approved by the FDA for the treatment of Fibromyalgia



## ALTERNATIVE THERAPIES

- ▶ None proven in large studies
- ▶ Some small studies have shown improvement with Tai Chi, Stretching, chiropractic
- ▶ Acupuncture has been disappointing
- ▶ IF not harmful encourage Pt to try

## FOLLOW UP

- ▶ Need to follow closely as relapse is common if Pts do not continue lifestyle changes
- ▶ Trigger point pain is not consistent in determining response
- ▶ Use severity scores, sleep quality, quality of life to determine response
- ▶ May be able to wean pharmacological interventions

## WHEN TO REFER

- ▶ Dx and treatment is clearly in the realm of Primary Care Provider
- ▶ Unclear Diagnoses
- ▶ Complicated medication regimen
- ▶ Often times a single consult with return to PCP can be valuable

## QUESTIONS

?????