Rheumatology pearls for the school nurse

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Overview of pediatric rheumatic diseases

Shoreline, By Moorthy LN

Role of the school nurse
...Interpreter or translator because he or she is “most familiar with the medical and health information” (Selekman & Gamel-McCormick, 2006, p. 640).
...Advocate for the family and child
...Eyes and ears in school for the family and doctors

Childhood arthritis is a leading cause of acquired disability in children

Arthritis Facts
Nearly 300,000 children have arthritis or a related disease.
That’s 1 in 250 kids.
Children with arthritis make an estimated 621,000 visits to health care professionals each year.

A 2007 CDC study found that 10% of all children aged 6 to 17 years had arthritis or a related condition.

Arthritis Facts
Nearly 300,000 children have arthritis or a related disease.
That’s 1 in 250 kids.
Children with arthritis make an estimated 621,000 visits to health care professionals each year.
Some types can interfere with growth.
Some can affect other parts of the body as well, including the skin, eyes and internal organs.
Often immunosuppressed on one or more medications

**Juvenile Idiopathic arthritis types**

- Oligoarticular / Pauciarticular-onset JIA (PaJIA)
- Polyarticular-onset JIA RF+ (PoJIA RF+)
- Polyarticular-onset JIA RF- (PoJIA RF-)
- Systemic-onset JIA (SoJIA)
- Psoriatic arthritis
- Enthesis-related arthritis
- Other arthritis undefined by above mentioned criteria (ILAR classification)

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**Oligo-articular onset JIA ~ Pauciarticular JIA (PaJIA)**

Clinical scenario 1: The limping child with a smile

- 4 year old Caucasian girl comes in limping to your office and she has a swollen knee on exam
- Mother noticed it a couple of weeks ago while giving her a bath- she initially thought her girl was chubby
- Mother recalls she has been limping in the mornings for a few months and has asked to be picked up
**Oligo-articular onset JIA**

- Commonest subtype (> 50%)
- 1-5 years of age
- Girls > boys
- Arthritis in ≤ 4 joints (mostly large joints)
- Treated with NSAIDs, steroid injections, methotrexate and biologics
- Discrepancy in limb lengths
  - Persistent inflammation
  - Orthotic/surgical correction
- Fixed flexion contractures
- PT & OT

**Uveitis**

Silent, chronic, anterior uveitis
- Mostly ANA positive girls (~ 20%)
- Irreversible damage & blindness
- Often require immunosuppressives
- Ophthalmologic monitoring

**Polyarticular JIA (PoJIA)**

*Clinical scenario 2: The girl who is tired, and cannot button her shirt*

- 12 year old girl has been very tired for the last several months
- She walks like grandma in the morning
- Has difficulty buttoning her shirt
- Fingers look puffy
- Teacher says she takes too long to write
Polyarticular JIA (PoJIA)

- 30%-40% of JIA
- Girls > boys
- 2-5 years & 10-14 years
- 5 or more joints in the first 6 months
- AM stiffness
- Increased fatigue
- May have low-grade fever
- Do not appear systemically ill
- Arthritis intermittent or persistent
- Pain with eating due to jaw arthritis
- Neck involvement
- RF+ve or RF-ve
- May lead to disability and contractures
- Short stature (more in children)
- NSAIDS, steroid injections, methotrexate and biologics

Enthesitis-related arthritis/ Spondyloarthropathy

Clinical scenario 3: The boy who limps off the soccer field

- 13 year old boy with foot pain and said that his shoes never fit him quite right.
- Dad says that he limps during and after soccer
- Dad says that he walks like grandpa and is stretching in the mornings. He usually feels better by the time he reaches school
- School is complaining that he is getting up to stretch very often
- You see that he has a swollen achilles and a swollen foot

Spondyloarthropathies

- Ankylosing Spondylitis
- Reactive arthritis
- Psoriatic Arthritis
- IBD associated arthritis
- Undifferentiated spondyloarthropathy
- Enthesitis-related arthritis
Enthesitis-related arthritis

- 10-19% of all pts classified as JIA
- 8-12 yrs, males
- Enthesitis and Arthritis
- Recurrent strains, sprains, heel pains
- Sacroiliac joint tenderness
- Inflammatory spinal pain
- Anterior uveitis associated with pain, redness, or photophobia
- +ve family history of anterior uveitis w/ pain, IBD or spondyloarthropathy
- HLAB27 +ve (90% AS, 60% spondyloarthropathies)

Spondyloarthropathy

- Wide variation in presentation
- Mild enthesitis to polyarthritis and axial involvement
- Psoriasis may not be evident initially
- Family history
- Dactylitis (sausage digits), wrist
- Arthritis mutilans
- Constitutional symptoms
- Uveitis
- Fingernail abnormality
- Use NSAIDS, steroid injections methotrexate, anti-TNF agents and or other biologic medications; do PT and OT

Psoriatic arthritis

- Review in UpToDate, P. Nigrovic, Psoriatic juvenile idiopathic arthritis: Pathogenesis, clinical manifestations, and diagnosis, J Pediatr 2015
Systemic arthritis (systemic juvenile idiopathic arthritis)

- Still's disease
- ~10-20% of JIA diagnoses
- Affects boys & girls equally
- Half of them have onset in childhood
- High spiking quotidian fevers at least 2 weeks, with arthritis in one or more joints
- Often with chills

- This form is now widely thought to be an autoinflammatory condition unrelated to other forms of childhood arthritis and requiring different therapy

Rash of sJIA mimics other infectious processes

- Systemic symptoms
  - Anorexia, arthralgias, hepatosplenomegaly, lymphadenopathy
  - Serositis
- Arthritis occurs concomitantly or later
  - Wrists, knees, ankles, hands, hips, C-spine & TMJ
8 y/o girl with fevers

8 y/o female presented at the time of her initial diagnosis with a 13 day history of fevers, polyarthritis and rash and lymphadenopathy

Course and complications

- Systemic features may persist for 4-6 months with varying degrees of joint involvement
- Monophasic/polyphasic/persistent
- Systemic and/or polyarthritis
- Persistent inflammation &/or chronic destructive arthritis
- Micrognathia, c-spine fusion and destructive hip disease
- Short stature & FTT
- Uveitis - rare
- Amyloidosis - rare in the US
- Macrophage Activation Syndrome
- NSAIDs, Steroids, Methotrexate, Anakinra, Canakinumab, and other biologics

1/5th with disability (Solari)

Growth failure most common extra-articular manifestation (Sarma)

The chief determinants of poor functional ability were younger age at disease onset and a greater restricted joint count (Magnimanzoni)

Motor performance and functional ability (van der Net 2008)

Physical activity (Lelieveld et al)

HRQOL (Oliveira, 2007)

Mental health (Mullick)

Pain, coping, podiatry, GI
Critical Window of Opportunity

Control of Disease Progression Should Start Early to Limit Joint Damage

- Early
- Established
- End Stage

Radiographic damage
- Progressive erosive joint disease
- Impairment of growth & development
- Chronic eye disease
- Psychosocial issues

Advocate early aggressive therapy
- Ophthalmologic monitoring
- Medication monitoring
- PT, OT, nutrition (Promote healthy choices) & counseling

Systemic lupus erythematosus: 1982 classification criteria --4/11

| Malar rash | Renal disorder |
| Distoid rash | Neurologic disorder |
| Photosensitivity | Hematologic disorder |
| Oral ulcers | Immunologic disorder |
| Arthritis | Antinuclear antibody |
| Serositis | |

Systemic lupus erythematosus: 1982 classification criteria
Fauci et al

The girl with fevers and weight loss

11 year old girl is feeling very tired and has a malar rash that is worsening. It is maculopapular erythematous with brownish plaques. She also has some photosensitivity and alopecia. For the last 2 months she has had intermittent fevers, fatigue, weight loss, and irritability.

Treatment: Oral steroids, Hydroxychloroquine
Rash, fatigue and alopecia improved
The girl with tea colored urine

- 14 yo girl has tea-colored urine, low grade fever and edema over face and feet in the past 2 weeks.
- Significant proteinuria
- CBC and diff – NL
- CMP- low albumin 2.5
- ESR 50mm/hr
- C3= 50, C4 = 7, ANA 1:1280, DS-DNA 1:160
- Diagnosed with Class IV SLE nephritis
- Started on Cyclophosphamide

Frequency of nephritis declines with increasing age of onset

[Graph showing frequency decline]

Adult onset SLE vs. Childhood onset SLE

- Malar rash, lymphadenopathy, cytopenias, and nephritis have a greater prevalence in cSLE.

[Table showing cumulative incidence of common SLE manifestations]

Levy et al, The Rheumatologist

Adult onset SLE vs. Childhood onset SLE

- Malar rash, lymphadenopathy, cytopenias, and nephritis have a greater prevalence in cSLE.
Percentage of patients with damage in the particular organ system

Similar data from our cohort also, Moorthy et al. PRYSM 2011

SLE (activity and damage) involvement of systems and Health Related Quality of Life (HRQOL)


Childhood –onset SLE versus adult onset SLE

Higher disease activity at onset and during the disease course (1)
Significant and early damage
Increased exposure to steroids, Longer disease duration
Higher frequency of organ involvement cSLE (1-4)
Cataracts, avascular necrosis, fractures, osteoporosis, low BMD (longer disease vs. steroids), premature atherosclerosis, persistent cognitive dysfunction in cSLE (1-2)
Incurable, potentially devastating disease during a vulnerable period of psychosocial development in cSLE
Increased and unique psychosocial stressors

(1) Brunner et al. (2) Kamphius et al (3) Watson et al (4) Mok et al
Mortality in childhood-onset SLE

Young adults with SLE: SMR 20 times higher than general population (vs. 2-5 fold in adults)

Accumulate disease damage more quickly

More aggressive course → increased exposure to immunosuppressive medications over a longer disease duration

19.5 vs. 16.5 years (p<0.0001)

56% vs. 24% and 21% vs. 8% (p<0.0001)


Predictors for early mortality

Mean age at death 33 (childhood-onset; n=98) versus 52 years (n=859)

Childhood-onset SLE
Education
ESRD

Male sex
Cardiovascular disease
Medicare or Medicaid insurance

Hersh 2010

Approach to management

General
- Routine/flare-worsening
- Team approach
- Adequate rest
- Sun-protection
- Immunizations (Pneumococcal; flu shot)
- Prompt management of infection
- Call doctor prior to any procedure—may need extra antibiotics or steroids
- Appropriate transitioning from pediatric to adult care

- Require medical home
- Reproductive health
- No piercing/tattoos
- Subspecialty visits
- Evaluate mood (patients may get depressed!!!)
- Medication interactions
- Exercise
- VitD and Ca
- Adherence
TREATMENT

- Glucocorticoids
  - Oral prednisone
  - IV methylprednisolone
- Methotrexate
- Hydroxychloroquine
- Rituximab
- Cyclophosphamide
- Azathioprine
- Tacrolimus
- Mycophenolate mofetil
- Benlysta

Juvenile Dermatomyositis

- Malaise, easy fatigue, muscle fatigue, fever & rash
- Insidious – symmetric progressive proximal muscle weakness & pain
- Milestone regression
  - Inability to hold head upright/maintain a sitting posture/stop walking/unable to dress/climb stairs/Gowers
  - Pharyngeal, hypopharyngeal, palatal, esophageal hypomotility - difficulty swallowing, dysphonia, palatal speech, nasal regurgitation - threat of aspiration

Gottron’s papules

Heliotrope rash

Clinical features

- Photosensitivity
- Sun exposures: onset/exacerbation
- Oral ulcers - pain on swallowing
- Vasculitic - eye corners, axillae, pressure points, stretch marks
- Steroids, Methotrexate, Cellcept, Rituximab, IVIG infusions, Azathioprine
Morphea and Linear scleroderma

Mask like-face and digital necrosis

Systemic sclerosis

Contractures of hands and thickening of skin
14 year old c/o cold hands and feet and hands turning pale with cold. She has hand pain also when cold.

Raynauds

- Can occur in different rheumatic diseases or in patients w/o rheumatic diseases
- Conservative approach (transportation, P.E, gloves, socks, etc)
- Calcium channel blockers

Immunizations in the immunosuppressed child

- Avoid live viral vaccines in children receiving systemic immunosuppression
- Encourage them to get killed flu vaccine
- SLE patients – should get the pneumococcal vaccine

Please advise your child's parent to check with your pediatrician before getting any vaccinations.
LIVE VIRAL VACCINES

- Live, attenuated vaccine list:
  - Measles, mumps, rubella (MMR combined vaccine)
  - Varicella (chickenpox)
  - Influenza (nasal spray)
  - Rotavirus

- Live virus vaccines use the weakened (attenuated) form of the virus. The measles, mumps, and rubella (MMR) vaccine and the varicella (chickenpox) vaccine are examples.
- Killed (inactivated) vaccines are made from a protein or other small pieces taken from a virus or bacteria. The flu vaccine is an example.
- Herd immunity is a form of indirect protection from infectious disease that occurs when a large percentage of a population has become immune to an infection, thereby providing a measure of protection for individuals who are not immune.

Fevers, rash or illness

- If the child with a rheumatic disease has a fever, rash, cough, nausea, vomiting, diarrhea, a very severe headache, dizziness, blurry vision, please call the parent and have the child see their pediatrician or rheumatologist.
- Infections and flares should be promptly managed.

Role of the school nurse

<table>
<thead>
<tr>
<th>Growth</th>
<th>Encourage healthy choices for snacks and lunch. Encourage regular activity based on how well the child is feeling.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gastrointestinal</td>
<td>Monitor for nausea/stomach upset as a result of side effects of medications. Monitor weight if on Prednisone for an extended period of time.</td>
</tr>
<tr>
<td>Vision</td>
<td>Encourage compliance with regular ophthalmology visits and eye drop administration if eyes are involved.</td>
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</tbody>
</table>

Eye guard if needed
Medication Side effects

- **NSAIDS** (Motrin, Relafen, Mobic, Diclofenac etc)
  - Liver and kidney function
  - Gastritis (take with food)
- **Antimalarial drugs** (Plaquenil)- Retinal damage
- **Steroids**- Cushingoid appearance, thinning of skin, acne, short stature, suppresses immunity, osteoporosis, diabetes, HTN, avascular necrosis (may need joint replacement), cataracts
  - Gastritis (take with food)
  - Very important to not miss any doses/ or drastically change without speaking to your physician!

Anti TNF agents- adverse events

- Generally effective, safe and well tolerated
  - Injection site reactions, URI symptoms
- **When on one of these agents, infections should be treated early**
- Serious infections (Glucocorticoid likely risk factor – Bukelman et al)
  - Tuberculosis
  - Opportunistic infections (e.g. histoplasmosis, listeriosis, coccidioidomycosis, PCP)
  - Non-opportunistic infections
- Demyelinating events
- Autoantibodies
- ?Malignancies - rate of malignancy higher in JIA compared to general population, Bukelman et al)

Other immunosuppressive medication side effects

Other immunosuppressive agents (Methotrexate, Rituximab, Mycophenolate mofetil, Cyclophosphamide, Tocilizumab, Anakinra, Canakinumab etc)

- Oral ulcers
- Hair loss
- Nausea, vomiting
- Immunosuppression
- Risk of infection
- Liver and kidney function
  - Blood counts
  - Risk for cancer
  - Risk for infertility
  - Affects bladder
Role of the school nurse

• Medications for breakthrough pain/trips

Always remember!

• HIPAA—Health Insurance Portability and Accountability Act of 1996—United States legislation that provides data privacy and security provisions for safeguarding medical information.

• FERPA—The Family Educational Rights and Privacy Act (FERPA)—A federal privacy law that gives parents certain protections with regard to their children's education records, such as report cards, transcripts, disciplinary records, contact and family information, and class schedules.

• School nurse is the member of the health care team!
Accommodations and the Law

• Helping students with the physical challenges of many rheumatic chronic illnesses (emotional, educational etc)

• Section 504 of the Rehabilitation Act of 1973 specifies that no one with a disability can be prevented from participating in federally funded programs – including elementary, secondary and post-secondary education.

• It also offers equal access to the curriculum and accommodations and legal protection for support to achieve that such as extra time to go from one class to another, adaptive physical education or ramp access.

Missing School Due to Illness or Hospitalization

• More than just missing schoolwork
• Missing important contact with friends and classmates.
• Create a plan to help him maintain contact with school friends
• Classmate can deliver missed assignments or get well cards from the class.
• Consider creating an email message board

If fingers, wrists or elbows are involved

- Holding a pencil- built up pencils
- Completing long writing assignments- allow computer use, give extra time
- Carry her schoolbooks- second set of books, roll on backpack, use computer, single binder
If knees or ankles are involved

- Trouble walking to class quickly
- Participating in some activities during P.E.
- Sitting cross-legged on the floor during story time/or prolonged sitting
- Allow child to move around in class

Problems Participating in P.E.

- Do not force your student to participate in any activities that cause pain or are prohibited by her health care provider.
- Do not force certain shoes or dress for P.E.
- Do not exclude her from activities she’s able to do.
- If her joints are painful or stiff, she should know to rest her joints and limit her activity.
- Help her participate in another way such as:
  - tossing a beach ball instead of a baseball
  - walking instead of jogging
  - dancing without jumping instead of jumping rope etc.

Difficulty Walking

- Difficulty getting to class on time
- Excuse your student early enough to get to his next class and avoid crowds in the hallways.
- Try to arrange your student’s classes are in the same area of the school to minimize the distance he has to walk between classes.
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Difficulty getting to class on time

Excuse your student early enough to get to his next class and avoid crowds in the hallways.

Try to arrange your student’s classes are in the same area of the school to minimize the distance he has to walk between classes.
Pain and Stiffness Due to Prolonged Sitting

Please look for ways to get him out of his seat and move around (e.g. Collect HW)

Children with arthritis should be encouraged to move around every 30 to 60 minutes.

Have the whole class stand up and stretch periodically. Everyone will benefit!

Hidden symptoms

- On “bad” days, your student may be-
  - slower
  - uncoordinated,
  - Irritable
    - barely able to walk or raise his or her hand
    - may avoid participating in class activities
    - feel isolated from classmates
- Children may try to ignore or hide their stiffness and pain because they want to be like their classmates.
Difficulty Carrying Heavy Books

- Arrange a locker on each floor
- Keep an extra set of books in the classroom
- Keep extra set to keep at home
- Provide textbooks on DVD
- Post assignments online

How can you help

Children should be allowed to adjust their level of activity.

Please help by watching for warning signs.

Please ask your student to let you know when she’s having a bad day and if there's anything you can do to help her on difficult days.

While arthritis often affects a student’s physical abilities, it’s important to remember that children with arthritis are just like other students in your class.

Having arthritis does not exclude them from having other learning needs or challenges such as learning disabilities or behavioral problems, just like any other student.

Tardiness, Absences (due to stiffness or doctors appointments)

BE AN ADVOCATE!

Accommodations

- Adaptations and equipment for the classroom.
- Examples:
  - Footstool
  - Desktop book holder
  - Floor pillow
  - Computer
  - Special pencil and pen grips
  - Someone take notes for them because writing is difficult.
Guidelines to School Programming

Date:

is followed by ________ for ______. We request the following school recommendations for this student based on the current status of his/her illness:

• [ ] Two sets of books for school and home use
• [ ] Use of elevator
• [ ] Door to door transportation
• [ ] Modified writing assignment and/or use of computer
• [ ] Modified physical education as tolerated
• [ ] Placement of classrooms and lockers in close proximity to minimize walking
• [ ] Allow students extra time to get to next class
• [ ] Rest period
• [ ] Avoidance of demanding classes first thing in the morning due to morning stiffness which usually improves throughout the day

Guidelines to School Programming

Cont’d

• [ ] Allow student to move around in class as needed to prevent inactivity stiffness
• [ ] Child Study Team
• [ ] 504 Plan

Please feel free to call ________ with questions or concerns.

Sincerely

(kgst.org/education)

Emotional Challenges and How You Can Help

• Feelings of isolation
• Whether and how to tell others about her arthritis
• Wanting to be treated like his peers
  – Sensitive to needs
  – Similar expectations
  – Emphasize the student’s strengths instead of weaknesses
• Frustration or fear for the future. Here are some of the more common emotional challenges children with arthritis experience and how you can help.
• Psychological Stress in the Family (finances, work, time, counseling)
Strengthening the school-parent relationship

• Schedule a meeting and keep lines of communication open.
• Designate a point person
• Any special accommodations
• The type of rheumatic condition your student has.
• Medications
  – for breakthrough symptoms
  – compliance
• Make an emergency plan
  – Contact
  – Missed medications
  – Pain
  – Another student coming down with chickenpox
• Upcoming absences (reasons)
• Teachers and substitute teachers

Resources

• http://www.arthritis.org
• Lupus Foundation of America
• National Resource Center on Lupus
• Teens Taking Charge
  Educational Rights Kit – Kids Get Arthritis Too
• Early Childhood and Elementary School Educational Rights
• 504 Plan | Section 504 | 504 Educational Plan | Arthritis Today
• Developing an Effective Plan | Educational Rights Tool Kit
• Juvenile arthritis at school
• Juvenile Idiopathic Arthritis Special Needs Factsheet
• JRA, Part 2 – Treatment, Restriction and Implications for School
• Education and JA: Get the Facts on 504 Plans and Juvenile Arthritis

Pediatric Alliance for Lupus (PAL) Initiative

• Focused on raising awareness and improving diagnosis and treatment of cSLE among racial and ethnic minorities in three geographic locations in NJ—Trenton, Newark-Union, and Edison-New Brunswick
• Seeks to achieve early diagnosis, effective management, and improved health outcomes and health related quality of life for cSLE patients nationwide and in the target areas by improving health literacy and enhancing the medical home for cSLE patients.
THANK YOU
NJAAP team
Patients and their parents
RWJMS team