Welcome to today’s webinar

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Juvenile Arthritis Webinar

Arthritis Foundation

“JIA 101”: An overview of juvenile idiopathic arthritis and its treatment

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What is arthritis?

• Arth-IT IS: joint inflammation
• Joints lie between two bones and allow movement, e.g. a knee
• Common arthritis symptoms:
  – Limping, joint swelling, warmth, pain or stiffness
  – Some children stop participating in activities, seem tired/“lazy”, or even weak
  – Many children are still active despite their arthritis
Causes of Arthritis in Children

• Many causes exist
  – Infection, mechanical, hereditary, or autoimmune disease
  – Lupus, dermatomyositis and other rheumatic diseases can be associated with arthritis
• Juvenile idiopathic arthritis (JIA) is most common chronic cause of arthritis in children

What is Juvenile IDIOPATHIC Arthritis?

• JIA is arthritis lasting >6 weeks beginning in a child <16 years old without a known cause
• JIA is not just one disease:
  – Oligoarticular (arthritis of <4 joints, often associated with uveitis)
  – Polyarticular (arthritis of 5 or more joints)
  – Systemic (fever, rash, and arthritis as well as other organ inflammation)
  – Enthesitis-related arthritis (AKA Spondyloarthritis)
  – Psoriatic arthritis
  – Undifferentiated arthritis

Children with JIA:
Not little adults with rheumatoid arthritis

• May (quietly) modify their behavior, often without complaining
• Growth disturbances can occur: short stature, leg length or small jaw
• Joint contractures, deformity and muscle wasting can occur
• Children are resilient
• Children tolerate medication fairly well
• JIA often doesn’t go away in adulthood
JIA is often associated with eye inflammation

- Uveitis is inflammation of the eye
- It can affect vision
- Uveitis can be symptomatic or asymptomatic
- Some JIA types are associated with high risk for uveitis, so needs frequent eye screening
- Treatment:
  - Steroid eye drops
  - Some may need medicine by mouth or injections
- Uveitis does not always correlate with the arthritis: uveitis can occur WITHOUT active arthritis

All Children with JIA Need Treatment

- Goals:
  - Eliminate active disease (both arthritis and uveitis)
  - Maintain full function
  - Prevent deformity/disability
  - Preserve normal growth and development
- Medications
  - NSAIDs, prednisone (oral and joint injections), DMARDs, Biologics
- Physical/occupational therapy
- Psychological support
- Diet and exercise
- School
- Family, friends, community support
- **GOAL= FULL FUNCTION...GET BACK IN THE GAME!**

NSAIDs

- Non-steroidal anti-inflammatory drugs (NSAIDs) are commonly used first
  - NSAIDs are generally safe and well-tolerated, for many years
    - Most common side effect: mild abdominal pain
  - Need monitoring for gastrointestinal, kidney and liver tests
  - Most NSAIDs have similar effects, but one may work better than another for individual children
  - Helps reduce pain and inflammation
  - NSAIDs may be used as a daily medication or “as-needed” for pain or limp
  - Dosing frequency, available liquid form and taste are important
  - Examples of NSAIDs: Ibuprofen (Advil, Motrin), naproxen (Naproxen, Aleve), and meloxicam (Mobic), indomethacin (Indocin), among many others
Joint injection can be the treatment of choice in Oligo-JIA

- Steroid joint injections of joints are often used if the arthritis persists
- Many children have long-lasting response
- Repeated injections are not associated with joint damage or growth impairment
- Skin thinning and discoloration may be seen (often transient)
- Young children may require anesthesia or sedation

DMARDs

- DMARDs (Disease Modifying Anti-Rheumatic Drug) actually treat the arthritis:
  - Decreases the likelihood of future joint destruction and deformity
- Who needs DMARD treatment?
  - Majority of poly-JIA
  - Some oligo-JIA
  - Some systemic JIA
  - Some patients with uveitis

Methotrexate is the most common DMARD

- Low-dose methotrexate by mouth or injection once a week
- As many as 80% respond well
- Well-tolerated and safe in children (used for more than 30 years)
- Not uncommon to have some mild abdominal discomfort
- Folic acid or leucovorin often prescribed to minimize side effects
- Regular blood test monitoring is needed
  - Liver test abnormalities are not uncommon
  - No cases of irreversible liver disease reported
  - Lung toxicity is rare
- Other DMARDs used:
  - Sulfasalazine, Leflunomide and Hydroxychloroquine
Biologics

- A newer and very effective group of medications
- Targets SPECIFIC molecules that cause inflammation and arthritis
- Can be given alone or with a DMARD
- Side effects include an increased risk of serious infections, especially if previously exposed to tuberculosis or certain fungal infections
- Other risks, such as cancer, are not known with certainty
- IMPORTANT: In adults with RA, cancer risk is not increased compared to RA patients who are not exposed to biologics

What do biologics do?

- Targets depend on the biologic, but can include:
  - Cytokines, which are little proteins that cause inflammation
    - E.g. TNF, IL-1, IL-6
  - Cells with certain molecules on their surface
    - Cells with CD-20 marker on their surface (B cells, which make harmful antibodies)
  - Communication between cells
    - Preventing B cells from activating T cells, which can cause/worsen arthritis

Anti-TNF biologics

- “Oldest” biologic medications:
  - Infliximab (Remicade)
  - Etanercept (Enbrel)
  - Adalimumab (Humira)
- Certolizumab (Cimzia) and golimumab (Simponi) have not yet been FDA approved in children
- Differences in how it’s given:
  - Infliximab is given as an IV infusion every month
  - Etanercept and adalimumab are given as a subcutaneous injection every 1-2 weeks
- Differences in treatment effects:
  - Infliximab and adalimumab are also effective in treating uveitis (not etanercept)
Other Biologics for JIA

- **Abatacept (Orencia)**
  - Blocks T cell activation which triggers inflammation
  - Monthly IV infusion
- **Tocilizumab (Actemra)**
  - Blocks IL-6, a key inflammatory cytokine
  - Given every 4 weeks in poly-JIA
- **Rituximab (Rituxan)**
  - Destroys B cells, which make antibodies (especially effective for JIA patients that are RF positive)
  - Given IV, sometimes every 6 months

**Blocking IL-1 and IL-6: effective for Systemic JIA**

- **IL-1 and IL-6 are elevated in systemic JIA**
- **Anti-IL-1 and IL-6 biologics:**
  - May be more appropriate, and can be given prior to DMARDs and prednisone in systemic JIA
  - May still be effective in patients who have failed methotrexate AND anti-TNF therapy
  - May result in less steroid use
- **Current biologics that target IL-1**
  - Anakinra (Kineret)-daily injection
  - Canakinumab (Ilaris)-injection every 4 weeks
- **Current biologics that target IL-6**
  - Tocilizumab (Actemra)-IV infusion every 2-4 weeks

**In addition to medication...**

- **Physical/occupational therapy**
  - Home exercise program, splinting of joints if necessary, pain management
- **Psychology**
  - Pain management, body image issues, peer/sibling relationships, risk taking behavior
- **Diet and exercise**
  - Prednisone often causes weight gain, patients often deconditioned due to illness
- **Regular school attendance always preferred**
  - Home schooling is often inadequate, and does not allow opportunity to interact with peers
- **Family, friends and community support**

**GOAL= FULL FUNCTION...GET BACK IN THE GAME!**
More research is needed to identify better treatments

• The good news:
  – We now have very effective treatments!

• The bad news:
  – We still have a lot to learn: which are the best treatments, which are the safest, which should be started first, how long to continue them, when to stop, etc.

• We need you and your child to help answer these questions!

Research 101:
Pediatric Rheumatology Research Update

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Childhood Arthritis & Rheumatology Research Alliance (CARRA)

JIA: 50 or more years ago
JA conferences 1990 to early 2000’s

JA Camps, 2014

Changes in Treatments for JIA
We Need More Pediatric Rheumatology Research

- Many urgent questions to ask:
  - How safe are these medications?
  - Which medications are the best for which disease?
  - For which particular patient?
  - When is it safe to stop medications?
- But research is challenging:
  - Rare diseases = relatively few patients
  - Information from research in adults is of limited use:
  - Kids are not just “small adults”!

Research Networks Help to Solve the Problem

- Patients with the same disease can be studied together in larger numbers
- Promotes a “culture” of research across all the sites
- Care of patients are improved though large clinical research studies: we learn from each other
- Treatments can be standardized to improve patient outcomes
- Gives all children the opportunity to participate in research

Lessons from Pediatric Oncology

![Annual US Cancer Mortality Rate Children <15 and Pediatric Cancer Cooperative Groups](chart.png)
CARRA’s Mission and Vision

• Mission ...  
  to improve the health, well-being and outcomes of children and adolescents with rheumatic disease through fostering and facilitating collaborative research in prevention, treatment and cure.

• Vision...  
  to identify the most effective and safe treatments for children with rheumatic diseases by enhancing and promoting pediatric rheumatology research in North America through collaborations between investigators, funding partners, and patients, making research participation available to all affected children and their families.

CARRA Membership

• >400 members at >100 sites in Canada and US  
• Includes >95% of pediatric rheumatologists in North America
What has CARRA Achieved?

- 4 large NIH sponsored clinical trials
- 23+ active research studies
- Successfully developed registry using NIH funding – the CARRA "Legacy Registry"
- A new CARRA Registry will begin in 2015:
  - Designed to help evaluate the long term safety of new biologic medications
  - Be a powerful foundation for innovative research in pediatric rheumatology

The CARRA Legacy Registry: Power in Numbers!

- JIA: 4
- Lupus: 27
- JDM: 12
- MCTD: 29
- Vasculitis: 1
- Uveitis: 11
- FM: 1
- Scleroderma: 1
- Sarcoidosis: 5
- Sjogren Syndrome: 1

Total: >9500

CARRA Registry: JIA patient exposure to biologics

- tocilizumab: 4
- rituximab: 27
- abatacept: 12
- infliximab: 326
- golimumab: 1150
- etanercept: 1
- apratuzumab: 1
- certolizumab: 5
- canulimumab: 4
- belimumab: 138
- adalimumab: 578
- abatacept: 237
The NEW CARRA Registry: Safety Studies
- Major goal: understanding safety of medications in real life
  - Enroll all patients regardless of what medications are taken
- Plan to enroll 10,000 or more children/adolescents
- Long term follow-up
  - 10+ years
  - Into adulthood
- CARRA partnership with pediatric rheumatologists, industry, patients, and government

The New CARRA Registry: Comparing Effectiveness of Treatments
- We now have many effective treatments!
- But how do we know which are the best treatments?
- CARRA has standardized the treatments for some diseases
- By collecting information about how patients do on standardized treatments through the CARRA Registry:
  - We can learn how treatments compare
  - What the best treatments for a disease should be
  - How treatments affect the lives of children and their families
- We can also learn even more by collecting blood and other samples
  - Understand the mechanisms underlying diseases
  - Markers that predict response, or remission, or cure

PARTNERS:
PPRN (Patient Powered Research Network)
- Patients, Advocates and Rheumatology Teams Network for Research and Service (PARTNERS) Consortium
- Funded by PCORI (Patient Centered Outcomes Research Institute)
  - Mission: A community of patients and healthcare professionals carrying out research on childhood rheumatic diseases with the patient’s voice at the center.
  - Vision: Improving the lives of children with rheumatic diseases through research that matters to YOU.
CARRA Registry Vision

Multicenter Clinical Trials
Translational studies (understanding disease mechanisms)
Comparative Effectiveness Research
Observational/natural history studies
Quality improvement
Sample repository

CARRA Registry—foundation for many studies going forward

Research 101: Summary

- CARRA is rapidly moving pediatric rheumatology research forward in North America
- CARRA is collaborating with partners around the world
- CARRA and the Arthritis Foundation together will find answers
- Research will allow your children to have healthier futures
- We need to hear from YOU!
  - Participate in research
  - Join/volunteer with the Arthritis Foundation
  - Be a voice that will help shape the research we do together

School Success

Making school a positive experience.... for everyone!

Doreen Tabussi, BSN, RN, CPN
Students with chronic health conditions might be scared about school

• Will the teachers know anything about my disease?
• Will my friends understand why I can’t do everything they can?
• What if I can’t keep-up with my assignments?
• What if I miss too many days of school?

Parents may feel the same way

• Will they think my child is dumb or lazy?
• What if my child is in pain at school and they don’t believe it?
• Who will help in my absence?

It can also be scary for educators & school healthcare professionals

• We have enough sick kids here. What help will this kid need?
• How am I going to know when this kid really has a problem?
• We’re short-staffed. I don’t have any extra time to offer.
How will the school know accommodations needed for a student to achieve academically?

Ask yourself and your child questions:

- Will attendance be affected?
- Is medication needed at school?
- Is additional hydration required?
- Are breaks possible?
- Who will help with ADLs?
- Does your building have stairs? An elevator?

Once you have gathered all of the information

It is time to formulate a plan
Who’s involved in formulating an appropriate accommodations plan?

- Family
- Medical team
- School Educators, Administrators, Medical staff, and Child Study Team; if needed.

Where to start

- Meet or speak with the school. See what accommodations could reasonably be provided.
- If an agreement is reached, inform everyone of the plan.
- If the school wants recommendations in writing, ask about setting-up a 504 plan.

If the school can easily meet needs:

- It is preferable to have open communication with school.
- Write to teachers, guidance counselors, and the school nurse to inform them about your or your child’s disease.
- Explain limitations, how condition changes will be communicated, and ask that each teacher receives a copy of communications. Staff often travel from different schools to teach.
What is a 504 plan?

• The American With Disabilities Act, under section 504, provides legal protection for students in federally-funded education programs.
• It allows disabled students to participate in school with well-abled peers.
• To qualify, a child needs to have one or more major life activities impacted by disease or illness.
• Some students do fine with a medical plan.

504 plans may include:

• Any necessary accommodations to help a student succeed in school, except for services that are provided through special education.
• Extra books for home use
• Modified physical education; No penalty when student is unable to participate.
• Permission to carry water bottle at school
• Ability to get up & stretch or go to nurse’s office or rest room without special permission.

Individualized Education Plan (IEP)

• May be written for a child with a chronic or acute illness under the term Other Health Impaired (OHI).
• Child must have a health problem that affects their vitality, strength or alertness.
• Educational performance is impacted secondary to health status, and the child needs special education resources.
If special education services are needed, the student must be evaluated.

• Evaluation is requested by a child’s teacher or in-writing by a parent or guardian.
• If a teacher requests an evaluation, parental consent must be obtained.
• Once a formal request and consent is completed, the school has 90 days to conduct a multi-disciplinary evaluation, and if eligible for services, develop and implement an IEP for the student.

Some of the people who are involved in the IEP evaluation process include:

• Teachers and educators
• The school’s administrative team
• Case Manager
• Guidance counselor
• Psychologist
• Physical & Occupational Therapist

What types of tests are included in an IEP evaluation?

• Vision and hearing screen
• Behavioral observation and scoring
• Social history intake, including medical records
• Individual intelligence and achievement test
• Assessment of communication skills
• Physical examination
• Learning process screens; auditory/visual
• Motor development and vocational assessment
What will be included in an IEP

- Long and short-term goals to be accomplished by the student.
- Goals are specific to the child’s needs and evaluated on an on-going basis.
- IEPs are written for a 3-year period, but may be modified, if necessary.
- Every 3 years, the child study team meets with parents/parents & child/or child.
- Plans are updated, continued or discontinued as warranted.

What happens when schools do not comply with 504/IEP plans?

Conservatively:
- Parents intervene and speak with teachers not complying with plan
- Primary medical team speaks with school to ask for assistance in enforcing plan
- Family asks for advocate to approach school on their behalf

Maximum Impact:
- Family files a complaint against the school system through the Department of Education
- Family pursues law suit or files federal complaint for violations related to Individuals with Disabilities Education Act

Academic success

- The road may seem long, but your healthcare team and the AF will help you.
- School success is a joint venture; achieving success depends as much on you, as it does on the school, and us!
Federal Education Office

• Office of Special Education and Rehabilitative Services
U.S. Department of Education
400 Maryland Avenue, S.W.
Washington, D.C. 20202-7100

• www.ed.gov/about/offices/list/osers/osep
• (202) 245-7468 (voice/TTY)

Juvenile Arthritis Programs

Family Day
JA Conference
Parent2Parent Network
ARTHRITIS FOUNDATION
JA Camps NY, NJ, EPA
Walk to CURE ARTHRITIS

To learn more: Visit www.kidsgetarthritis.org
Your VOICE Counts!

Become an E-advocate!
Get e-alerts on arthritis related legislation and ways you can make your voice count.
To learn more, visit:
www.arthritis.org/advocate/join-the-movement

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