Ultrasound in Rheumatological Conditions: Status and Perspectives

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SPR 2016 Pediatric Ultrasound Course
Orlando, Florida
No disclosures
Goals

Briefly review pathophysiology of JIA
Review US techniques to optimize imaging & interpretation
Identify the appearance of normal MSK structures in children
Discuss the current role of US in JIA
Future directions of imaging
Juvenile Idiopathic Arthritis

Most common chronic rheumatologic disease of childhood

**Definition:**

“Arthritis that begins before age 16y, continues for approximately 6 weeks and involves 1 or multiple joints”

1/1,000 children in North America

Juvenile Idiopathic Arthritis

Historically, two main classification systems:
  European – Juvenile Chronic Arthritis (JCA)
  American – Juvenile Rheumatoid Arthritis (JRA)

ILAR (International League of Associations for Rheumatology)
  1995; Juvenile Idiopathic Arthritis (JIA)
    Classification updated in 2004
    7 major subtypes – ”umbrella term”

### 7 Major Subtypes

<table>
<thead>
<tr>
<th>Subtype</th>
<th>Frequency</th>
<th>Findings</th>
<th>Sex Ratio</th>
</tr>
</thead>
<tbody>
<tr>
<td>Systemic Arthritis</td>
<td>4-17%</td>
<td>Systemic symptoms (fever, rash, organ)</td>
<td>F=M</td>
</tr>
<tr>
<td>Oligoarthritis</td>
<td>27-56%</td>
<td>Up to 4 joints in 6 mo</td>
<td>F&gt;&gt;&gt;M</td>
</tr>
<tr>
<td>RF + Polyarthritis</td>
<td>2-7%</td>
<td>5 or more joints</td>
<td>F&gt;&gt;M</td>
</tr>
<tr>
<td>RF – Polyarthritis</td>
<td>11-15%</td>
<td>5 or more joints</td>
<td>F&gt;&gt;M</td>
</tr>
<tr>
<td>Enthesitis-Related Arthritis</td>
<td>15-20%</td>
<td>Insertion sites of tendons and ligaments &amp; axial disease</td>
<td>M&gt;&gt;F</td>
</tr>
<tr>
<td>Psoriatic arthritis</td>
<td>2-11%</td>
<td>Typical skin and nail findings</td>
<td>F&gt;M</td>
</tr>
<tr>
<td>Undifferentiated arthritis</td>
<td>11-15%</td>
<td></td>
<td></td>
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JIA Pathophysiology

*Hallmark:* Synovial thickening & inflammation

Role of Imaging

• JIA diagnosis based on clinical & laboratory findings
• Imaging may detect early manifestations of arthritis
  • Synovitis, cartilage abnormalities, erosions
• Evaluate the extent of disease/joint involvement
• Monitor disease progression & treatment response

*Imaging can more accurately distinguish between arthritis and tenosynovitis than clinical examination alone and in some cases, it can help define the subtype of JIA*

*Provides opportunity to implement therapy at an early stage*
Role of Imaging

Radiographs – traditional method of evaluating joints
  Poor sensitivity in depicting active arthritis
  Rarely show erosive disease until late in disease course

Increase use of US & MRI

*Ultrasound has a higher sensitivity and specificity for the presence of synovitis than clinical assessment along and is the most sensitive method for detection of tenosynovitis*

Imaging Obstacles in Children

- Sparsity of normative data
  - To correctly diagnose pathology, need to be familiar with normal age-dependent changes of the growing skeleton
  - Large amount of data in adults – apply to children?

- Unique features of the growing skeleton
  - Physiologic features of recently ossified bones can be misinterpreted as cortical erosions
  - Appearance, thickness & vascularity of the epiphyseal cartilage varies with skeletal maturation

- Suboptimal reader agreement on validation studies

Ultrasound

• Rapid & inexpensive evaluation
• Survey multiple joints – *advantage over MRI*
• No sedation or exposure to radiation
• Uses:
  • *Synovial proliferation, joint fluid, cartilage thickness, erosions, tenosynovitis*
• Color & Power Doppler – synovial vascularity & hyperemia
• US contrast agents may further improve evaluation of synovial hyperemia

US Limitations

• No validated US scoring scales for JIA
• Highly operator dependent and lack of normative data
• Unable to assess for bone marrow edema
• Deeper portions of joints are unassessable by ultrasound
  • TMJ, sacroiliac joints
  • Poor at evaluating subtalar disease\textsuperscript{2,3}

\textit{US is not reliable in the assessment of active TMJ arthritis & MRI should be performed}\textsuperscript{1}

• No established imaging protocols

\textsuperscript{1} Muller et al. \textit{Rheumatology (Oxford)}. Jun 2009;48(6):680-685.
## Protocol for JIA

<table>
<thead>
<tr>
<th>Institution</th>
<th>Ultrasound Approach</th>
<th>Established Protocols</th>
<th>Radiology Specialization</th>
<th>Rheumatology POC US</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>Targeted US exams when recommended by radiologist in rheum-rad case conference</td>
<td>Yes</td>
<td>Yes*</td>
<td>No</td>
</tr>
<tr>
<td>B</td>
<td>Targeted US exams</td>
<td>In progress</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>C</td>
<td>Typically MRI – but for multi-joint US screening &amp; targeted US exams</td>
<td>No</td>
<td>Yes</td>
<td>Beginning phase</td>
</tr>
<tr>
<td>D</td>
<td>US for looking for an effusion</td>
<td>No</td>
<td>Yes</td>
<td>Not sure</td>
</tr>
<tr>
<td>E</td>
<td>Targeted US exams</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>F</td>
<td>Targeted US exams</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>G</td>
<td>Targeted US exams</td>
<td>No</td>
<td>No**</td>
<td>Yes</td>
</tr>
<tr>
<td>H</td>
<td>Mostly MR, few targeted US exams</td>
<td>No</td>
<td>Yes</td>
<td>Not sure</td>
</tr>
</tbody>
</table>

*1 radiologist in group to do US exams
**A few radiologists like to do US exams
Imaging Techniques

Good contact between the transducer and skin:
Use of coupling gel or standoff pad

Achilles tendon

Stand-off pad
Water Bath Techniques

- Poor contact with curved contours of the hands and feet
- Compression of clinically relevant superficial structures
  - Patient discomfort caused by transducer contact

Balloon Technique

Right third toe Long
Knob-ology

Make use of machine presets

Focal zones – point of interest

Achilles Tendon
Keger's Fat
Calcaneus
Contralateral Side for Comparison

Helps differentiate normal from abnormal

Particularly useful in pediatric population with changing appearance of the growing skeleton

Be cautious of bilateral pathology, especially in subclinical disease wrist, PIP, subtalar and foot joints

“Enthesis Organ”

Components:

Tendon
Various portions of fibrocartilage
Sub-tendinous bursae
Fat

Insertion of the fibers into the bone through the fibrocartilage

Weiss PF, Chauvin NA, Roth J. Current Rheumatology Reports, In press.
Tendons

Well defined structure. Echogenic compared to the adjacent fat and muscle.

Longitudinal: Parallel lines, densely packed fibrillary pattern

Transverse: “Fiber cable wire” appearance
Enthesis

Surrounded by a connective tissue called *paratenon* – elastic sleeve for the tendon - *Lined on its inner surface by synovial cells*

Other tendons have synovial sheaths - parietal and visceral sheets - lined by synovial cells

Anisotropy

Property of all tendons (direction dependent)

Occurs when the US beam is not at 90 degrees

Demonstrates false hypoechogenicity within the tendon

“Heel to Toe” maneuver
Two Imaging Planes

Quadriceps Tendon

Longitudinal

Transverse
Common Flexor Tendon

Elbow, 5 year-old girl
14 year old girl. There is a thin rim of cartilage (*) in the region of the plantar fascia insertion.
Lateral Elbow – Normal PDI

Longitudinal PDI of the common extensor tendon in a healthy 5-year old
The OMERACT Ultrasound Working Group 10 Years On: Update at OMERACT 12

George A. Bruyn, Esperanza Naredo, Annamaria Iagnocco, Peter V. Balint, Marina Backhaus, Frederique Gandjbakhch, Marwin Gutierrez, Andrew Filer, Stephanie Finzel, Kei Ikeda, Gurjit S. Kaeley, Silvia Magni Manzoni, Sarah Ohrndorf, Carlos Pineda, Bethan Richards, Johannes Roth, Wolfgang A. Schmidt, Lene Terslev, and Maria Antonietta D’Agostino, on behalf of the OMERACT Ultrasound Task Force

**Abstract.** Musculoskeletal ultrasound (US) now thrives as an established imaging modality for the investigation and management of chronic inflammatory arthritis. We summarize here results of the Outcome Measures in Rheumatology (OMERACT) US working group (WG) projects of the last 2 years. These results were reported at the OMERACT 12 meeting at the plenary session and discussed during breakout sessions. Topics included standardization of US use in rheumatic disease over the last decade and its contribution to understanding musculoskeletal diseases. This is the first update report of WG activities in validating US as an outcome measure in musculoskeletal inflammatory and degenerative diseases, including pediatric arthritis, since the OMERACT 11 meeting. (First Release March 15 2015; J Rheumatol 2015;42:2172–6; doi:10.3899/jrheum.141462)
Joint effusion – OMERACT definitions

Abnormal hypoechoic or anechoic (relative to subdermal fat), or in some cases isoechoic or hyperechoic, intra-articular material that is displaceable and compressible but does not demonstrate Doppler signal.

**Physiologic fluid** is common in children and it can be difficult distinguishing between normal amounts of joint fluid from a joint effusion.

Ankle
Anterior ankle; at the dome of the talus
Elbow

“Crab position”

Hum
Cap
Olecranon
Elbow Effusion

Anterior Fat Pad Sign
Synovial thickening—OMERACT definitions

Solid, non-compressible, abnormally hypoechoic tissue associated with joint lines or surrounding tendons.

Can be difficult to discern from adjacent hypoechoic epiphyseal cartilage.

Images courtesy of Monica Epelman, MD
Synovial thickening may not represent ongoing disease – Doppler ultrasound to depict increased synovial blood flow

Healthy children may exhibit some Doppler signal due to physiologically enhanced blood flow

....need for better knowledge of ‘normal’

6–year old girl with pauciarticular JIA and knee pain. Transverse images of suprapatellar region.
Ultrasound - Synovitis

US assessment of disease activity can be more informative than clinical examination

Subclinical synovitis is frequently observed by US, particularly within the hands and feet

A semi-quantitative system for grading synovial thickening is used in adult rheumatology but no such system has been validated in JIA
Bursitis

15-year old girl with JIA and shoulder pain
Tenosynovitis - OMERACT definitions

Hypoechoic or anechoic thickened tissue with or without fluid in the tendon sheath which may or may not exhibit Doppler signal

In children, tenosynovitis is most commonly seen around the ankle joint and along the extensor tendons of the wrist

12-year old boy with JIA and medial left ankle swelling
Enthesitis—OMERACT definitions

Inflammation at the tendinous or ligamentous insertion - abnormally hypoechoic foci and/or thickened tendon or ligament at its bony attachment that is seen in two perpendicular planes

May exhibit abnormal Doppler signal and/or bony changes such as enthesophytes, erosions or cortical irregularities, however these features are less commonly seen in children

Within the tendon, perientheseseal and cartilage


Weiss PF, Chauvin NA, Roth J. Current Rheumatology Reports, In press.
Enthesitis

Lateral epicondyle

Longitudinal image of the CET in a 13 year old girl

Low flow settings with a low PFR and low WF

Gain adjusted to allow maximum sensitivity without creating artifacts
Ultrasound - Enthesitis

US can detect subclinical enthesitis

Doppler-US revealed enthesitis in 50% of clinically normal entheses


Standard dolorimeter examination for the detection of enthesitis in children with ERA and found that dolorimeter testing had poor accuracy and reliability, as compared with power Doppler US


Prognostic significance of subclinical inflammation still needs to be determined
Bone erosion is defined as discontinuity of the bone surface visible in two perpendicular planes.

Assessment of erosive changes in children is challenging - physiologic irregularities in recently ossified bone that can be misinterpreted as cortical erosions, highlighting the need for further knowledge of normal bone anatomy throughout pediatric age groups.

Ultrasound - Cartilage

Age and gender normal US reference standards for cartilage thickness of the knee, ankle, wrist, MCP and PIP joints in children


Further validated by demonstrating good agreement between MRI & US for measurement of cartilage thickness in healthy children

Ultrasound - Cartilage

It has been shown that patients with JIA have reduced cartilage thickness when compared to age- and gender-matched controls, although interestingly, this is observed in both clinically affected and non-affected joints.


Work in progress...
Nail Disease - Psoriasis

Long, DIP joint in affected nail disease

NP – nail plate, DP – distal phalanx – DIPJ – distal interphalangeal joint

Current Status

• No defined imaging recommendations for JIA
  • OMERACT – Outcome Measures in Rheumatology
  • Health-e-Child Radiology Group

• Timing & utilization of imaging in JIA is tailored to the individual patient

• Established adult protocols and standardized scoring systems must be modified before adopted for children

Large, long-term innovative research must be established by collaboration with rheumatologists & radiologists

Lingering Questions

• Which US findings are preferred for establishing a definite diagnosis?
• Can ultrasound predict/evaluate remission?
• How frequently do US scans have to be repeated?
• Which joints need to be screened?
• What is a sufficient protocol?
Ultrasound - Summary

US is a powerful tool that may allow differentiation between synovial, tendinous and entheseal inflammation

Prognostic significance of subclinical information depicted by US still needs to be determined

Subclinical disease may potentially alert the physician towards more aggressive treatment & close monitoring of the patient
THANK YOU!

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