Scoring and Grading B-Mode Synovitis and Doppler findings in pediatric MSKUS

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Pathology - Definition Synovitis

• Synovitis on ultrasonography in children B-mode and Doppler

• Depending on the joint, synovitis can be diagnosed on the basis of B-mode abnormalities alone

• Abnormal B-mode includes synovial effusion or synovial hypertrophy
Pathology - Definition Synovitis ctd.

- Synovial effusion is defined as abnormal, intraarticular, an- or hypoechoic fluid that is displaceable
- Synovial Hypertrophy is defined as abnormal, intraarticular, hypoechoic material that is non-displaceable
- Abnormal Doppler signals within an area of synovial hypertrophy
- Physiologic Doppler signals can be present in any area of the joint
Scores

B-Mode and Doppler

Rationale:

1) Serial clinical assessment and comparability

2) Research
How to report?

1) Descriptive – “Effusion in the tibiotalar joint with increased Doppler”, measure?

2) Qualitative – Mild, moderate, severe

3) Semiquantitative Scoring

4) Pixel count or other (RI)?
Challenges

• Variability in equipment

• Solution: Ensure optimization of settings, especially frequency and PRF

• Do not use standardized settings !

• But: pay great attention to low flow settings and frequency
Ultrasound Scoring Systems
Semi-quantitative US-Score

4 step semi quantitative grading system (Grad 0 – 3) separately for:

- Joint effusion
- Synovial hypertrophy
- Bone erosions
- Power Doppler activity

of 5 selected joints unilateral: MCP II, III, PIP II and MTP I and II from dorsal

Interobserver Kappa agreement 0.48 – 0.68

Semi-quantitative Synovitis – Score

Grade 0: no elevation of joint capsule, no synovitis
Grade 1: slight elevation of joint capsule, mild synovitis, parallel to bone line
Grade 2: parallel elevation of joint capsule to skin, moderate synovitis
Grade 3: convex elevation of joint capsule severe synovitis

Sum scores of synovitis of MCP II – V and PIP II – V
Assess dorsal and volar

Scheel AK, Backhaus M et al., Arthritis Rheum 2005: 52; 733-43
Pathology – Scores Adults B-Mode

Semi-quantitative GS-Synovitis-Score at the MCP-joints II/III in dorsal long axis

Grade 0: normal

Grade 1: mild synovitis (*)

Grade 2: moderate synovitis

Grade 3: severe synovitis (*)

Scheel AK, Backhaus M et al., Arthritis Rheum 2005: 52; 733-43
Pathology – Scores Adults Doppler

Semi-quantitative PD-Synovitis Score at wrist in dorso-median long axis

Grade 0: no signal / no blood flow

Grade 1: single signal / mild blood flow

Grade 2: more single and confluent signals / moderate blood flow, <50% of intra-articular area

Grade 3: confluent signals / severe blood flow, >50% of intra-articular area

Scheel AK, Backhaus M et al., Arthritis Rheum 2005: 52; 733-43
Composite score

Grade 0 or normal: no synovial hypertrophy, no Doppler signal

Grade 1 or minimal:
minimal synovial hypertrophy, with (or without) no more than grade 1 Doppler signal

Grade 2 or moderate:
moderate synovial hypertrophy with no more than grade 2 Doppler signal
minimal synovial hypertrophy and grade 2 Doppler signal

Grade 3 or severe:
severe synovial hypertrophy with or without Doppler signal
minimal/moderate synovial hypertrophy and grade 3 Doppler signal

How many joints to assess?

US Scores 7 – 12 – 28 – 44 – 78 joints

High correlation coefficients between all these scores for grey scale and Power Doppler

Hammer-H et al ACR 2011
Which joints to assess?

Loeuille D AR 2006 Frequency of Synovitis in RA
62 RA patients

PD positive synovitis:

Wrist > MCP2 > MCP3 > MTP2 > MTP3 > MTP5 > MCP5

Arctic Group ACR 2016
using the proposed selection of 7 joints from Backhaus er dataset, we analyzed data from Naredo and colleagues by order to evaluate the applicability of the 7-joint count in another the global synovitis scores of 50% (7 of 14) of the articles. In metatarsophalangeal (MTP)-2, and MTP-5] were included in Naredo, to present good validity issues: the 12-joint count proposed by logistic regression model counts are presented in Table 4. The use of the 7-joint count in Comparative results on responsiveness by using the 2 joint ly Joints selected in the proposed 12-joint count are wrist, MCP2, MCP3, PIP2, PIP3, MTP2, MTP5 (bilateral)

Table 4. Evaluation of responsiveness of the 7-joint score developed by Backhaus, et al25 in an additional dataset obtained from Naredo, et al26. Data for this analysis is courtesy of Marina Backhaus and Esperanza Naredo.

<table>
<thead>
<tr>
<th>Joint Count</th>
<th>Gray-scale Synovitis</th>
<th>Power Doppler Activity</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean decrease‡ (95% CI)</td>
<td>SRM</td>
</tr>
<tr>
<td>Wrist, MCP2, MCP3, knee ankle, elbow (bilateral)*</td>
<td>2.5 (2.0–2.9)</td>
<td>0.925</td>
</tr>
<tr>
<td>Simplified 12-joint PDUS model (12 joints 24 recesses)†</td>
<td>2.3 (1.9–2.8)</td>
<td>0.881</td>
</tr>
<tr>
<td>Wrist, MCP2, MCP3, PIP2, PIP3, MTP2, MTP5 (unilateral right)‡</td>
<td>1.5 (1.2–1.7)</td>
<td>0.842</td>
</tr>
<tr>
<td>Wrist, MCP2, MCP3, PIP2, PIP3, MTP2, MTP5 (unilateral left)‡</td>
<td>1.3 (1.0–1.6)</td>
<td>0.732</td>
</tr>
<tr>
<td>Wrist, MCP2, MCP3, PIP2, PIP3, MTP2, MTP5 (bilateral)†</td>
<td>2.8 (2.3–3.4)</td>
<td>0.890</td>
</tr>
<tr>
<td>44 joints‡‡</td>
<td>7.4 (6.1–8.8)</td>
<td>0.934</td>
</tr>
</tbody>
</table>

Conclusion: Small and at least one large joint bilateral
Challenges

• Scores differ in the views they include, MCP dorsal vs volar

• Does the scoring system work for all views?

• Does the scoring system work for large joints?

• 4 views are summarized for the MCP resulting in a max score of 12 compared to one view for the knee resulting in a max score of 3

• No images are kept, Kaeley et al AR 2016 significant reader drift
Backhaus Large Joint Score
BMC Musculoskeletal Disorders 2013

GSUS
Synovitis by GSUS analyzed semiquantitatively from 0 to 3
(0 = absence, 1 = mild, 2 = moderate, 3 = severe)
Grade 1 small abnormal hypoechoic/anechoic line beneath capsule.
Grade 2 joint capsule elevated parallel to the joint area.
Grade 3 strong convex distension of the joint.

PDUS
Grade 0 = no intraarticular color signal,
Grade 1 = up to 3 color signals representing only low flow
Grade 2 = greater than grade 1 to < 50% of the intraarticular area
Grade 3 = > 50% of the intraarticular area filled with color signals.
<table>
<thead>
<tr>
<th>Region</th>
<th>Plane</th>
<th>Grade 0</th>
<th>Grade 1</th>
<th>Grade 2</th>
<th>Grade 3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shoulder</td>
<td>Dorsal transverse</td>
<td>Normal</td>
<td>Effusion/synovitis in external rotation only</td>
<td>Effusion/synovitis in external and internal rotation</td>
<td>Remarkable convex JCD</td>
</tr>
<tr>
<td>Shoulder</td>
<td>Axillary longitudinal</td>
<td>Normal</td>
<td>Joint capsule distension (JCD) concave</td>
<td>JCD straight</td>
<td>JCD convex</td>
</tr>
<tr>
<td>Elbow</td>
<td>Humero-radial</td>
<td>Normal</td>
<td>JCD parallel to the humerus</td>
<td>JCD straight</td>
<td>JCD convex</td>
</tr>
<tr>
<td>Elbow</td>
<td>Humerulnar</td>
<td>Normal</td>
<td>JCD parallel to the humerus</td>
<td>JCD straight</td>
<td>JCD convex</td>
</tr>
<tr>
<td>Elbow</td>
<td>Olecranon fossa</td>
<td>Normal</td>
<td>Olecranon fossa partially filled</td>
<td>Fossa olecrani completely filled</td>
<td>JCD convex above the olecranon fossae</td>
</tr>
<tr>
<td>Hip</td>
<td>Anterior longitudinal</td>
<td>Normal</td>
<td>JCD concave</td>
<td>JCD straight</td>
<td>JCD convex</td>
</tr>
<tr>
<td>Knee</td>
<td>Suprapatellar longitudinal</td>
<td>Normal</td>
<td>JCD parallel to the humerus</td>
<td>JCD straight</td>
<td>JCD convex</td>
</tr>
<tr>
<td>Knee</td>
<td>Medial/lateral longitudinal</td>
<td>Normal</td>
<td>JCD parallel to bone no distension over the joint space</td>
<td>JCD parallel to bone distension above the joint space</td>
<td>JCD convex above the joint space</td>
</tr>
<tr>
<td>Knee</td>
<td>Posterior</td>
<td>Normal</td>
<td>Slight JCD over the jointspace</td>
<td>JCD parallel to bone, distension over the joint space</td>
<td>JCD convex above the joint space</td>
</tr>
</tbody>
</table>
Where Do We Look for Synovitis?

Recesses are lax capsular areas where fluid and synovial hypertrophy accumulate. Favorite joints and PD sensitivity:

MCP : Dorsal > Volar

PIP : Volar > Dorsal

Wrist: Dorsal

Elbow: Lateral and Annular Recess > Posterior > Anterior

Knee: Parapatellar > Parameniscal >> Suprapatellar

MTP – Dorsal recesses contain small amounts of fluid normally
Synovitis MCP
Grade 2 or 3?
Within recess or synovial hypertrophy?
Which color should the Pixels be in?
Kaeley et al – Tocilizumab study

Blue color for Power Doppler

Detailed scoring system B-mode and Doppler
Scores Pediatric

reduced joint power Doppler US (PDUS) score

validity, feasibility, reliability and sensitivity to change compared with a 44 joints PDUS assessment in JIA

42 children with active JIA requiring modified therapy

greyscale (GS) synovitis and power Doppler signal
4-point semiquantitative scale

calculation of US composite indices and US composite joint counts

Collado-P et al
Rheumatology 2013
PedSynS 4-point semiquantitative scale of GS synovitis

0  absence, normal joint recess

1  mild, synovitis filling the joint recess between periarticular epiphyses change from angle-shaped to a plateau-shaped recess

2  moderate, convex shape of the joint recess without extension over the bone diaphysis

3  marked, convex shape of the joint recess with extension to at least one of the bone diaphyses

Collado-P et al
Rheumatology 2013
The problem:

0 absence, normal joint recess

1 mild, synovitis filling the joint recess between periarticular epiphyses, change from angle-shaped to a plateau-shaped recess

2 moderate, convex shape of the joint recess without extension over the bone diaphysis

3 marked, convex shape of the joint recess with extension to at least one of the bone diaphyses
Score suggestion

- Grade 0 = normal

- Grade 1 = mild, synovitis filling the joint recess between periarticular epiphyses that leads to a change from the angle-shaped recess to a plateau-shaped recess

- Grade 2 = convex shape of the recess without extension to the bone diaphysis nor to the whole length of a short bone

- Grade 3 = convex shape of the recess with extension to the bone diaphysis or the whole length of a short bone
Power Doppler Score

0 absence, no synovial flow

1 mild, single-vessel signal

2 moderate, confluent vessels

3 marked, vessel signals in more than half of the Intraarticular area

Tenosynovitis and bursitis were scored using a dichotomous assessment (0, absence; 1, presence) in both GS and PD scoring systems

Collado-P et al
Rheumatology 2013
Current Pediatric Projects

1) Omeract ongoing – global scoring system

2) PRES

3) PANLAR – enthesis

4) CARRA – joint by joint

5) Collaborative projects using 12 joint score
<table>
<thead>
<tr>
<th>Joint</th>
<th>Imaging specification</th>
<th>Total Number of Images</th>
</tr>
</thead>
<tbody>
<tr>
<td>Elbow</td>
<td>• A sagittal scan over the humeroulnar, humeroradial and dorsal elbow joint will be done.</td>
<td>• 3 images in B-Mode and 3 images in Doppler-Mode, total 6 images.</td>
</tr>
</tbody>
</table>
| Wrist   | • A sagittal scan over the dorsal aspect of the medial and ulnar part of the wrist will be done and the radiocarpal, midcarpal as well as the carpometacarpal joints will be evaluated.  
  • The radioulnar joint will be evaluated as well in an additional scan.  
  • A dorsal transverse scan of the wrist will be done to evaluate the extensor tendon sheaths | • 4 images in B-Mode and 4 images Doppler-Mode, total 8 images. |
| Fingers | • The subject will be in a sitting position with the hands palm-side down or dorsum-side down (depending on whether dorsal or volar assessment is done) in a neutral position on an examination table.  
  • A sagittal scan over the dorsal and volar aspect of the MCP 2 and 3 joint will be done. In addition a transverse scan on the dorsal and volar aspect will be done for MCP 2 and 3. | • 8 images in B-Mode and 8 images Doppler-Mode, total 16 images |
| Knee    | • The subject will be in a supine position with the lower extremities parallel to each other. A measurement will be taken in 30 degrees flexed position.  
  • The measurement will be done sagittally over the suprapatellar recess, with the probe positioned cranial to the superior edge of the patella.  
  • Subsequently a medial and lateral parapatellar scan will be done. | • 3 images in B-Mode and 3 images Doppler-Mode, total 6 images |
| Ankle   | • The subject will be in supine position with the knee in 90 degrees flexion.  
  • A sagittal scan over the medial aspect of the tibiotalar joint will be done. In addition in 3 transverse scans the tendon sheath and cross sectional area of the Tibialis anterior, Extensor Hallucis Longus, Extensor Digitorum at the level of the talar cartilage, the Tibialis Posterior, Flexor Digitorum, Flexor Hallucis Longus at the level of the sustentaculum and the Peroneus Tendons at the level of the supramalleolar and trochlea (where they separate) will be assessed.  
  • In a separate medial and lateral scan the subtalar joint will be assessed.  
  • Finally the talonavicular joint will be assessed in a sagittal scan. | • 7 images in B-Mode and 7 images Doppler-Mode, total 14 images |
Grade 1 – mild

Distension of synovial recess originating from the radiocarpal or midcarpal joint or both with the superior border of the recess remaining parallel to the respective bone line and filling less than 50% of the potential space below the beta line.

Grade 2 – moderate

Distension of synovial recess originating from the radiocarpal or midcarpal joint or both with the superior border of the recess convex and filling 50% or more of the potential space below the beta line and remaining deep to the beta line. Distension of the recess may be present in the radiocarpal joint only, in the intercarpal joint only, or in both joints.

Grade 3 – severe

Distension of synovial recess originating from the radiocarpal or midcarpal joint or both with the superior border of the recess convex and extending above the beta line.
Scoring Doppler

Grade 0
No Doppler signals except for feeding vessels crossing synovial recesses. A feeding vessel is defined as a single vessel showing continuity in entering the bone/cartilage

Grade 1
Single Signals (up to three)

Grade 2
Confluent signals in less than 50 % of synovial hypertrophy/hyperplasia

Grade 3
Confluent signals in more than 50 % of synovial hypertrophy/hyperplasia
B-mode

Grade 0

The parapatellar recesses are empty. A minimal bulge of synovium may be found at the patellofemoral joint line.

Grade 1

Synovium fills the proximal part of the parapatellar recess
**Grade 2**

Synovium fills part of the parapatellar recess or extends throughout the recess filling <50% of its expected full volume.

**Grade 3**

Synovium occupies >50% of the expected full volume of the recess
Huge variability in scores

B-mode vs Doppler, Doppler in area of abnormal B-mode ?

Tendon, Enthesis, Bursae, other structures

Inclusion of parameters of damage ?

Various sum scores weighing parameters differently
Scoring Enthesis Adults - Option

**Thickness**

<1 mm of increase exceeding the threshold of normal values grade 1

1 mm or greater but less than 2 mm of increase grade 2

2 mm or greater grade 3

**Erosions**

max diameter (grade 1, >0 mm but <2 mm; grade 2, ≥2 mm and <3 mm; grade 3, ≥3 mm)

**Hypoechogenicity, enthesophytes, calcifications and bursa** semiquantitative scores (mild changes: grade 1, moderate changes: grade 2 and severe changes: grade 3).

**Intraentheseal Doppler signals**

0: no Doppler signal, 1: mild (≤ 2 punctiform Doppler signals with no confluent Doppler signal), 2: moderate (2–4 punctiform Doppler signal or 1 confluent Doppler signal), 3: marked (> 4 punctiform Doppler signals or > 1 confluent Doppler signal).
Pediatric Enthesis

• No consensus yet

• No need to include parameters of damage (Weiss & Roth 2016)

• Focus on Doppler signals in area of B-mode abnormality

• Important in light of physiologic Doppler in Enthesis

• Scoring system: Semiquantitative scoring for Doppler signals
Tusentakk