Preliminary Definitions for Sacroiliac Joint Pathologies in the OMERACT Juvenile Idiopathic Arthritis Magnetic Resonance Imaging Score (OMERACT JAMRIS-SIJ)


ABSTRACT. Objective. To develop definitions for the assessment of magnetic resonance imaging (MRI) pathologies of the sacroiliac joints (SIJ) in juvenile idiopathic arthritis.

Methods. An Outcome Measures in Rheumatology (OMERACT) consensus-driven methodology consisting of iterative surveys and focus group meetings within an international group of rheumatologists and radiologists.

Results. Two domains, inflammation and structural, were identified. Definitions for bone marrow edema, joint space inflammation, capsulitis, and enthesis were derived for joint inflammation; sclerosis, erosion, fatty lesion, and ankylosis were defined for assessing structural joint changes.

Conclusion. Preliminary consensus-driven definitions for inflammation and structural elements have been derived, underpinning the ongoing development of the OMERACT Juvenile Arthritis MRI SIJ scoring system (OMERACT JAMRIS-SIJ). (First Release May 15 2019; J Rheumatol 2019;46:1192–7; doi:10.3899/jrheum.181115)

Key Indexing Terms:
OMERACT MAGNETIC RESONANCE IMAGING JUVENILE IDIOPATHIC ARTHRITIS OUTCOME ASSESSMENT SACROILIITIS
Juvenile idiopathic arthritis (JIA) is a chronic inflammatory disease that affects joint function, resulting in significant effects on quality of life by limiting physical and psychosocial integration. JIA has an estimated worldwide prevalence of 0.07 to 4.01 cases and an incidence rate of 0.008 to 0.226 cases per 1000 children per year, respectively, highlighting the global disease burden and the need for new clinical trials for JIA management.

Magnetic resonance imaging (MRI) has been reported to be sensitive in detecting early inflammatory changes in the sacroiliac joints (SIJ) in JIA long before radiography, thus allowing for intervention before the onset of irreversible joint damage. Clinical assessment of the SIJ is limited by its location and anatomy. The quantification of interval change of pediatric SIJ using an MRI-based scoring method may serve as an important objective outcome measure. The new JIA Clinical Core Set recently endorsed at the OMERACT 2018 conference includes joint inflammation and damage domains as variables in the middle circle, recommending these as important measurement domains in JIA clinical trials. Improving these outcome measures calls for an international, multidisciplinary effort toward development and validation of suitable and specific measurement instruments to quantify disease and assess treatment effects in JIA.

A recent systematic review by Swami, et al of existing MRI-based SIJ scoring systems reported the lack of established MRI instruments for assessment of SIJ in the pediatric population. Conversely, SIJ MRI scoring systems are well developed and validated in adults. Current attempts to extrapolate adult SIJ MRI scoring systems into the pediatric group reported significant feasibility and reliability, but the construct validity remains a limitation, although clinical assessment of disease activity and damage also remains problematic in the pediatric population. Given the lack of standards for normality and pathology in growing SIJ, there is an urgent need for the development and validation of a pediatric-specific MRI scoring system for SIJ. We used the Outcome Measures in Rheumatology (OMERACT) consensus-driven methodology for iteratively developing definitions as a prelude to the development of an MRI-based scoring system for the assessment of SIJ in JIA, and as a first step we aimed to decide on the main domains and elementary pathology definitions of the MRI-based scoring system.

**MATERIALS AND METHODS**

**Conceptual framework of the OMERACT JAMRIS-SIJ.** The OMERACT JAMRI Working Group developed the Juvenile Arthritis MRI SIJ scoring system (JAMRIS-SIJ) as an outcome measure for the quantification of interval changes in the SIJ of children and adolescents with JIA in effectiveness trials. The components of this newly developed scoring system should identify and define the measured construct, and they need not correlate with each other. Thus, the conceptual framework of the construct and measurement components are in keeping with a formative model. This model together with the OMERACT Filter 2.0 shall guide the instrument’s validation framework and specify the use and interpretation of the JAMRIS-SIJ in the context of other concurrently used outcome measures in clinical trials.

**Measurement domains of the OMERACT JAMRIS-SIJ.** After a working group discussion and review of the existing literature, joint inflammation and structural change were selected because they measured/evaluated separate measurement constructs for the assessment of active and chronic SIJ disease in JIA. This separation is in keeping with known pathophysiology and disease history and follows from the domains in adult SIJ and other inflammatory arthritis scoring systems that have shown to be valid and feasible in the adult population.

**Developing definitions.** An anonymous survey (https://goo.gl/forms/ehnV5bs2WPv62xt32) containing multiple choice, dichotomous, and open-ended questions was developed to select the constructs, component definitions, MRI data acquisition protocol, and scoring method of an ideal pediatric SIJ MRI scoring system. The survey questions and component definitions were developed based on published adult SIJ MRI scoring systems. Respondents provided suggestions to enrich the components and component definitions template, applying relevant content expertise.

The survey response was analyzed and presented to the OMERACT JAMRI Special Interest Group meeting during the Radiological Society of North America scientific conference in November 2017 in Chicago, Illinois, USA, for refinement of definitions and consensus upon final definitions. In May 2018, the preliminary JAMRIS-SIJ was presented to a broader group of experts at the OMERACT meeting in Terrigal, Australia, for additional input. In addition, a Steering Committee constituted of members from the adult SIJ study group was formed to align pediatric definitions with existing definitions for the adult OMERACT SIJ MRI scale. After 2 iterations of video conferences held in August and September 2018, final deliberations and consensus of items and definitions of the JAMRIS-SIJ were reached. Each item definition was voted for agreement among the members of OMERACT JAMRI Working Group as summarized in Table 1. A consensus was deemed to have been achieved if > 70% agreement was reached among the respondents.
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consideration” for all definitions and a caveat for the inter-
complexities and pitfalls of interpreting the growing/maturing
SIJ, the group decided to include a statement of “overarching
orating centers. Participating experts voluntarily elected to
surveys, and research agreements were made among collab-

MRI acquired using a dedicated acquisition protocol and
lesion components without the use of contrast for acquisition
of MRI sequences. This allows for the avoidance of unnec-
cessary use of contrast in pediatric imaging because of
concerns of potential brain accumulations and nephrogenic
ystemic fibrosis17. Twenty-eight international multidisci-

sions that occur because
of physiologic maturity unique to sex and age 18,19,20.
articular cartilage and bone marrow development are an ongoing
process in children, resulting in changes that occur because
of physiologic maturity unique to sex and age18,19,20.
Transformation between hematopoietic and fatty marrow
occurs with advancing age, with earlier skeletal maturation
noted in girls than in boys19,21.

RESULTS
Our study reported an all-inclusive definition of SIJ MRI
lesion components without the use of contrast for acquisition
of MRI sequences. This allows for the avoidance of unnec-

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DISCUSSION
Our study aimed at facilitating the development of a
pediatric-specific MRI scoring system for measuring disease
activity and structural joint changes in the SIJ of JIA. The
study identified components that encompass both constructs,
active joint inflammation and structural joint changes in JIA,
through an iterative consensus process that allowed for both
anonymized and in-person discussions and consensus quan-
tification. The experts agreed that the preliminary
JAMRIS-SIJ definitions have an acceptable face and content
validity, allowing for a comprehensive measurement of the
underlying construct14. Data-driven assessment of the lesion
definitions is to follow.

However, as stated above, the aim of this process is to
develop an outcome measure, not a diagnostic tool. The
working group noted that there is a need to define an internal
comparator to differentiate variations because of MRI acquisi-
tion and inter-subject variation. These can be used
whenever available as normal references for bone marrow
pretation of bone marrow edema in Table 1. These contextual
considerations require pediatric-specific experience in image
interpretation. The working group acknowledges that
cartilage and bone marrow development are an ongoing
process in children, resulting in changes that occur because
of physiologic maturity unique to sex and age18,19,20.
Transformation between hematopoietic and fatty marrow
occurs with advancing age, with earlier skeletal maturation
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Table 1. Consensus definitions of components of the JAMRIS-SIJ.

<table>
<thead>
<tr>
<th>Feature</th>
<th>Definition</th>
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<tbody>
<tr>
<td><strong>Inflammation MRI components</strong></td>
<td></td>
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<tr>
<td>BME</td>
<td>An ill-defined area of high bone marrow signal intensity† on fluid-sensitive sequences within the subchondral bone of the ilium or sacrum</td>
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<tr>
<td>Joint space inflammation</td>
<td>Increased signal intensity on fluid sensitive and/or contrast-enhanced T1-weighted images within the joint space of the cartilaginous portion of the SIJ</td>
</tr>
<tr>
<td>Capsulitis</td>
<td>Increased signal intensity on fluid sensitive and/or contrast-enhanced T1-weighted images involving the SIJ capsule</td>
</tr>
<tr>
<td>Enthesitis</td>
<td>Increased signal intensity within the bone marrow and/or adjacent soft tissue on fluid sensitive and/or contrast-enhanced T1-weighted images at sites where ligaments and tendons attach to a bone</td>
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<tr>
<td><strong>Structural MRI components</strong></td>
<td></td>
</tr>
<tr>
<td>Sclerosis</td>
<td>A substantially wider than normal (of ≥ 5 mm in adolescents) area of low subarticular bone signal on T1-weighted and fluid sensitive images</td>
</tr>
<tr>
<td>Erosion</td>
<td>Bony defect (or irregularity with associated BME, sclerosis, or fatty lesion) at the osteochondral interface involving the bone contour and presenting with low signal intensity on T1-weighted and fluid-sensitive images</td>
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<tr>
<td>Fatty lesion</td>
<td>Homogeneous increased signal intensity within the subchondral bone on T1-weighted non-FS image presenting with a distinct border of the lesion</td>
</tr>
<tr>
<td>Ankylosis</td>
<td>Presence of signal equivalent to regional bone marrow continuously bridging a portion of the joint space between the iliac and sacral bones</td>
</tr>
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</table>

Statement of overarching consideration for all definitions: “ […] in comparison to physiological changes normally seen on MRI examinations of age- and sex-matched children, and visible in 2 planes wherever available.” † Caveat for BME: “[…] compared to the signal intensity of the iliac crest, edges of the vertebrae, and triradiate cartilage wherever available.” JAMRIS-SIJ: Juvenile Arthritis Magnetic Resonance Image Sacroiliac Joint Scoring System; MRI: magnetic resonance imaging; BME: bone marrow edema; SIJ: sacroiliac joint; FS: fat suppressed.
signal: the sacral interforaminal area, the iliac crest, ischial pubic synchondrosis, and triradiate cartilage.\textsuperscript{11}

Notwithstanding the consensus obtained for lesion definitions among the expert group, there were areas of disagreement. The definition of erosion remains a contentious topic, because erosions may be solitary or in association with...
other lesions. Also, topics such as presence of erosive changes in association with differences in skeletal maturation according to sex and other common pitfalls should be reconsidered by the expert group in the future to improve the usability of the JAMRIS-SIJ. The development of an atlas for MRI of SIJ in the future should enable definition of thresholds between normality and abnormality of SIJ in the pediatric population. Further studies are required to define the minimum, ideal, and recommended data acquisition protocol, measurement scale, and methods for the OMERACT JAMRIS-SIJ.

We have reported the lesion component definitions for a preliminary pediatric MRI SIJ scoring system according to OMERACT standards — the OMERACT JAMRIS-SIJ — for the assessment of inflammatory and structural joint changes in JIA. The results of this consensus-driven methodologic and technical survey conducted among a multidisciplinary international expert group are the first steps toward the development of the OMERACT JAMRIS-SIJ scoring system. Further assessment is needed, including derivation of lesion scoring method and component score, determination of MRI data acquisition protocol, exercises to systematically evaluate reliability, validity, responsiveness, and feasibility of the lesion definitions.

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REFERENCES

APPENDIX 1. Summary of the methodology and consensus framework for the development of an MRI-based scoring system including its constructs, components, and the consensus process for component definition. MRI: magnetic resonance imaging; SIJ: sacroiliac joint; BME: bone marrow edema; JAMRIS-SIJ: Juvenile Arthritis MRI SIJ scoring system.