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ORIGINAL ARTICLE

High frequency power doppler ultrasonography in oligoarticular juvenile idiopathic arthritis: Correlation with disease severity

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KEYWORDS

Juvenile idiopathic arthritis; High resolution power Doppler ultrasonography; Disease severity score; Juvenile arthritis functionality scale

Abstract *Aim of the work:* Juvenile idiopathic arthritis (JIA) is the most common chronic rheumatic disease in children and is a leading cause of acquired disability in the pediatric age. We aimed to detect subclinical synovitis in children with oligoarticular JIA using high frequency power Doppler ultrasonography (US) and correlate US scores with clinical disease severity.

Patients and methods: High frequency power Doppler ultrasonography was done for both knees and ankles of 20 oligoarticular JIA patients. Assessment of clinical disease severity by articular index scores and functional assessment by the modified juvenile arthritis functionality scale (JAFS) were calculated for all patients.

Results: We found 24 clinically active knees and 20 clinically active ankles. On comparison between clinical and ultrasonography activity, we found 13 clinically inactive joints, but they were active joints by ultrasonography. There was a highly significant difference ($p < 0.001$) between clinical and US activity as regards the number of affected joints. There was a highly significant ($p < 0.01$) positive correlation between clinical disease severity scores and all US scores except clinical score of range of motion (ROM); that did not show a significant positive correlation with US score of effusion ($p > 0.05$).

Conclusions: High frequency power Doppler US may be useful in detecting subclinical synovitis of joints in JIA patients with clinically defined inactive joints. Also, it can indicate the degree of disease severity in JIA patients. So, high frequency power Doppler US can be used in standard clinical practice for monitoring JIA patients for better assessment and management of the disease.

1. Introduction

The term juvenile idiopathic arthritis (JIA) encompasses a clinically heterogeneous group of disorders characterized by arthritis that begins before 16 years of age, persists for more than 6 weeks and is of unknown cause [1]. JIA is the most

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common chronic rheumatic disease in children and is a leading cause of acquired disability in the pediatric age. It is an important cause of short and long-term disability [1]. The persistence of synovial inflammation may cause cartilage and bone damage and, ultimately, lead to permanent alterations in joint structures and serious impairment of physical function [2].

The International League of Associations for Rheumatology (ILAR) has subclassified the disease entity into seven distinct categories, based on the number of affected joints and the presence of particular extraarticular manifestations. They are systemic, oligoarticular (persistent and extended), polyarticular (rheumatoid factor negative), polyarticular (rheumatoid factor positive), psoriatic, enthesitis-related and undifferentiated [3].

Musculoskeletal ultrasound (MSUS) has emerged as an indispensable tool for physicians involved in musculoskeletal medicine, and lately it has become more attractive to pediatric rheumatologists as well. Two important aspects have resulted in increased interest in using MSUS in JIA: firstly, the evolution of high frequency linear transducers that depict superficial musculoskeletal structures with unsurpassed resolution. Secondly, the need for imaging techniques that can detect the slightest traces of soft tissue inflammation. So far, only few studies have investigated power Doppler assessments of children with JIA [4–9].

In general, US has gained an important position in rheumatologists' clinical practice. This is due to its intrinsic characteristics, such as low costs, rapidity and lack of radiation [10]. For these reasons, US could be a valuable tool in the assessment of JIA, as it is a simple examination that can offer a significant amount of information on a large number of joints in a relatively short time and with minimal discomfort to the patient. It is easy to perform on children of all ages, because agitation of the patient is rarely a problem [11,12].

Ultrasound may demonstrate a greater sensitivity in identifying joint synovitis than clinical examination alone. This is of major importance, as clinical classification and therapeutic strategy in JIA are guided by the number of joints involved [1]. So, In JIA, US assessment of disease activity may be more informative than clinical examination [13]. Ultrasound images are analyzed in real time, and the information that is acquired can be used directly to adjust the clinical assessment, which can be particularly useful if there are few verbal complaints, e.g., in infants [14]. Subclinical synovitis is frequently detected by US, particularly in the hands and feet [15]. The prognostic significance of such subclinical inflammation still needs to be determined [11].

The aim of this study was to detect subclinical synovitis (hypertrophied vascular synovium) in children with oligoarticular JIA using high frequency Doppler US as a new imaging microvascular sensitive method of disease prediction, then correlate the US scores with clinical disease severity.

2. Patients and methods

Twenty patients with oligoarticular JIA were selected from those presenting to the Pediatric Rheumatology outpatient clinics of the Ain Shams University Hospitals. Those patients were all fulfilling the ILAR classification. They presented with arthritis of 4 or fewer joints during the first 6 months of disease. Patients were further classified as either persistent (if no more than four joints are affected during the disease course) or extended (if, after the initial 6 month period, the total number of affected joints exceeds four) [3]. The included patients had arthritis in knees and/or ankles.

We excluded patients with age more than 16 years old, other six types of JIA according to ILAR, systemic connective tissue disease: systemic lupus erythematosus, vasculitis, growing pain, trauma of joints or bones, infective arthritis, reactive arthritis, hemophilic arthropathy, tumor of joints or bones, avascular necrosis, Perthes' disease and osteochondritis. The study protocol was in accordance with Helsinki declaration of human rights, and was approved by the local Ethics Committee. The written informed consent from all the patients' parents was obtained.

All patients were subjected to the following:

1. *Full history taking and thorough physical examination* including detailed musculoskeletal examination.
2. *Assessment of disease severity by articular index (severity score)* according to Kakati et al. [16]: Both knees and ankles of all patients were assessed for swelling, tenderness/pain on motion and restricted motion. Clinically active arthritis was defined as swelling or limitation of movement with either pain upon movement or tenderness. *Swelling* was scored as 0 = none; 1 = mild, definite swelling but with no blurring of normal skeletal outlines; 2 = moderate, definite obscuring of skeletal landmarks; 3 = severe, no discernible skeletal landmarks; *Tenderness and/or pain on motion* was scored as 0 = none; 1 = mild, patient complains on joint movement or palpation; 2 = moderate, patient withdraws or changes facial expression on movement or palpation; 3 = severe, patient responds severely to movement or palpation); *Restricted motion* was scored as 0 = full range; 1 = 1–25% limitation; 2 = 26–50% limitation; 3 = 51–75% limitation; 4 = 76–100% limitation. In overall, severity score is calculated to each joint as the sum of the severity ratings obtained for the scores of pain on motion/tenderness, swelling and limited range of motion; severity score to each joint ranged from 0 to 10 and to all joints from 0 to 40.
3. *Functional assessment:* The parent is asked to assess a child's functional ability by completing the modified juvenile arthritis functionality scale (JAFS) [17]. It is a 5-item questionnaire (for lower limbs only) in which the ability of the child to perform each task is scored as follows: 0 = without difficulty, 1 = with difficulty and 2 = unable to do. It is ranged from 0 to 10.
4. *High frequency ultrasonographic and power Doppler assessment:* By 12 MHz probe Esaote MyLab in Al Demerdash hospital, is performed on both knees and ankles of all patients, which are scanned for the presence of synovial hyperplasia, joint effusion, and power Doppler (PD) signal. All of the US findings were interpreted using both longitudinal and transverse planes [18]. Synovial hyperplasia was graded as follows: 0 = < 2 mm, 1 = 2–5 mm, 2 = 6–8 mm, and 3 = > 8 mm. Joint effusion was graded as follows: 0 = absent, 1 = < 5 mm, 2 = 5–10 mm, and 3 = > 10 mm.Power Doppler signal was graded as follows: 0 = absent, 1 = presence of single/vessel dots, 2 = presence of confluent vessel dots in less than half of the synovial area, and 3 = presence of confluent vessel dots in more than half of the synovial area. Ultrasound active joint scores ranged from 1 to 9, except if 1 was for PD alone, then it was considered to be inactive [13]. Ultrasound