The Arthritis of Familial Mediterranean Fever

To The Editor:

We read with great interest the article by Lidar, et al\(^1\) on arthritis as the sole episodic manifestation of familial Mediterranean fever (FMF). We would emphasize an additional feature that may be considered in the variables characterizing FMF related arthritis, namely the synovial fluid cell count. We recently reviewed the articular manifestations of 74 patients with FMF seen at the American University of Beirut Medical Center over an 18-year period\(^2\), and reported the case of a young Lebanese woman who also presented with recurrent episodes of left knee and calf swelling as the sole manifestation of FMF\(^3\). The diagnosis was intriguing initially because the synovial fluid leukocyte count was suggestive of septic arthritis [172,000 cells/mm\(^3\), 94% polymorphonuclear (PMN)]; however, bacteriologic cultures were negative. FMF was suspected in view of a positive family history and genetic analysis for the mutations in the pyrin/marenostrin (MEFV) gene revealing a homozygote mutation at methionine-694-valine. The arthritis was controlled with prophylactic colchicine therapy.

In the earliest reviews on articular manifestations of FMF, Heller, et al\(^4\) reported that synovial fluid leukocyte counts ranged from 200/mm\(^3\) to 1,000,000/mm\(^3\) with high predominance of PMN\(^5\). This elevated leukocyte count usually suggests infection, and patients are often subjected to prolonged antibiotic therapy despite negative cultures\(^6\).

Our case supports the conclusion of Lidar, et al\(^1\) on the importance of considering FMF in the differential diagnosis of acute monoarthritis in young adults, keeping in mind that this arthritis may be associated with an elevated synovial fluid leukocyte count in regions with high incidence of FMF.

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REFERENCES


Dr. Lidar and Dr. Livneh reply

To the Editor:

We agree with Dr. Uthman’s comment concerning the very high leukocyte count that occasionally may be seen in the sterile synovial fluid drawn from an active joint of a patient with FMF. Others have reported a comparable observation, although only scantily\(^1,2\), and in this regard Dr. Uthman’s contribution is important. High white blood cell counts in sterile synovitis, however, are not specific for FMF and may occur in other monoarthritides, e.g., Behcet’s disease, gout, reactive arthritis, etc\(^3,4\). We were unable to evaluate this feature for sensitivity and specificity due to incomplete data in the files of the patients and controls. However, we believe that the criteria established are valid in spite of the absence of white blood cell counts in joint fluid. For instance, the arthritis in the patient reported by Dr. Uthman could have been assigned to FMF long before genetic and synovial cell analyses were obtained, because it complies with the criteria of FMF solo arthritis defined in our work\(^5\).

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REFERENCES


Can Hand Assessments Designed for Persons with Scleroderma Be Valid for Persons with Rheumatoid Arthritis?

To the Editor:

Two recently developed assessments have been designed for persons with scleroderma. One, the UK Scleroderma Functional Score (UKFS) or Scleroderma Functional Assessment Questionnaire (Table 1), is a self-report of functional ability\(^1\) and has been shown to be reliable and valid for persons with scleroderma\(^1\). The other assessment is the Hand Mobility in Scleroderma Test (HAMIS)\(^2\). The HAMIS is a performance test of joint motion (Table 1) and has also been shown to be reliable and valid for per-
sons with scleroderma. Since rheumatoid arthritis (RA) results in deformi-
ties of the hand, as does scleroderma, we wondered if the SFQ and the
HAMIS were valid assessments to measure hand function and joint motion
in persons with RA. We compared SFQ and HAMIS scores to other self-
reports and performance based tests of hand function and range of motion
in persons with RA.

A convenience sample of 40 subjects who had been diagnosed with RA
according to the American College of Rheumatology criteria\textsuperscript{4} participated
in this study. The mean age of subjects was 49.5 years (range 22–76 yrs),
with a mean disease duration of 13.1 years (range 1–42 yrs). Subjects consis-
ted of 34 women and 6 men. Thirty-eight subjects were right-handed,
one was left handed, and one reported being ambidextrous.

Subjects were administered the following assessments: the UKFS\textsuperscript{1}, the
HAMIS\textsuperscript{4,5}, the Duruöz Hand Index (DHI; also referred to as the Cochin Scale),\textsuperscript{7} the Health Assessment Questionnaire (HAQ),\textsuperscript{8} the Arthritis Hand Function Test (AHFT),\textsuperscript{9} and the Keital Functional Test (KFT).\textsuperscript{10} The concepts measured, question structure, and possible range of scores for each
assessment are shown in Table 1. The UKFS, DHI, and HAQ are all self-
reports, while the HAMIS, AHFT, and KFT are performance based tests.

Table 1 also shows the means and standard deviations for the 40 sub-
jects on the UKFS, HAMIS, DHI, HAQ, AHFT, and KFT. Spearman rho
correlation coefficients were calculated to examine the concurrent validity
of the UKFS and HAMIS with the DHI, HAQ, and KFT. Table 2 shows
good to excellent correlations of the UKFS with the DHI and the HAQ.\textsuperscript{11} Fair to moderate correlations were ascertained between UKFS and the
HAMIS, AHFT, and KFT.\textsuperscript{11} For the HAMIS, only the correlation with the
KFT was in the good to excellent range; correlations with the other tests
were in the fair to moderate range.

One way to determine validity of instruments is to correlate scores with
variables known to have a converging relationship and with variables con-
sidered to have a moderate to minimal or divergent relationship. Thus, the
UKFS would be expected to have higher relationships with the DHI and
HAQ and less strong relationships with the measures of joint motion
(HAMIS, KFT), hand strength, and dexterity (AHFT). Indeed, the
strongest correlations for the UKFS were with the other self-reports, the
DHI and HAQ, which have similar questions. However, the correlations
with the sections on the AHFT are good, suggesting that perceptions of
ability do reflect actual hand skills. The lower correlations between the
UKFS scores and the HAMIS and KFT suggest a divergent relationship,
and are not surprising as these 2 tests correlated highly with each other and
assess joint motion rather than hand function. Thus, the findings support
the convergent validity of the HAMIS. On the other hand, the HAMIS only
moderately correlated with the 3 self-reports of function. The slightly
stronger correlations between the HAMIS and scores from the AHFT sug-
gest that range of motion is related to hand strength and dexterity.
However, from the moderate correlations, it seems that measures of joint
motion, strength, and dexterity are not synonymous with hand function.
These findings are similar to other studies with persons with RA, which
found that self-reports of hand function had convergent relationships with
other self-reports of functional ability, but divergent relationships with
variables such as pain, stiffness, tenderness and swelling, and joint motion.\textsuperscript{12,13} The divergent relationships suggest that measures of impair-
ment such as joint motion, hand strength, and dexterity do not adequately
address hand function, and impairment measures should be accompanied
by assessments of hand function. Thus, the UKFS and HAMIS comple-
ment each other and are simple and easy to administer. They are valid for
use with persons with RA and may be useful as outcome measures of hand
function and joint motion.

Table 1. Description and descriptive statistics for the performance of 40 subjects on the UKFS, HAMIS, DHI, HAQ, AHFT, and KFT.

<table>
<thead>
<tr>
<th>Concept Measured</th>
<th>Question Structure</th>
<th>Possible Score Range</th>
<th>Sample Mean (SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td>UKFS\textsuperscript{1}</td>
<td>9 upper extremity function items, 2 muscle strength items</td>
<td>Self-report: ability rated from 0 (able to perform in normal manner) to 3 (impossible to do). Scores for each item summed to obtain a total score</td>
<td>0–33</td>
</tr>
<tr>
<td>HAMIS\textsuperscript{4,5}</td>
<td>9 items of joint motion: finger flexion, extension, abduction; wrist flexion, extension; thumb abduction, pincer grip, pronation, supination</td>
<td>Performance test: ability scored from 0 (no impairment) to 3 (cannot do). Scores for each item summed to obtain a total score</td>
<td>0–54</td>
</tr>
<tr>
<td>DHI\textsuperscript{7}</td>
<td>18 items in 5 categories: kitchen, dressing, hygiene, office, and other</td>
<td>Self-report: ability rated from 0 (no difficulty) to 5 (impossible). Scores for each item summed to obtain a total score</td>
<td>0–90</td>
</tr>
<tr>
<td>HAQ\textsuperscript{8}</td>
<td>20 items in 8 categories: dressing and grooming, arising, eating, walking, hygiene, reach, grip, outside activity</td>
<td>Self-report: ability rated from 0 (no difficulty) to 3 (unable to do). High scores for each category are summed and divided by the number of categories answered</td>
<td>0–3</td>
</tr>
<tr>
<td>AHFT items: Strength total (pounds)\textsuperscript{9}</td>
<td>3 items performed with each hand: grip strength, 2-point pinch strength, 3-point pinch strength</td>
<td>Performance test: score is the mean of 3 trials for the grip and 2 pinch items. Scores are summed</td>
<td>0–260</td>
</tr>
<tr>
<td>Applied Dexterity total (seconds)\textsuperscript{9}</td>
<td>5 items: fasten and unfasten 4 buttons, lace a shoe and tie a bow, pin and unpun 2 safety pins, pick up and manipulate coins, cut a piece of putty with a knife and fork</td>
<td>Performance test: score is the sum of times for the 5 items</td>
<td>0–400</td>
</tr>
<tr>
<td>Pegboard total (seconds)\textsuperscript{9}</td>
<td>1 item performed with each hand: 9-hole peg test</td>
<td>Performance test: score is the time to place and remove 9 pegs from a board with each hand. Score for each hand is summed</td>
<td>0–120</td>
</tr>
<tr>
<td>Total applied strength (milliliters)\textsuperscript{9}</td>
<td>2 items: lift a tray filled with cans of soup, pour water from a pitcher</td>
<td>Performance test: score is the number of cans on the tray (summed in ml) and total amount of water lifted in the pitcher</td>
<td>0–5800</td>
</tr>
<tr>
<td>KFT total\textsuperscript{10}</td>
<td>9 items of joint motion: finger flexion, wrist flexion and extension, forearm pronation and supination, elbow flexion</td>
<td>Performance test: depending on the item, ability rated from 0 (item performed fully or normally) to 2 (performed partially) or 3 (unable to do)</td>
<td>0–21</td>
</tr>
</tbody>
</table>

UKFS: United Kingdom Scleroderma Functional Score; HAMIS: Hand Mobility Test in Scleroderma; DHI: Duruöz Hand Index; HAQ: Health Assessment Questionnaire; AHFT: Arthritis Hand Function Test; KFT: Keital Functional Test.

Letters

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Table 2. Spearman rho correlation coefficients for the SFAQ and HAMIS (n = 40).

<table>
<thead>
<tr>
<th>Variable</th>
<th>SFAQ</th>
<th>HAMIS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Self-report measures</td>
<td></td>
<td></td>
</tr>
<tr>
<td>UKFS</td>
<td>1.00</td>
<td>0.47**</td>
</tr>
<tr>
<td>DHI</td>
<td>0.85**</td>
<td>0.36*</td>
</tr>
<tr>
<td>HAQ</td>
<td>0.86**</td>
<td>0.44**</td>
</tr>
<tr>
<td>Performance based measures</td>
<td></td>
<td></td>
</tr>
<tr>
<td>AHFT</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Strength total (pounds)</td>
<td>–0.65**</td>
<td>–0.65**</td>
</tr>
<tr>
<td>Pegboard total</td>
<td>0.63**</td>
<td>0.57**</td>
</tr>
<tr>
<td>Applied dexterity total</td>
<td>0.45**</td>
<td>0.55**</td>
</tr>
<tr>
<td>Applied strength total (milliliters)</td>
<td>0.53**</td>
<td>–0.36*</td>
</tr>
<tr>
<td>HAMIS Total</td>
<td>0.47**</td>
<td>1.00</td>
</tr>
<tr>
<td>KFT</td>
<td>0.37*</td>
<td>0.87*</td>
</tr>
</tbody>
</table>

*p < 0.05; ** p < 0.01.

Corrections

The Journal of Rheumatology Table of Contents, volume 32, number 8, August 2005. The title of the article at page 1426 should be, "Evaluation of the MDHAQ in Finnish Patients with RA." We regret the error.

The Journal of Rheumatology Table of Contents, volume 32, number 8, August 2005. The title of the article at page 1432 should be, "Further Development of a Physical Function Scale on a MDHAQ for Standard Care of Patients with Rheumatic Diseases." We regret the error.

Hudson M, Baron M, Rahme E, Pilote L. Ibuprofen may abrogate the benefits of aspirin when used for secondary prevention of myocardial infarction. J Rheumatol 2005; 32:1589-93. Table 2 should appear as follows. We regret the error.