The modified Nail Psoriasis Severity Index: validation of an instrument to assess psoriatic nail involvement in patients with psoriatic arthritis.

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J Rheumatol 2007;34;123-129
http://www.jrheum.org/content/34/1/123

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Psoriatic arthritis (PsA) is a chronic, inflammatory arthropathy characterized by the association of arthritis and psoriasis. Psoriasis affects about 2% of the population and PsA has been reported in 7%–42% of those with psoriasis. Estimates of the prevalence of PsA are variable, due in part to the heterogeneity of the disease, as well as lack of validated diagnostic criteria.

Nail involvement is common in patients with psoriasis and PsA, and can be severe and disfiguring. The frequency of psoriatic nail involvement in persons with PsA has been reported from 63% to 83%. One study of 1728 psoriatic patients found 93% considered nail psoriasis a cosmetic handicap, 48% felt it interfered with their jobs, and 52% reported it was associated with pain.

Psoriasis can affect all components of the nail. The nail plate is composed of hard, translucent, dead keratin. Four epithelial structures surround the nail plate: the proximal nailfold, the matrix, the nailbed, and the hyponychium. The matrix, located at the proximal end of the nail, synthesizes 90% of the nail plate. The distal edge of the matrix extends from underneath the proximal nailfold and is seen as the white half-moon called the lunula. The proximal matrix produces the superficial nail plate and distal matrix produces the deeper nail plate. The proximal nailfold overlies the matrix and extends onto the proximal nail bed. The nailbed and plate are firmly attached, most likely by longitudinal ridges and furrows in the bed and plate meshing together. The hyponychium is the short segment beginning at the distal nailbed and ending at the distal groove, a ridge adjacent to the volar skin of the finger.

Characteristic features of psoriasis affecting the nail matrix include: pitting, leukonychia, red spots in the lunula, and nail plate crumbling. Pits are sharply defined depressions in the plate caused by shedding of nail plate cells, much the same way psoriatic scale is shed. Superficial pitting is produced by parakeratosis or temporary, ineffectual keratinization in the proximal matrix, while deeper lesions in the nail, such as smooth-surfaced leukonychia, come from a perakeratotic focus in the middle matrix. Crumbling, gross alteration of the nail plate, comes from extensive matrix involvement. Red spots in the lunula are small pink or red macules in the lunula. Characteristic features of psoriasis affecting the nailbed...
include oil-drop discoloration, onycholysis, nailbed hyperkeratosis, and splinter hemorrhages. Oil spots are local separations of the nail plate from the bed, with accumulation of cellular debris and serum in this space. Splinter hemorrhages are small extravasations of red blood cells in the dermal ridges that lodge between the epidermis and nail plate, and are dragged as the plate streams distally. Onycholysis and subungual hyperkeratoses reflect psoriasis of the hyponychium. Compared to skin psoriasis, nailbed psoriasis has the same layering of parakeratosis and accumulation of polymorphonuclear leukocytes in the stratum corneum, but has more spongiosis in the epidermis and more serum accumulation in the stratum corneum. The availability of highly effective therapies for psoriasis and PsA has raised the possibility of treating nail psoriasis in a more effective way. Quantitative assessments of response of treatments require a validated tool for assessing the activity and extent of psoriatic nail involvement. This would allow quantification of disease and response. Thus, the Nail Psoriasis Severity Index (NAPSI) was recently developed as a psoriatic nail grading instrument. This instrument involves rating the presence or absence of features of psoriatic nail matrix disease (pitting, leukonychia, red spots in the lunula, and nail plate crumbling) and psoriatic nailbed disease (oil-drop discoloration, onycholysis, splinter hemorrhage, and subungual hyperkeratosis) in each of 4 quadrants of each fingernail. This leads to a possible total score of 0–80. While correlation of NAPSI scores among different graders has been assessed, the instrument has not been formally validated. In addition, the NAPSI was developed using real-time assessments of patients.

Our goal was to validate a psoriatic nail disease assessment instrument, beginning with the NAPSI, and modify it as necessary to enhance its performance characteristics to measure psoriatic nail disease in patients in clinical trials. To accomplish this and allow recording of nail assessments at different times, we chose to use digital photographs of individual nails of patients with PsA to test the tool. As a secondary aim, we wanted to determine the extent to which psoriatic nail disease severity correlated with other disease indicators in PsA.

**MATERIALS AND METHODS**

The protocol for this study was reviewed and approved by the University of California, San Diego (UCSD) Institutional Review Board in accord with the requirements of the Code of Federal Regulations on the Protection of Human Subjects, including its relevant subparts. Digital macrophotographs were taken of a convenience sample of 29 patients with PsA from the UCSD Arthritis Clinic. Each patient had a separate photograph taken of each nail on both hands using a Nikon Coolpix5000 5-megapixel camera on macro setting. In preliminary assessments, photographs including more than a single nail did not allow adequate identification of psoriatic nail characteristics. Individual macrophotographs were taken using the same ambient overhead fluorescent light with the nail placed at a standard fixed distance from the camera lens on a wall against a blue cloth background. Because of factors such as greater heterogeneity in size and shape and technical issues making individual photographs difficult, we did not include toenails.

Other clinical data collected about the patients, concomitant with the nail photography, included: swollen joint count (scored 0–66), tender joint count (0–68), Health Assessment Questionnaire (HAQ), Psoriasis Area and Severity Index (PASI), presence of dactylitis or enthesitis, medications used, pertinent medical histories, duration of morning stiffness, physician visual analog scales (VAS) of overall PsA disease and nail activity, and patient VAS for skin, joint and nail activity. In the initial phase, after study and review of the NAPSI scoring system, 6 physicians scored the nail photographs using the original NAPSI instrument. Analysis of this data revealed significant differences in scoring among evaluators. Of note, interrater variability accounted for only 52% of the variance in nail scores. Therefore, 2 sequential focus sessions were held. During the focus sessions, the physicians involved in the initial phase of the study discussed specific reasons they had scored a particular nail and feature in a certain way. Reasons for differences between scores were discussed as a group, as were potential solutions to achieve consensus. Based on these focus sessions, the original NAPSI was modified in an attempt to enhance its reliability and its face validity. First, the division of the nail into quadrants was eliminated because quadrants were felt to be difficult to precisely quantify in many cases, and hence varied among observers. Additionally, it was noted that patients’ disease was often asymmetric, leading to the possibility that if a patient had severe nail disease, but in only half of the nail, the score might not reflect the true severity. Second, a more quantitative aspect was added to the scoring of several features. In addition to scoring for the presence or absence of some features, as in the original NAPSI, the modified NAPSI (mNAPSI) takes into account the amount or severity of the most commonly seen features. Nail pitting was scored 0–3 depending on the number of pits present — a score of 0 if no pits were present, a score of 1 if 1–10 pits were present, a score of 2 if 11–49 pits were present, and a score of 3 if ≥ 50 pits were present. Crumbling and onycholysis were scored 0–3 depending on the percentage of the nail involved. For crumbling, the score was determined as follows: 0 if no crumbling was present, 1 if 1–25% of the nail had crumbling, 2 if 26–50% of the nail had crumbling, and 3 if > 50% of the nail had crumbling. For onycholysis the score was determined as follows: 0 if no onycholysis, 1 if 1–10% of the nail had onycholysis, 2 if 11–30% had onycholysis, and 3 if > 30% of the nail had onycholysis. It was felt that this would add to the sensitivity of the overall grading. The other features evaluated in the original NAPSI — splinter hemorrhages, leukonychia, red spots in the lunula, oil-drop dyschromia, and nailbed hyperkeratosis — were individually scored in the mNAPSI as 1 if they were present and 0 if absent.

Oil-drop dyschromia is a very important feature of nail psoriasis and is part of the same pathologic process as onycholysis. Therefore, after further discussion, it was decided that oil-drop dyschromia and onycholysis should be graded together. The combined area (as percentage) of the nail involved by either onycholysis or oil-drop dyschromia would be graded by the schema as previously described for onycholysis. Additional analysis of the scoring of individual nail photographs was carried out using these further modifications, and there was no significant change in the results. In the end, the range of possible scores using the mNAPSI was 0–14 for each fingernail, or 0–140 for all 10 fingernails (see Appendix). In addition to the mNAPSI, global nail psoriasis severity ratings from both patients and physicians were added using a VAS (0–10).

The original photographs of 20 patients were subsequently regraded by 5 physicians using the new mNAPSI. Inter- and intrarater reliability were assessed with Cronbach’s alpha and intraclass correlation coefficient (ICC). Values for Cronbach’s alpha > 0.7 are generally considered a marker of high reliability, and ICC values > 0.9 are considered to indicate excellent correlation. Correlations between mean nail scores and physician global severity VAS scores and between mean nail scores and clinical markers of disease were assessed using Spearman’s rho. All analysis was carried out using JMP (SAS Institute, Cary, NC, USA) The data presented here represent scores using the mNAPSI.

**RESULTS**

Patient characteristics are shown in Table 1. Overall, the pop-
A population included more men than women. The average duration of psoriasis and PsA was about 18 and 11 years, respectively. Most patients (90%) were receiving treatment with a disease modifying antirheumatic drug (DMARD; methotrexate, sulfasalazine, leflunomide, hydroxychloroquine), a biologic agent (etanercept, infliximab, adalimumab, or alefacept), or both a DMARD and biologic agent.

The range of mNAPSI scores (mean of the 5 graders’ scores) in our cohort of PsA patients was 8.2–69.6. The mean mNAPSI score was 22.96 (SD 15.62) and the median mNAPSI score was 18.10. The range for physician global nail severity VAS scores for the cohort was 0.44 to 8.4 (mean 2.54, SD 2.33, median 1.55). The mean patient global nail severity VAS was 2.22 (SD 3.12, median 0.85).

Comparison of the individual physician graders’ mNAPSI scores showed excellent internal consistency and interrater reliability (Cronbach’s alpha 0.98, ICC 0.92, 95% confidence interval 0.87–0.97). These results are illustrated by the scatterplot in Figure 1. Internal consistency and interrater reliability of individual physician global nail severity VAS scores were also excellent (Cronbach’s alpha 0.98, ICC 0.93, 95% CI 0.87–0.97). These results are illustrated by the scatterplot in Figure 2. Correlation between an individual grader’s mNAPSI scores and VAS scores was strong (Spearman’s rho for the 5 graders: 0.87–0.97).

### Table 1. Patient characteristics (n = 20).

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex, M/F</td>
<td>16/4</td>
</tr>
<tr>
<td>Age, mean ± SD yrs</td>
<td>52.6 ± 14.7</td>
</tr>
<tr>
<td>Psoriasis duration, mean ± SD yrs</td>
<td>18.8 ± 16.3</td>
</tr>
<tr>
<td>Psoriatic arthritis duration, mean ± SD yrs</td>
<td>11 ± 9.2</td>
</tr>
<tr>
<td>Tender joint count, range 0–68</td>
<td>7.4 ± 12.3</td>
</tr>
<tr>
<td>Swollen joint count, range 0–66</td>
<td>3.0 ± 3.8</td>
</tr>
<tr>
<td>PASI score, range 0–72</td>
<td>5.0 ± 9.9</td>
</tr>
<tr>
<td>HAQ score, range 0–3</td>
<td>0.59 ± 0.67</td>
</tr>
<tr>
<td>Patients with ≥ 30 min morning stiffness, % (n = 19)</td>
<td>36.8</td>
</tr>
<tr>
<td>Patients with enthesitis, %</td>
<td>15</td>
</tr>
<tr>
<td>Patients with dactylitis, %</td>
<td>5</td>
</tr>
<tr>
<td>Patients treated with tumor necrosis factor inhibitor (etanercept, adalimumab, or infliximab), %</td>
<td>40</td>
</tr>
<tr>
<td>Patients treated with methotrexate, %</td>
<td>65</td>
</tr>
<tr>
<td>Patients treated with sulfasalazine, %</td>
<td>10</td>
</tr>
<tr>
<td>Patients treated with alefacept, %</td>
<td>5</td>
</tr>
<tr>
<td>Patients treated with hydroxychloroquine, %</td>
<td>5</td>
</tr>
<tr>
<td>Patients treated with prednisone, %</td>
<td>5</td>
</tr>
<tr>
<td>Patients treated with leflunomide, %</td>
<td>5</td>
</tr>
<tr>
<td>Patients treated with NSAID or COX-2 inhibitor, %</td>
<td>55</td>
</tr>
<tr>
<td>Patients treated with no DMARD or biologic agent, %</td>
<td>10</td>
</tr>
</tbody>
</table>

PASI: Psoriasis Area and Severity Index, HAQ: Health Assessment Questionnaire.

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![Image of scatterplot](https://www.jrheum.org/...)

**Figure 1.** Individual scores for mNAPSI scores paired among 5 evaluators. The ellipse indicates the 95% confidence interval for the intraclass correlation coefficient.
observers: 0.90, 0.93, 0.94, 0.96, and 0.99; p < 0.001 for all). Interrater correlations between mNAPSI and VAS scores were also strong (Spearman’s rho 0.85; p < 0.01; Figure 3). Additionally, there was significant correlation between the physician and patient global nail severity VAS scores (Spearman’s rho 0.47; p = 0.04; Table 2).

Several clinical measures of psoriasis and PsA were significantly correlated with the mNAPSI scores (Table 2). These included (p < 0.05) physician global PsA disease severity VAS, swollen joint count, tender joint count, and patient global nail severity VAS. Physician global PsA disease severity and nail severity VAS scores correlated significantly (p < 0.01). PASI scores, patient global arthritis and skin severity VAS scores, and HAQ scores did not correlate with mNAPSI scores. Interestingly, patient global skin and nail disease severity VAS scores also did not correlate. The presence of active enthesitis and dactylitis did not correlate with nail severity scores; however, there were very few patients with these features in our database, impairing the ability to assess such associations. Also, presumably because the majority of patients were on active treatment, total joint counts were low. Therefore, there were not enough patients with distal interphalangeal (DIP) joint arthritis to adequately assess correlations with nail severity.

DISCUSSION
Nail involvement in PsA is a common, important problem. Many studies have evaluated the effectiveness of various treatments for nail psoriasis. Some have tried to determine correlations between psoriatic nail disease and other features of PsA. Each study developed a different outcome measure, some more subjective and some more objective, to assess psoriatic nail disease severity and response to treatment. This makes comparison between trials and treatments very difficult. Additionally, to our knowledge, none of the outcome measures has been validated.

The authors of the NAPSI made great strides in developing a tool that had excellent content validity, as it incorporated all clinically relevant aspects of psoriatic nail disease. Recently, several studies have used the original NAPSI as an outcome measure for the treatment of nail psoriasis. However, when we used the original NAPSI to grade macrophotographs of psoriatic nail involvement, we found significant interobserver variability. Therefore, modifications were made to enhance its face validity and feasibility based on discussions raised in our focus groups.

Using the mNAPSI, the correlations between the different graders’ scores indicate good reproducibility. Construct validity is shown by the correlations between mNAPSI scores and global nail severity VAS scores and by correlation between the physician and patient global assessments of nail disease activity.

Parrish, et al described modifications they made to the NAPSI. They comment on some similar aspects of the orig-
The modifications suggested by Parrish, et al have not been validated. In the mNAPSI, macrophotographs of individual nails were used for scoring, rather than live assessments as in the original NAPSI. This allowed numerous assessors to grade the same nails at different timepoints. The goal was to develop a tool useful for clinical trials. Photographs allowed standardization and would allow for review of data during a clinical trial. We recognize that using photographs, rather than live

Table 2. Correlations between clinical parameters and measurements.

<table>
<thead>
<tr>
<th>Comparison</th>
<th>Spearman’s rho</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Physician global disease severity VAS vs physician global nail disease severity VAS</td>
<td>0.57</td>
<td>&lt; 0.01</td>
</tr>
<tr>
<td>mNAPSI score vs patient global nail disease severity VAS</td>
<td>0.55</td>
<td>0.01</td>
</tr>
<tr>
<td>mNAPSI score vs physician global disease severity VAS</td>
<td>0.53</td>
<td>0.02</td>
</tr>
<tr>
<td>mNAPSI score vs tender joint count</td>
<td>0.53</td>
<td>0.02</td>
</tr>
<tr>
<td>Patient global skin disease severity VAS vs PASI</td>
<td>0.53</td>
<td>0.02</td>
</tr>
<tr>
<td>Patient global nail disease severity VAS vs physician global nail disease severity VAS</td>
<td>0.47</td>
<td>0.04</td>
</tr>
<tr>
<td>mNAPSI score vs swollen joint count</td>
<td>0.44</td>
<td>0.05</td>
</tr>
<tr>
<td>mNAPSI score vs patients global arthritis severity VAS</td>
<td>0.42</td>
<td>0.07</td>
</tr>
<tr>
<td>mNAPSI score vs HAQ</td>
<td>0.41</td>
<td>0.07</td>
</tr>
<tr>
<td>mNAPSI score vs PASI</td>
<td>0.38</td>
<td>0.10</td>
</tr>
<tr>
<td>mNAPSI score vs patient global skin disease severity VAS</td>
<td>0.34</td>
<td>0.14</td>
</tr>
<tr>
<td>Patient global skin disease severity VAS vs patient global nail disease severity VAS</td>
<td>0.07</td>
<td>0.76</td>
</tr>
</tbody>
</table>

mNAPSI: modified Nail Psoriasis Severity Index, PASI: Psoriasis Area and Severity Index.
assessments where an assessor could look at a nail from different sides and angles, probably decreases the sensitivity of the tool, particularly when assessing onycholysis and hyperkeratosis. However, in the setting of clinical research trials, photographs provide the benefits of being able to archive photographs and make comparisons across sites. The mNAPSI is also a feasible measure, as a set of 10 photographs can be scored in just a few minutes.

Our study has several potential limitations. All the patients in the study had PsA. The original NAPSI was developed using patients with psoriasis, but not necessarily with PsA. Additionally, the majority of the subjects were actively treated. The original NAPSI was developed by dermatologists in the clinic. The modifications on the original NAPSI to create the mNAPSI were made by rheumatologists, with dermatologists’ input, as a tool for clinical trials. Our goal was to develop a tool to assess disease severity and response to treatment in clinical trials, keeping in mind that the assessor in a clinical trial most likely would not be a trained dermatologist. Outcome tools used in clinical trials are often slightly different than those used in the clinic, as with the case of joint counts in clinical trials, and this likely will be true for a psoriatic nail disease assessment tool as well.

An additional goal of our study was to further characterize any correlations between psoriatic nail disease and other disease measures in PsA. Using the mNAPSI score as the “gold standard” measure of psoriatic nail disease severity, several significant correlations with other disease measures were noted. These included the physician global PsA disease severity score, the swollen joint count, and the tender joint count. Previously, Elkayam, et al found a correlation of nail disease and tender and swollen joint counts in a series of 70 patients with PsA5. In our study, other clinical indicators, such as the HAQ and PASI, did not correlate with mNAPSI scores. In a study of 100 patients with PsA, Jones, et al did find a correlation between PASI and nail scores (which were determined based on presence or absence of pitting, onycholysis, hyperkeratosis, and severe nail deformity with involvement of both sides of the nails)17. Williamson, et al assessed 69 PsA patients and found correlations between nail disease severity and skin disease, enthesis, polyarticular disease, HAQ, higher depression and anxiety scores, and unremitting and progressive arthritis7. There were not enough patients in our database with active DIP joint arthritis to assess for correlations between nail disease severity and DIP joint arthritis. In the literature there are mixed reports of correlations of DIP joint arthritis and psoriatic nail disease, with some showing no relationship5,18 and others showing a positive correlation6,7,17. While finding no correlation with presence of nail disease and DIP joint arthritis, Scarpa, et al did find those with onychopathy had more marked bone involvement at the level of the DIP joint18. A recent study using magnetic resonance imaging highlighted diffuse involvement of the nailbed in PsA patients who had DIP involvement, a finding not observed among patients with osteoarthritis of the DIP joints19. PsA is a heterogeneous disease, and understanding some correlations will add to knowledge of the disease and how to better care for our patients.

The mNAPSI needs to be validated with more patients, patients with heterogeneous levels of disease activity, and with patients who have psoriasis without PsA. The mNAPSI also must be tested longitudinally to assess its discriminant validity or responsiveness to change.

Patients consider psoriatic nail disease a significant problem and therefore understanding and treating nail involvement are important. New effective treatments of PsA and psoriatic nail disease require outcome measures that are standardized, validated, reproducible, and reliable, and that allow for comparison of outcomes between trials. We modified the original NAPSI tool and have shown that the mNAPSI has reproducibility and construct validity. Prospective, longitudinal testing of the tool to assess discriminant validity is under way. The mNAPSI may be a valuable tool for evaluating psoriatic nail disease.

Appendix. Modified NAPSI Instructions
This tool will ask you to assess each nail abnormality for each of a subject’s nails. If you question which grade to give, your answer should be the lower of the grades. Three features or groups of features (pitting, onycholysis and oil-drop dyschromia, and crumbling) of each fingernail will be graded on a scale from zero to 3, according to the directions below. Four features (leukonychia, splinter hemorrhages, hyperkeratosis, and red spots in the lunula) will be graded as either present or absent for each fingernail. After you have viewed all the nails of a subject, consider all aspects of all of the subject’s nails and place a mark on the visual analog scale giving a global assessment of their nails.

1. Onycholysis: Separation of the nail plate from the nail bed. The separated part of the nail is opaque and can have white, yellow, or greenish tinge. If there is a piece of nail missing, estimate where the nail normally would have ended at the end of the nail bed, and count that missing part as involved in onycholysis.

   Oil-drop (salmon patch) dyschromia: Reddish-brown discoloration under the nail plate.

   Onycholysis and oil-drop dyschromia are considered together. When looking at the nail, combine the total percentage area of the nail that is affected by either and use that combined total to score the nail.

2. Pitting: Small, sharply defined depressions in the nail surface. Pits are discrete abnormalities (“ice-pick-like”). If there is nail plate crumbling that is confluent with pits, do not score for pits. If the pits are separate from crumbling, they may be scored regardless of whether crumbling is present or not.

3. Nail plate crumbling: Crumbling or fragmentation of friable nail plate which may be associated with confluent pitting. Crumbling involves alteration of the nail plate surface. Horizontal ridging of the nail, “wave-like” appearance, and horizontal lines are all features of crumbling.
Score | Percent of nail with crumbling present
--- | ---
0 | No crumbling
1 | 1–25% of the nail has crumbling
2 | 26–50% of the nail has crumbling
3 | > 50% of the nail has crumbling

The next 4 abnormalities are scored only by their presence or absence. A score of 1 indicates present and a score of zero indicates not present.

1. Leukonychia: White spots in the nail plate due to psoriasis in the mid matrix. Leukonychia are just color changes. If it appears that there is depression or irregularity to the nail surface, this may be pitting or crumbling, not leukonychia. If the leukonychia is adjacent to, or confluent with crumbling or pits, it is counted as part of the crumbling or pitting and not as a separate abnormality.

2. Splinter hemorrhages: Small, longitudinal, linear, dark brown hemorrhage under the fingernail.

3. Nail bed hyperkeratosis: Thickened keratin in the nail bed.

4. Red spots in the lunula: Small pink or red macules in the lunula.

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