A Novel Severity Assessment Scoring System for Hidradenitis Suppurativa

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IMPORTANCE  The variation in both clinical appearance and responses to diverse treatment options emphasize the importance of an accurate, clinically relevant, yet easy-to-use scoring system in hidradenitis suppurativa.

OBJECTIVE  To propose and provide validation data for the newly designed Severity Assessment of Hidradenitis Suppurativa score.

DESIGN, SETTING, AND PARTICIPANTS  We prospectively assessed disease severity using Hurley staging and the modified Hidradenitis Suppurativa Score in 355 patients referred to Ruhr-University Bochum Department of Dermatology between March 2016 and June 2017. We also assessed disease severity via the Severity Assessment of Hidradenitis Suppurativa score.

MAIN OUTCOMES AND MEASURES  Evaluation and assessment of convergent validity and responsiveness to treatment of the Severity Assessment of Hidradenitis Suppurativa score.

RESULTS  Eighty-eight of the 355 patients (134 [37.7%] men and 221 [62.3%] women with a median [IQR] age of 40 [30-49] years) were classified as Hurley stage I, 221 were Hurley stage II, and 46 were Hurley stage III, with an overall median modified Hidradenitis Suppurativa Score of 31 (interquartile range [IQR], 19.3-53). The median total Severity Assessment of Hidradenitis Suppurativa score was 6 (IQR, 4-9), significantly different among the 3 Hurley groups. The median SAHS score for patients in Hurley stage I was 5 (IQR, 3-6), 6 (IQR, 5-9) for patients in Hurley stage II, and 9 (IQR, 7-12) for patients in Hurley stage III (P < .001, Kruskal-Wallis test). Correlation analysis showed a significant correlation between the modified Hidradenitis Suppurativa Score and the Severity Assessment of Hidradenitis Suppurativa score (r = 0.79, P < .001). Disease severity assessment before and after 3 months of conservative systemic treatment showed a significant correlation between the Severity Assessment of Hidradenitis Suppurativa score and modified Hidradenitis Suppurativa Score. Both the mHSS (P = .001) and the SAHS score (P < .001) significantly differed between the baseline visit (median mHSS, 33 [IQR, 24-52]; median SAHS score, 6 [IQR, 5-9]) and the 3-month visit (median mHSS, 28 [IQR, 15-43.5]; median SAHS score, 5 [IQR, 4-6.3]). The 2 patient-reported items demonstrated excellent test-retest reliability with intraclass correlation coefficient values greater than 0.8.

CONCLUSIONS AND RELEVANCE  Our validation data demonstrated that the Severity Assessment of Hidradenitis Suppurativa score is a disease severity instrument that significantly correlates with Hurley staging and the modified Hidradenitis Suppurativa Score, and is responsive enough to measure treatment outcome.
Hidradenitis suppurativa (HS) is a chronic skin disease that predominantly affects the skin of the intertriginous area through an inflammatory process and results in recurrent nodules, abscesses, sinuses, fistulas, and scarring. Because the precise description of HS may provide a challenge, considerable efforts have been made to formulate diagnostic criteria and validate clinical descriptive terms for the lesions occurring in patients with HS. In addition, an international initiative (HS core outcomes set international collaboration) has started to develop a list of mandatory outcomes that should be measured in clinical trials on the treatment of HS. However, the variation in clinical appearance and the variability in response to a wide range of proposed treatments underline the importance of an accurate disease severity assessment tool.

The first severity classification of HS proposed by Hurley in 1989 is routinely used in the clinical setting to categorize patients into 3 stages based mainly on the presence of sinus tracts and scarring. Although the Hurley scoring system is a simple tool, it is nonquantitative and static. Sartorius et al described a more complex and detailed severity scoring method better suited to assess disease severity and grade of inflammation. Although it has been modified, it is still difficult to use and time-consuming in routine clinical practice. In addition, its practical applicability is difficult in severe cases with more confluent lesions. Although there is no gold standard, the Hurley staging and the Sartorius scoring systems are the most commonly used assessment tools in clinical trials and daily practice. Later, several other clinical measures for assessing HS disease severity were described, including the recently proposed HS Severity Score System (IHS4) and the Hidradenitis Suppurativa Clinical Response. Notably, the latter evaluates the inflammatory response and treatment effectiveness. Although each described disease severity assessment has its advantages, our clinical experience has resulted in the following criteria for a scoring system: (a) it should be easy to use in clinical routine (ie, the identification of skin lesions, which are assessed by the score, should be clear and practicable); (b) it should be dynamic (ie, able to measure the chronic relapsing nature of the disease); (c) it should be responsive enough to measure treatment outcome; (d) it should be able to assess disease burden (eg, as measured with patient-reported items); and (e) it should be time effective (ie, it should be able to be performed quickly).

Based on these criteria, we designed a new scoring system for HS, the Severity Assessment of Hidradenitis Suppurativa (SAHS) score, and conducted this prospective study to evaluate and assess its validity.

### Methods

In this prospective study, 355 patients who were referred to our HS center from March 2016 to June 2017 were consecutively included.

The severity of HS was assessed by the Hurley staging and the modified Hidradenitis Suppurativa Score (mHSS), as previously described. In addition, disease severity was assessed by the new SAHS score, for which the following items were surveyed: number of involved regions (axilla left, axilla right, submammary left, submammary right, genitral, abdominal, mons pubis, groin left, groin right, genital, perianal or perineal, gluteal left, gluteal right, and others [eg, neck, retroauricular]), number of inflammatory and/or painful lesions other than fistula (ILOF), and number of fistula (Table 1). These physician-rated items were completed by 2 patient-reported items: patients were asked for number of new boils or number of existing boils, which flared up during the past 4 weeks and to rate the current severity of pain of the most symptomatic lesion in the course of their daily activities (eg, sitting, moving, or working) on a numerical rating scale.

### Table 1. Severity Assessment of Hidradenitis Suppurativa Score

<table>
<thead>
<tr>
<th>Score Category</th>
<th>Regions Involved, No.</th>
<th>ILOF, No.</th>
<th>Fistula, No.</th>
<th>New or Flared Existing Boils in Past 4 Weeks, No.</th>
<th>Severity of Pain (NRS-11)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0-1</td>
</tr>
<tr>
<td>1</td>
<td>1-2</td>
<td>1-4</td>
<td>1-2</td>
<td>1-2</td>
<td>2-4</td>
</tr>
<tr>
<td>2</td>
<td>3-4</td>
<td>5-9</td>
<td>3</td>
<td>3-4</td>
<td>5-6</td>
</tr>
<tr>
<td>3</td>
<td>≥5</td>
<td>≥10</td>
<td>≥4</td>
<td>≥5</td>
<td>≥7</td>
</tr>
</tbody>
</table>

Abbreviations: ILOF, inflammatory and/or painful lesions other than fistula; mHSS, modified Hidradenitis Suppurativa Score; NRS-11, numerical rating scale; SAHS, Severity Assessment of Hidradenitis Suppurativa.

* Classification: mild disease, SAHS score of 4 or less; moderate disease, SAHS score of 5 to 8; severe disease, SAHS score of 9 or higher.

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Table 2. Baseline Characteristics of the Included 355 Patients With Hidradenitis Suppurativa

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>No. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Sex</strong></td>
<td></td>
</tr>
<tr>
<td>Men</td>
<td>134 (37.7)</td>
</tr>
<tr>
<td>Women</td>
<td>221 (62.3)</td>
</tr>
<tr>
<td><strong>Age, median (IQR), y</strong></td>
<td>40 (30–49)</td>
</tr>
<tr>
<td><strong>Current smoker</strong></td>
<td>252 (71)</td>
</tr>
<tr>
<td><strong>BMI, median (IQR)</strong></td>
<td>29.8 (26.3–33.9)</td>
</tr>
<tr>
<td>Underweight (BMI&lt;18.5)</td>
<td>0</td>
</tr>
<tr>
<td>Normal weight (BMI≤24.9)</td>
<td>62 (17.5)</td>
</tr>
<tr>
<td>Overweight (BMI≥25)</td>
<td>121 (34.1)</td>
</tr>
<tr>
<td>Obesity (BMI≥30)</td>
<td>172 (48.5)</td>
</tr>
<tr>
<td><strong>Hurley stage</strong></td>
<td></td>
</tr>
<tr>
<td>I</td>
<td>88 (24.8)</td>
</tr>
<tr>
<td>II</td>
<td>221 (62.3)</td>
</tr>
<tr>
<td>III</td>
<td>46 (13)</td>
</tr>
<tr>
<td><strong>mHSS, median (IQR)</strong></td>
<td>31 (19.3–53)</td>
</tr>
</tbody>
</table>

Abbreviations: BMI, body mass index (calculated as weight in kilograms divided by height in meters squared); IQR, interquartile range; mHSS, modified Hidradenitis Suppurativa Score.

(NRS-11) ranging from 0 to 10. Each item is scored according to the criteria in Table 1. For example, in the item “number of regions involved,” 1 or 2 involved regions is scored with 1 point, 3 or 4 regions is scored with 2 points, and more than 4 regions is scored with 3 points. Each item is scored accordingly and calculated for the total SAHS score with an upper limit of 15. Mild disease is defined as an SAHS score of 4 or less, moderate is defined as an SAHS score of 5 to 8, and severe disease is defined as an SAHS score of 9 or more. The standard of the criterion validation was based on Hurley staging and mHSS.

To evaluate the test-retest reliability of the 2 patient-reported items in the SAHS score, 48 additional patients were included and completed the 2 patient-reported items. In a second visit 7 to 14 days later (this time interval was set to limit the recall of responses at retest and to limit potential change in disease severity) patients were asked again to complete the 2 patient-reported items. In addition, they determined on a 3-point Likert scale (deterioration, no change, and improvement) if any substantial change in disease severity had occurred between the first and second visit. Only data from participants who reported no change were included in test-retest reliability analysis.

Statistical analysis was made with MedCalc statistical software (version 15.2, MedCalc). We used the Kolmogorov-Smirnov test to analyze data distribution, and we used the Wilcoxon test to compare disease severity (as assessed by the mHSS and SAHS score) obtained at baseline and the 3-month visit. Differences among the 3 Hurley groups were examined using the Kruskal-Wallis test, including the Conover post hoc test for pairwise comparison. We used Spearman’s coefficient of rank correlation (r) to determine correlations between variables. To examine the test-retest reliability, the intraclass correlation coefficient (ICC, one-way random effects model, single measure, and absolute agreement definition) with a 95% confidence interval (CI) was calculated. An ICC of 0.75 or greater indicates excellent test-retest reliability, ICC between 0.6 and 0.74 indicates good, ICC between 0.41 and 0.59 indicates fair, and ICC less than 0.4 indicates poor reliability. 

The ethics review board of the Ruhr-University Bochum, Germany, approved this study, which was conducted according to the Declaration of Helsinki. Written informed consent was obtained from all study participants.

Results

The data of 355 patients were included for validation of the SAHS score (Table 2). Of these patients, 221 (62.3%) were women, and 134 (37.7%) were men, with a median age of 40 years (interquartile range [IQR], 30–49 years). Overall, 252 (71%) of 355 patients were current smokers. The median body mass index (calculated as weight in kilograms divided by height in meters squared) was 29.8 (IQR, 26.3–33.9). Regarding severity of disease, 88 (24.8%) were classified as Hurley stage I, 221 (62.3%) were Hurley stage II, and 46 (13%) were Hurley stage III with an overall median mHSS of 31 (IQR, 19.3–53).

The middle line represents the median. The whiskers extend from the minimum to the maximum value and outliers are represented with separate dots.

Multiple-comparison box-and-whisker plots showing SAHS score vs the 3 Hurley groups. The median SAHS score was significantly different between each Hurley group and increased with the degree of severity (P < .001, Kruskal-Wallis test). The central box represents the values from the 25th to 75th percentiles. The middle line represents the median. The whiskers extend from the minimum to the maximum value and outliers are represented with separate dots.

Figure 1. Comparison of the Severity Assessment of Hidradenitis Suppurativa (SAHS) Score Between the 3 Hurley Groups

0.75 or greater indicates excellent test-retest reliability, ICC between 0.6 and 0.74 indicates good, ICC between 0.41 and 0.59 indicates fair, and ICC less than 0.4 indicates poor reliability. 

The ethics review board of the Ruhr-University Bochum, Germany, approved this study, which was conducted according to the Declaration of Helsinki. Written informed consent was obtained from all study participants.
3 months to analyze the responsiveness of the SAHS score during treatment (Figure 3). Both the mHSS ($P = .001$) and the SAHS score ($P < .001$) significantly differed between the baseline visit (median mHSS, 33 [IQR, 24-52]; median SAHS score, 6 [IQR, 5-9]) and the 3-month visit (median mHSS, 28 [IQR, 15-43.5]; median SAHS score, 5 [IQR, 4-6.3]). In addition, there was a significant correlation between the SAHS score and mHSS obtained at the baseline visit ($r = 0.62$, $P < .001$) and 3-month visit ($r = 0.78$, $P < .001$).

In the eFigure in the Supplement, the assessed patient-reported items (severity of pain and number of new boils or number of existing boils that flared up during the past 4 weeks) are plotted against the SAHS score. Correlation analysis showed a significant correlation between the SAHS score and severity of pain as measured by a NRS-11 ($r = 0.56$, $P < .001$) (Figure 2), and number of new boils ($r = 0.62$, $P < .001$).

For the test-retest reliability, 48 additional patients (eTable in the Supplement) were included and retested within 7 to 14 days (median, 7 [IQR, 5-9] days). Data for 38 patients who reported that no change in disease severity had occurred between the first and second visit were included for test-retest reliability analysis. The ICC value was 0.95 (95% CI, 0.92-0.98) for number of new boils or number of existing boils that flared up during the past 4 weeks and 0.82 (95% CI, 0.68-0.90) for severity of pain.

Discussion

Disease severity assessment of patients with HS is a challenge for the physician owing to the lack of a standard assessment tool and the wide variability in the clinical appearance of HS. In daily clinical practice and as HS awareness rises, the number of consultations with patients with HS seems to be increasing.21,22 Thus, a scoring system is needed that can be easily and quickly implemented in clinical routine and provide an accurate, responsive, and clinically relevant representation of the disease severity.

Our data showed that the proposed SAHS score correlated significantly with the mHSS and differs significantly between the 3 Hurley groups, both representing the most commonly used measurements, with the mHSS being partly validated.8-11 This is, to the best of our knowledge, the largest number of patients with HS reported in the current literature in which disease severity assessment and a score validation were performed.

The SAHS score was designed to be a simple, easy-to-use, quick assessment tool to evaluate HS disease severity. Therefore, the SAHS score is not based on the identification of different inflammatory HS lesions (eg, inflammatory nodule vs abscess) because such a differentiation can present a challenge in daily clinical routine for some patients with HS, and its clinical impact is questionable. Thus, the SAHS score simply differentiates between ILOF and fistula. For the latter, the difference between draining and nondraining fistula is not necessary because it needs tactile examination with pressure on the skin, which is often painful and has only limited impact on treatment decisions. Notably, we refer to the recently presented concept of mandatory surgical indications (MIBHS) to distinguish between medical and surgical treatment in HS, which is based on the identification of 5 defined mandatory surgical indications and provides valuable assistance for the physician in daily routine for choosing between (a) surgical treatment, (b) combined surgical and conservative treatment, and (c) conservative treatment alone.23,24 The SAHS score can be well implemented in this concept.

Hidradenitissuppurativa has a considerable negative impact on the quality of life of patients, and a variety of questionnaires have been used for measurement, among which, the Dermatology Life Quality Index (DLQI) is the most commonly used.25 Although it is important to quantify the impact of a disease on the quality of life of patients (eg, during clinical studies), implementing the DLQI in a scoring system may be difficult. It was not designed specifically for HS, and
if not completed by the patient in advance of the consultation, the total score can only be calculated after the return of the completed questionnaire, which can be an organizational problem in daily clinical routine.

However, patient-reported items are indeed important to evaluate the burden of this disabling disease on daily activities and distinguish the SAHS score from the recently proposed IHS4.15 Thus, we supplemented the physician-based items with 2 patient-reported items, which, from our experience, appropriately represent the disease burden.

The first patient-reported item concerns the severity of pain. Owing to painful lesions and predilection sites (namely the groin, axilla, and gluteal areas), the severity of pain is an important factor influencing the daily activities of patients with HS.26 To assess the severity of pain using the SAHS score, the patients are asked to rate the highest intensity of pain of the most painful lesion during activities of daily living on a range between 0 to 10 (NRS-11). The NRS-11 is a valuable measurement and commonly used for measuring pain intensity.27 Based on available data, a difference of 2 points in the NRS-11 represents the best correlation with a clinically important difference, which is essential to evaluate a relapsing chronic disease and treatment outcome.28,29 In addition, scoring of the pain intensity in the SAHS score is based on the described thresholds for mild, moderate, and severe pain on the NRS.30-33

An accurate scoring system for HS should be designed to measure the chronic, relapsing inflammatory nature of the disease. Therefore, the second patient-reported item concerns this relapsing aspect of HS. Patients are asked to report the number of new boils or number of existing boils that flared up during the past 4 weeks. According to our clinical experience and also described by Sartorius et al30 (but not included in the Sartorius score), this item provides a good representation of the inflammatory load and disease-burden experienced by patients with HS that has a strong impact on the patients’ discomfort. With the currently available conservative treatment options, a decrease in the number of new boils and a lengthening of the period between new boils is one of the important goals of therapy that should be monitored by a scoring system. The 2 patient-reported items demonstrated excellent test-retest reliability with ICC values greater than 0.8.

Limitations
Although the current study assessed 2 important validation criteria of a clinical disease severity instrument, namely convergent validity and responsiveness to treatment, future studies are needed to provide more validation evidence (ie, inter-rater reliability), evaluate the usability, and interpretability (minimal clinically important difference) of the SAHS score in clinical studies.34-36

Conclusions
The SAHS score significantly correlates with Hurley staging and the mHSS. The SAHS is a satisfactorily valid and responsive treatment scoring system for health care professionals treating patients with HS.
Gland on Disease of the Mammary Areola Preceding Cancer of the Mammary

Paget published his findings as a general treatment. Furthermore, he found that a mammary gland tumor following a mysterious illness. He observed that “calcified particles” frequently ingested by consuming uncooked pork were actually worms in their capsules.

A New Severity Assessment for Hidradenitis Suppurativa

Original Investigation Research

NOTABLE NOTES

Sir James Paget—Contributions of a Surgeon and Pathologist

Manthan Patel, MS; Varun Ayyaswami, BS; Arpan V. Prabhu, BS

Sir James Paget is remembered as one of the greatest English surgeons and fathers of pathology. After passing the College of Surgeon’s Examination in 1836, he struggled to find a job practicing medicine and spent 7 years doing various odd jobs for minimal compensation.1

In 1843, Paget joined St Bartholomew’s Hospital; while working there as a professor and assistant surgeon, he encountered 15 women 40 years or older who presented with “an eruption on the nipple and areola.” In most of these women, the erythematous eruption “had the appearance of a florid, intensely red, raw surface, very finely granular, as if nearly the whole thickness of very acute diffuse eczema, or like that of an acute balloonitis.”2(p303) Paget noticed clear, yellowish, and viscid exudation on the whole thickness of very acute diffuse eczema, or like that of an acute dermatitis.2(p315) Paget described the disease as “calcified bodies” frequently ingested by consuming uncooked pork were actually worms in their capsules.

He worked to impose a ban on the sale of uncooked pork.3 Outside of medicine, Paget had a penchant for languages and translated German, Dutch, Italian and French journals into English.1

For his accomplishments, Paget received a knighthood in 1871 and was named the vice-chancellor of the University of London, president of the Royal College of Surgeons of England, and president of the International Congress of Medicine in 1881. He died in 1899 at the age of 85 years.1

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