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# New insights into the diagnosis of hidradenitis suppurativa: Clinical presentations and phenotypes

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Current classifications for hidradenitis suppurativa (HS), such as the Hurley staging system, are not very specific. The absence of rigorous classifications is incongruous with the clinical variability of the condition. There is no pathognomonic sign for HS that leads to a diagnosis that is unquestionable. Consequently, diagnosis is made largely through clinical presentation of the condition. The use of several validated tools assists in confirming the diagnosis and the severity of the disease. In future investigations, the identification of phenotypes and clinical subtypes—taking into account genetic variants—will serve to identify subpopulations of patients who are responsive to particular therapies, thereby improving the overall therapeutic picture for patients with HS. There is a potential for personalized, tailored delivery of therapy in the HS setting. (J Am Acad Dermatol 2015;73:S23-6.)

**Key words:** clinical subtypes; hidradenitis suppurativa; Hurley staging; inflammatory; noninflammatory; Physician Global Assessment; Sartorius score; severity.

## INTRODUCTION

The diagnosis of hidradenitis suppurativa (HS) is based on the modified Dessau definition, describing the constellation of diagnostic lesions, topography, and history of the disease<sup>1</sup> and the terminology of HS has recently been reviewed.<sup>2</sup> For the diagnosis of HS, 3 criteria must be present. First, typical lesions must be present (ie, deep-seated painful nodules). These are often described as “blind boils” in the early lesions. Other lesions are abscesses, draining sinus (inflamed tunnels), bridged scars, and postinflammatory “tombstone” double-ended pseudocomedones. Most often, multiple elements are present simultaneously. It is, however, important to avoid confusion with nondiagnostic elements, such as simple folliculitis, when making the diagnosis. Second, these elements mainly occur in  $\geq 1$  of the areas for which HS has a predilection: the axillae, groin, perineal region, buttocks, and infra- and intermammary folds. Lesions may appear ectopically, but they must involve the areas for which the disease has a predilection to meet the diagnosis. Third, there must be a clear history of chronicity and

recurrence. Lesions initially recur in the areas for which HS has a predilection, only to turn chronic later on in the course of the disease. Arbitrarily, 2 recurrences over a period of 6 months have been used as a qualifier for a diagnosis.<sup>3</sup> All 3 criteria must be present for the definitive diagnosis, and because of the diagnostic requirement for recurrence/chronicity, an observation period may be necessary before the definitive diagnosis is made. Suspicion of the diagnosis can be strengthened by other factors (Table 1) that are not, however, pathognomonic.

It has been suggested that the clear symptomatology (painful lesions recognized by patients as boils), the specific areas affected, and the recurrence/chronicity of the lesions are sufficient for reliable, self-reported diagnosis, but this approach requires additional validation.<sup>4</sup>

## SCORING OF DISEASE SEVERITY

### Hurley staging

In 1989, a severity classification for HS was first proposed by Hurley<sup>5</sup> as follows:

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- Stage I—Abscess formation, single or multiple, without sinus tracts and cicatrization
- Stage II—Recurrent abscesses with tract formation and cicatrization, single or multiple, and widely separated lesions
- Stage III—Diffuse or near-diffuse involvement or multiple interconnected tracts and abscesses across the entire area

Stage I disease is most common, affecting 68% of patients, while stage II occurs in 28% of patients, and 4% of HS patients develop stage III.<sup>6</sup>

The Hurley classification is useful for rapid classification of HS severity, but it has serious limitations. First, it cannot be considered a surgical classification because it does not incorporate inflammatory features, such as erythema and discharge. It is based on static disease characteristics, such as scarring and fistulas, which are only treatable with surgery. It is therefore static and not useful to monitor the efficacy of medical therapy. Second, because scarring is a key feature of stage II and III patients and irreversible, the score can never fall below II, even if the disease is in complete remission. Third, the classification, with just 3 stages, is not a precise monitoring tool in the clinical trial setting.

### Sartorius score

A more detailed and dynamic HS severity score for measuring clinical severity was created by Sartorius et al<sup>7-9</sup> and was later modified and partially validated. The original Sartorius score included counting individual scars and pustules, but this was abandoned in the modified Sartorius score.<sup>7-9</sup> The parameters in the modified Sartorius score include counting of individual nodules and fistulas, measuring the longest distance between 2 lesions, and adding extra points to Hurley III areas.<sup>8</sup> Although the severity of mild HS can be measured quite readily with the Sartorius score and in a reasonable amount of time, its use may be limited in more severe cases (ie, patients with Hurley stage III disease). In these cases, distinguishing separate lesions becomes challenging, particularly when lesions become confluent and sinuses become interconnected. Still, even with these shortcomings, the Sartorius score is frequently used in clinical trials.

### CAPSULE SUMMARY

- Hidradenitis suppurativa is currently attracting significant interest from the scientific community.
- No pathognomonic test exists, leaving the diagnosis to be a clinical one.
- A clear definition of diagnosis is needed to facilitate further developments in the treatment of hidradenitis suppurativa.

### Physician Global Assessment

An anchored 6-stage, HS-specific Physician Global Assessment (PGA) has been defined for use in a phase II trial.<sup>10</sup> These are the defined stages:

- Clear—No inflammatory or noninflammatory nodules
- Minimal—Only the presence of noninflammatory nodules
- Mild—Fewer than 5 inflammatory nodules without abscesses and draining fistulas or 1 abscess or draining fistula without additional inflammatory nodules
- Moderate—Fewer than 5 inflammatory nodules, or 1 abscess or draining fistula and  $\geq 1$  inflammatory nodules, or 2 to 5 abscesses or draining fistulas and fewer than 10 inflammatory nodules
- Severe—Two to 5 abscesses or draining fistulas and  $\geq 10$  inflammatory nodules
- Very severe— $>5$  abscesses or draining fistulas

### The Hidradenitis Suppurativa Severity Index

The Hidradenitis Suppurativa Severity Index (HSSI) was created by Kerdell et al<sup>11</sup> and is a HS-specific severity score. This score incorporated categorical objective parameters with categorical subjective patient-reported parameters. The HSSI score has been used in 2 studies examining the efficacy of biologic agents.<sup>11,12</sup>

### Hidradenitis Suppurativa Clinical Response

The most recently developed and validated HS-specific severity score is the Hidradenitis Suppurativa Clinical Response (HiSCR) score.<sup>13</sup> It was developed based on data gathered using the PGA described earlier and data from the phase II study to which it was applied. The HiSCR score provides a valid, responsive, and meaningful clinical endpoint when studying the inflammatory manifestations of HS.<sup>13</sup> The HiSCR is defined as a  $\geq 50\%$  reduction in inflammatory (transient) lesion count (sum of abscesses and inflammatory nodules) and no increase in abscesses or draining fistulas (chronic inflamed lesions) in HS when compared with baseline. The cutoff point has been validated by the use of patient-reported outcomes, such as Dermatology Life Quality Index and the PGA. This score was only recently created, and therefore has

**Table I.** Nonpathognomonic factors that can strengthen the diagnoses of hidradenitis suppurativa

Familial history of hidradenitis suppurativa
Recurrent inflammatory noncharacteristic lesions (eg, folliculitis or open comedones) in a typical location
Typical lesions in atypical locations (usually pressure points and locations of enhanced mechanical friction, such as thighs or the belt region of the abdomen)
Presence or history of a pilonidal sinus
The absence of pathogenic microbes on routine culture

only been used in 1 randomized, controlled trial in HS investigating the efficacy of adalimumab in HS.<sup>10</sup>

### Different clinical presentations

In 2013, Canoui-Poitrine et al<sup>14</sup> identified HS subgroups by latent class analysis with a priori hypotheses in a cohort of 618 patients. They identified 3 phenotypes (Table II).<sup>14</sup> The classification has not been further validated against risk factors or treatment outcomes.

Clinical experience suggests several other possible subtypes that also require additional exploration. The authors suggested that the possible subtypes below be considered in future studies.

**The regular type.** Patients with regular HS fulfill all of the diagnostic criteria. This is probably the most common type, and all HS patients who lack other specific characteristics belong in this category.

**Frictional furuncle type.** These patients are usually overweight; in addition to regular HS, they are characterized by the presentation of multiple deep nodules and abscesses on sites exposed to enhanced friction, such as the abdomen, thighs, and buttocks. The formation of tunnels and fistulas in these areas is unusual.

**Scarring folliculitis type.** Patients with the scarring folliculitis type of HS have, in addition to regular HS, pustules, cysts, superficial nodules, depressed cribriform scarring, and double-ended comedones. These lesions are frequently seen on the buttocks, inguinal region, and pubic region. The formation of sinus tracts and fistulas in these sites may be unusual, and although the inflammatory lesions are small and superficial (Hurley stage I), scarring typically occurs. These patients are also frequently overweight and often smoke.

**Conglobata type.** These patients are characterized by cyst formation and acne conglobata lesions on the back especially, but also the face. HS usually runs in the family in this type, and is moderate to severe, Hurley stages II to III. Patients are usually men and are not overweight. The Chinese cases in

**Table II.** Proposed hidradenitis suppurativa subgroups by Canoui-Poitrine et al<sup>14</sup>

Type	Proportion	Locations	Lesion types
Axillary-mammary	48%	Breast and armpit	Hypertrophic scars
Follicular	26%	Breast and armpit, ears, chest, back, or legs	Epidermal cysts, pilonidal sinus, comedones, and severe acne
Gluteal	26%	Gluteal involvement	Follicular papules and folliculitis

whom the initial finding of gamma-secretase mutations were found may belong to this putative group.<sup>15</sup>

**Syndromic type.** Patients with syndromic type HS are characterized by concomitant manifestations, such as pyoderma gangrenosum and arthritis, in the syndromic constellation recognized as pyoderma gangrenosum, acne, and suppurative hidradenitis syndrome or pyogenic arthritis, pyoderma gangrenosum, acne, and hidradenitis suppurativa syndrome.<sup>16</sup>

**Ectopic type.** The existence of an ectopic type involving the face has been proposed.<sup>17</sup>

These proposed clinical subtypes are not based on disease severity as outlined in the Hurley staging system. All of the 3 Hurley stages can occur in these subtypes. Future studies should show the ratios between these clinical subtypes in the HS population and whether or not these clinical subtypes can be validated or whether new ones need to be added.

### IMPACT ON TREATMENT

Some level of heterogeneity exists for all major dermatoses. Clearly, not all psoriasis patients present in the same fashion, and patients with HS are no different in this regard. Clinical experience suggests variability in clinical appearance and in response to an array of therapies. Currently, a more detailed subclassification of HS is not strongly supported by data, but it is reasonable to anticipate that future studies may identify HS subtypes that respond exceptionally well to a given therapy, particularly as more data are generated in larger, longitudinal follow-up studies and the typology becomes better developed.

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