

Back to basics: understanding hidradenitis suppurativa

KEY WORDS

- ▶ Dermatology
- ▶ Fistulae
- ▶ Hidradenitis Suppurativa
- ▶ Scarring
- ▶ Sinus tracts

Hidradenitis Suppurativa (HS) is a chronic recurrent debilitating skin disease of the hair follicle. It is a condition that has been overlooked in wound care publications, with most articles found in dermatological journals. However, the condition affects 1% of the population in Europe and produces painful nodules in one or more of the apocrine-gland bearing aspects of the skin that can ulcerate and produce pain and a foul odour and can multiply and eventually develop sinus tracts and fistulae. HS is often misdiagnosed as alternative skin ulcerating conditions, leaving the individuals with many years of suffering from the physical symptoms and their psychological consequences. The disease often begins in puberty and burns out by middle age, leaving the individual with unsightly scarring. This article examines the pathophysiology, clinical presentations and comorbidities associated with the disease. The treatment options focus on controlling the comorbidities, moderating life-style behaviours and arresting the disease. The medical and surgical options are discussed along with their limitations.

Hidradenitis Suppurativa (HS) was first described by the French surgeon Velpeau in 1839. The origin of the term HS comes from the Greek hidros meaning sweat and aden denoting glands (Ather et al, 2006). Initially it was thought to be due to infection of the sweat glands however it is now recognized as an acneform disorder which begins with follicular occlusion. Therefore, apocrine involvement is incidental and not essential to the pathogenesis (Ather et al, 2006).

The following Dessau definition of HS was created at the first international conference on HS in Dessau, Germany in 2006; HS is a chronic inflammatory, recurrent, debilitating skin disease of the hair follicle that usually presents after puberty with painful deep-seated, inflamed lesions in the apocrine gland-bearing areas of the body, most commonly the axillae, inguinal and anogenital regions (Zouboulis et al, 2015a). HS is sometimes referred to as apocinitis, acne Inversa and pyoderma Fistulans significa (World Union of Wound Healing Societies [WUWHS], 2016).

The reported prevalence varies across geographical areas due to misclassification and

selection bias, however is thought to be 1% in Europe (Gulliver et al, 2016; WUWHS, 2016). Prevalence is rare in children and when HS does occur in this population it is often associated with hormonal disorders such as metabolic syndrome, precocious puberty, adrenal hyperplasia and premature adrenarche (Vivar and Kruse, 2017).

There is a predominance of HS in the female population with women being three times more affected. Incidence is higher in African and Afro-Caribbean populations (WUWHS, 2016).

PATHOPHYSIOLOGY

The aetiology of HS is uncertain (Jianbing et al, 2013). It can be an acute or chronic disease and recurrence is common (Jianbing et al, 2013; Persaud et al, 2017). It is a skin disease of the hair follicles, specifically the follicular pilosebaceous unit (Beltrame and Staffolani, 2017; Persaud et al, 2017). The upper part of the hair follicles is occluded by keratinized stratified squamous epithelium that then progresses to dilation and rupture of the follicles with their contents deposited into the surrounding dermal tissue. This

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Figure 1. Armpit of a patient in phase III of HS, showing multiple interconnected sinus tracts and abscesses.

process initiates an inflammatory cascade with the attraction and deposition of white blood cells and subsequently abscess formation and destruction of the pilosebaceous unit (Ather et al, 2006; Zouboulis et al, 2015a).

HS is not a classic infectious disease there is no unique bacterial agent, it is a chronic disease with secondary infections of an initially sterile process, bacterial sampling of suppurations is frequently sterile. A distinctive feature of infection in HS is the lack of lymph node enlargement near the lesions despite inflammation and bacterial infection (Zouboulis et al, 2015a).

CATEGORISATION OF HS

Hurley's staging system has been used clinically since 1989 and is composed of three phases:

- ▶▶Phase I: single or multiple isolated abscesses without sinus tracts or scarring
- ▶▶Phase II: one or more recurrent abscesses, single or multiple widely separated lesions with sinus tract formation and scars
- ▶▶Phase III: multiple deep fistulas and abscesses, diffuse or broad involvement across a regional area with multiple interconnected sinus tracts and abscesses (*Figure 1*), secondary infection and fistulae may be present (Jianbing et al, 2013).

A criticism of the Hurley categorisation is that it is a static system and not sufficiently responsive to changes in the inflammatory component of HS (Gulliver et al, 2016).

The Sartorius staging system is said to be more sophisticated than Hurley's staging system especially when attempting to assess the treatment effects in a clinical trial situation. It was the first disease specific instrument for dynamically measuring the clinical severity of HS. The elements of this staging system include:

- ▶▶The anatomical region (axilla, groin, gluteal, inframammary or other regions)
- ▶▶The number and types of lesions involved (abscesses, nodules, fistulae/sinuses, scars)
- ▶▶The distance between lesions and the presence of normal skin in-between lesions (Sartorius et al, 2003).

The limitation of this staging system becomes apparent when the lesions coalesce and become confluent (Zouboulis et al, 2015a). A reduction in the Sartorius score demonstrates clinical efficacy

of HS treatments (Maarouf et al, 2017).

CLINICAL PRESENTATION OF HS

There is frequently a delay in diagnosing HS as it can be mistaken for simple infected lesions (Beltrame and Staffolani, 2017). An Irish study identified an average of 8.5 years from the onset of symptoms to receiving a diagnosis of HS, with the average age at diagnosis of 31 years (Delany et al, 2017). Clinicians are advised to use the following question to which a positive answer can identify patients with HS (sensitivity 90%, specificity 97%): "Have you had outbreaks of boils during the last 6 months with a minimum of 2 boils in one of the following 5 locations: axilla, groin, genitals, under the breasts and other locations, e.g. perianal, neck and abdomen?" (Zouboulis et al, 2015b).

There is no definitive histological or other test to diagnose HS, the diagnosis is made on the clinical examination and clinical presentation of the affected areas along with a medical history and presence of comorbidities (Saunte and Jemec, 2017). It is important to examine all apocrine gland-bearing areas of the body and not just those areas of reported lesions (Zouboulis et al, 2015a; Beltrame and Staffolani, 2017). Ultrasonography and MRI scans are helpful to establish abscess borders and the presence of fistulae (Zouboulis et al, 2015b).

Initially, HS may present with painful nodules that can suppurate and ulcerate and produce a foul odour (Alavi et al, 2017). The nodules are often referred to as boils by patients with HS (Zouboulis et al, 2015a). The nodules can coalesce and cover a wide area of skin. In addition to the nodules there may be sinus tracts, epithelial cysts, pyogenic granulomas and disfiguring scarring (Persaud et al, 2017).

Common sites for HS are groin, armpit, perineum, buttocks area and submammary /inframammary fold. More uncommon sites for HS include areola of the breast, periumbilical skin, scalp, zygomatic and malar areas of the face, thighs and popliteal fossa (WUWHS, 2016). The diagnostic criteria for HS are shown in *Table 1*.

Most patients will have the milder form of the disease (Hurley stage 1), with a third having Hurley stage II and only 4% having the more severe stage III (WUWHS, 2016).

The course of the disease is variable (Zouboulis et al, 2015b). Flare ups of the disease may be

Table 1. Diagnosing HS (adapted from Zouboulis et al, 2015b)

Obligatory criteria	
1	History: recurrent painful or purulent lesions more than twice/6 months
2	Location: groin, armpit, perineum, buttocks area and submammary intramammary fold (women)
3	Clinical signs: <ul style="list-style-type: none"> • Primary lesions like follicular papule/pustule (folliculitis), nodule (inflammatory or non-inflammatory), abscess • Secondary lesions like cyst, fistula/sinus (exudative or non-exudative), double pseudocomedone, scar (atrophic, net-like, erythematous, hypertrophic, linear or bridged)
Additional criteria (not obligatory)	
1	History: HS-positive family history
2	Microbiology: no evidence of pathogens or presence of normal skin microflora at the predominant primary type of lesions
Diagnosis of HS if	
<ul style="list-style-type: none"> • All three obligatory criteria are present • One or more obligatory locations are involved • One or more clinical signs are present (i.e. nodule, abscess, fistula/sinus, scar) 	

exacerbated by stress and heat (Ather et al, 2006). Eventually the disease burns itself out leaving scarred and fibrotic skin, with the average duration of the disease being 19 years (Ather et al, 2006).

DIFFERENTIAL DIAGNOSES

Due to the clinical similarities HS may be misdiagnosed as Staphylococcal infection (the lesions are spread in a random fashion and more pustular), Cutaneous Crohn’s disease (associated with intestinal Crohn’s), abscesses (usually a single lesion), primary or metastatic tumours (confirmed by histology), pilonidal and dermoid cysts and tuberculosis of the skin (Ather et al, 2006; Zouboulis et al, 2015b). However, squamous cell carcinoma can co-exist with HS and surgical intervention is recommended in these situations (Jourabchi et al, 2016).

RISK FACTORS AND COMORBIDITIES

HS is strongly associated with smoking and obesity (Saleem et al, 2017). In addition, comorbidities include arthropathy, inflammatory bowel disease and metabolic syndrome (Dini et al, 2015; Saunte

and Jemec, 2017). It is proposed that there is a genetic (family history) and hormonal aspect to the condition (Ather et al, 2006).

PSYCHOLOGICAL IMPACT

It is important to establish the effects of HS on an individual’s quality of life. The Dermatology Quality of Life Index (DLQI) is a non-specific tool in terms of unique disease characteristics that has been used in HS populations. However, Skindex-29 is a tool that measures the following; the impact of symptoms, emotion and functioning, and side effects of the treatment of a disease on quality of life. In HS Skindex-29 has identified the problems of pain and skin irritation in the physical domain and fatigue in the functional domain (WUWHS, 2016).

The chronicity of the disease results in embarrassment, self-consciousness, and social isolation. It is important to reassure the individuals that HS is not linked to poor hygiene and is not contagious (Ather et al, 2006).

Due to the distribution of the disease, it is associated with a lack of sexual health and difficulty with intimate relationships (WUWHS, 2016; Delany et al, 2017; Zouboulis et al, 2015b). The younger population suffering with HS report that it influenced their choice of clothing (Delany et al, 2017). The disease is said to be debilitating and has an adverse effect on an individual’s ability to function in employment and domestic roles (Nicoli et al, 2013; Patel et al, 2017).

In a recent Danish study of over 7,500 patients, Thorlacius et al (2018) identified an increased risk of completed suicide and a high prevalence of anxiety and depression in a HS population. There are support groups and information sites such as:

- ▶▶ The HS Trust
- ▶▶ The British Skin Foundation.

They provide help and guidance for HS sufferers (Malcolm et al, 2015).

TREATMENT OPTIONS

The choice of therapy is based on severity of the disease, types of lesions, extension of involved area and resistance to previous treatments (WUWHS, 2016). It is important to treat the comorbidities and offer lifestyle modification such as weight loss guidance and smoking cessation to improve the HS and reduce the associated cardiovascular risks



Figure 2. Armpit of a patient with healed HS.

(Zouboulis et al, 2015b; Saleem et al, 2017; Persaud et al, 2017).

Pain relief is a priority with a positive response to non-steroidal anti-inflammatory drugs (NSAID) however opioids may be used if the pain is not relieved by the NSAID (Persaud et al, 2017).

The first line of treatment is medical intervention. Topical clindamycin is advised for superficial and localised infection, progressing to systemic antibiotic therapy (often for prolonged periods) with clindamycin alone or in combination with rifampicin for cases of deep and surrounding infection. Additional specific medical interventions include the use of retinoids (isotretinoin), immunosuppressants (Cyclosporin), systemic corticosteroids (prednisone), other anti-inflammatory drugs (Dapsone) and hormonal therapy e.g. anti-androgens (Persaud et al, 2017). Further patient information on the applicability of these additional treatments can be found at <http://www.hstrust.org/treatments.html>.

The biologic drugs are making a significant impact on the treatment of HS. Humira adalimumab was the first drug approved by the USA Food and Drug Administration for the treatment of HS (Maarouf et al, 2017). This is a member of the group of drugs that inhibit tumor necrosis factor-alpha (TNF-alpha) which also includes etanercept and infliximab. TNF-alpha is a proinflammatory cytokine produced by white blood cells (Romanelli, 2015).

A Cochrane review examined which treatments have been shown to be effective in randomised clinical trials (RCTs) for HS in adults. They concluded that there is high-quality evidence of benefit from adalimumab given weekly and moderate-quality evidence suggests that infliximab is beneficial; RCT evidence for other interventions was lower in quality or absent, limiting further conclusions (Ingram, 2017).

Biologic drugs may be an effective treatment for severe HS, however recurrences often occur following discontinuation of therapy (Dini et al, 2015). Selection of suitable patients for biological treatment is imperative and helps to minimize the impact of adverse effects, especially with the high cost of the therapy (Romanelli, 2015).

If patients do not respond to medical intervention surgical treatment is the alternative option. Initially localised excision and drainage may be considered however this will not halt the progress of the disease

and is viewed as a temporary fix. The main surgical intervention is wide excision of the affected areas followed by healing by secondary intention (*Figure 2*). A survey of patients who had undergone wide excision and healing by secondary intention for HS reported that patients were glad they had undergone the surgery and would recommend it to other HS sufferers as a treatment option (Deckers et al, 2017). The surgery can be quite extensive depending on the amount of diseased tissue and the resulting scars may produce a poor cosmetic result, in these situations The Camouflage Service may be able to conceal the scarring <https://www.changingfaces.org.uk/skin-camouflage>. The service is part of the Changing Faces charity that has trained camouflage practitioners who find the best colour match for each person's skin tone and then teach them how to self-apply the specialist cover products. The products are fully waterproof and are available on prescription from a GP. There are over 80 clinics in England and Scotland, with some requiring a GP referral, and some operating with a self-referral system.

Negative pressure wound therapy has been used successfully in combination with wide excision (Jianbing et al, 2013). A small case series report successful treatment of HS with wide excision combined with topical autologous platelet-rich plasma and split-thickness skin grafts (Vossen et al, 2016).

Guidelines contain information on further treatments such as Skin Tissue-sparing Excision with Electrosurgical Peeling (STEEP) for Hurley's stage II and II presentations. Deroofing carbon dioxide laser treatment is suggested for mild to severe disease. It can be performed under local and general anesthetic and is said to result in less scarring and pain than conventional surgery (WUWHS, 2016).

The challenge when applying local wound dressings to areas affected by HS is trying to get a fit in difficult areas with body contours whilst managing the localised pain and wound odour (Alavi et al, 2017). The general principles of moist wound healing should be applied, and advice sought from the wound care team if the symptoms of pain, exudate and odour are not resolving. The literature also advises using antiseptic washes, not shaving or using deodorants and wearing loose clothing, the latter to avoid trauma and friction to the affected areas (Malcolm et al, 2015; Persaud et al, 2017).

Although the literature concentrates on the management of the physical aspects of HS, counselling and psychological support play an equivalent part in the management of this distressing disease.

CONCLUSION

HS is a chronic inflammatory recurrent debilitating disorder located in the apocrine-bearing aspects of the skin. It is a skin disease of the hair follicles initiated by occlusion, progressing to inflammation and destruction of the hair follicle and deposition of waste products in the adjacent dermis. The initial presentation is of nodules that are painful and may suppurate however bacterial sampling is frequently sterile. As the disease progresses the abscesses and nodules will increase in number and sinus tracts and fistulae may form. The disease may be contained to one area of the body or present at multiple sites.

HS is often misdiagnosed as other skin eroding/ulcerating conditions. There is a predominance of HS in the female population and strong associations with smoking and obesity. Hurley's staging system and The Sartorius staging system are methods of classifying the status of HS. There is no definitive histological or other test to formally diagnose HS, diagnosis is based on medical history, comorbidities and clinical presentation of the affected areas.

The physical problems of pain, itching and malodorous weeping lesions have a significant impact on the individual's quality of life.

The first line therapies consist of antibiotics, immunosuppressants, hormonal and biological therapies. Surgical intervention is considered if there is a poor response to the initial treatments. There are very few randomised controlled trials in this patient population with a large amount of the evidence to support individual interventions at the anecdotal and case series level. However, new developments in the field are starting to emerge.

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REFERENCES

- Alavi A, Farzanfar D, Lee, RK, Almutairi D (2017) The contribution of malodour in quality of life patients with hidradenitis suppurativa. *J Cutan Med Surg* 1203475417745826
- Ather S, Chan DSY, Leaper DJ, Harding KG (2006) Surgical treatment of hidradenitis suppurativa: case series and review of the literature. *Int Wound J* 3: 159–69
- Beltrame A, Staffolani S (2017) Clinical diagnosis of hidradenitis suppurativa. *N Engl J Med* 377(25):2474
- Deckers IE, Dahi Y, van der Zee HH, Prens EP (2017) Hidradenitis suppurativa treated with wide excision and second intention healing: A meaningful local cure rate after 253 procedures. *J Eur Acad Dermatol Venereol* doi:10.1111/jdv.14770
- Delany E, Gormley G, Hughes R et al (2017) A cross-sectional epidemiological study of hidradenitis suppurativa in an Irish population (SHIP). *J Eur Acad Dermatol Venereol* doi:10.1111/jdv.14686
- Dini V, Oranges T, Romanelli M (2015) Hidradenitis suppurativa and wound management. *Int J Low Extrem Wounds* 14(3):236–44
- Ingram JR (2017) Interventions for hidradenitis suppurativa: Updated Summary of an original Cochrane Review. *JAMA Dermatol* 153(5): 458–59
- Gulliver W, Zouboulis CC, Prens E et al (2016) Evidence-based approach to the treatment of hidradenitis suppurativa/acne inversa, based on the European guideline for hidradenitis suppurativa. *Rev Endocr Metab Disord* 17(3):343–51
- Jianbing T, Biao C, Qin L, Yanhong W (2013) Topical negative pressure coupled with split-thickness skin grafting for the treatment of hidradenitis suppurativa: a case report. *Int Wound J* 12(3):334–7
- Jourabchi N, Fischer AH, Cimino-Mathews A et al (2016) Squamous cell carcinoma complicating a chronic lesion of hidradenitis suppurativa: a case report and review of the literature. *Int Wound J* 14(2):435–38
- Maarouf M, Clark AK, Lee DE, Shi VY (2017) Targeted treatments for hidradenitis suppurativa: a review of the current literature and ongoing clinical trials. *J Dermatolog Treat* 10: 1–9 doi: 10.1080/09546634.2017.1395806
- Malcolm C, Javed MU, Dickson (2015) *Support Groups for Hidradenitis Suppurativa*. Available at: <https://www.thepmfajournal.com/features/post/support-groups-for-hidradenitis-suppurativa> (1.03.2018)
- Nicoli F, Balzani A, Lazzeri D, Gentile P et al (2013) Severe hidradenitis suppurativa treatment using platelet-rich plasma gel and Hyalomatrix. *Int Wound J* 12(3):338–43
- Patel ZS, Hoffman LK, Buse DC et al (2017) Pain, Psychological Comorbidities, Disability, and Impaired Quality of Life in Hidradenitis Suppurativa. *Curr Pain Headache Rep* 21(12):49
- Romanelli M (2015) Biological drugs in wound management. *J Wound Care* 24(6):243
- Saunte DML, Jemec GBE (2017) Hidradenitis suppurativa advances in diagnosis and treatment. *JAMA* 318(20):2019–32
- Persaud R, Elliott JA, Goodman L et al (2017) Hidradenitis suppurativa: A retrospective review of 13 patients and literature summary. *EMJ Dermatology* 5(1):90–97
- Saleem MD, Arnold DL, Feldman SR (2017) Hidradenitis and smoking. *Br J Dermatol*. doi:10.1111/bjd.16214
- Sartorius K, Lapins J, Emtestam L, Jemec GB (2003) Suggestions for uniform outcome variables when reporting treatment effects in hidradenitis suppurativa. *Br J Dermatol* 149(1):211–3
- Thorlacius L, Cophen AD, Gislason GH et al (2018) Increased suicide risk in patients with hidradenitis suppurativa. *J Invest Dermatol* 138(1):52–7
- Vivar KL, Kruse L (2017) The impact of pediatric skin disease on self-esteem. *International Journal of Women's Dermatology* 4(1): 27–31
- Vossen ARJV, van der Zee HH, Prens EP (2016) Letter to the Editor: Accelerated wound healing after wide excisions in hidradenitis suppurativa using autologous split-thickness skin grafting and platelet-rich plasma. *International Wound Journal* DOI: 10.1111/iwj.12673
- World Union of Wound Healing Societies (2016) *Understanding Hidradenitis Suppurativa*. Available at: http://www.wuwhs2016.com/files/WUWHS_HS_Web.pdf (accessed 1.03.2018)
- Zouboulis CC, Desai N, Emtestam L et al (2015a) European S1 guideline for the treatment of hidradenitis suppurativa/acne inversa. *J Eur Acad Dermatol Venereol*. 29(4): 619–44
- Zouboulis CC, Del Marmol V, Mrowietz U et al (2015b) Hidradenitis suppurativa/acne inversa: Criteria for diagnosis, severity assessment, classification and disease evaluation. *Dermatology* 231(2): 184–90