# Hidradenitis suppurativa: A comprehensive review

Ali Alikhan,[a] Peter J. Lynch, MD,[b] and Daniel B. Eisen, MD[b]
*Sacramento, California*

Hidradenitis suppurativa, also known as acne inversa, is a chronic, often debilitating disease primarily affecting the axillae, perineum, and inframammary regions. Prevalence rates of up to 4% have been estimated. Our understanding of the disease has changed over time. It is now considered a disease of follicular occlusion rather than an inflammatory or infectious process of the apocrine glands. Clinically, the disease often presents with tender subcutaneous nodules beginning around puberty. The nodules may spontaneously rupture or coalesce, forming painful, deep dermal abscesses. Eventually, fibrosis and the formation of extensive sinus tracts may result. The location of the lesions may lead to social embarrassment and the failure to seek medical treatment. Therapies in the past have consisted of long-term antibiotics, antiandrogens, and surgery. New treatments like tumor necrosis factor—alfa inhibitors have given clinicians more options against this difficult disease. (J Am Acad Dermatol 2009;60:539-61.)

***Learning objectives:*** After completing this learning activity, participants should be able to describe the clinical presentation, demographics, and prevalence of hidradenitis suppurativa, be familiar with current controversies regarding the pathogenesis of this complex, and be able to discuss potential treatments and their outcomes.

***Key words:*** acne inversa; etanercept; hidradenitis suppurativa; infliximab; Verneuil disease.

**Key points**

- **HS is a chronic, recurrent, inflammatory disease presenting as painful subcutaneous nodules**
- **Double comedones, deep sinus tracts, and abscesses are characteristic for HS**
- **A diagnosis of HS is made clinically without the use of laboratory tests**

*Abbreviations used:*

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| AD: | autosomal dominant |
| CD: | Crohn disease |
| CNS: | coagulase-negative Staphylococcus |
| DDD: | Dowling—Degos disease |
| DLQI: | Dermatology Life Quality Index |
| FFD: | Fox—Fordyce disease |
| HS: | hidradenitis suppurativa |
| IK: | interstitial keratitis |
| KID: | keratitis-icthyosis-deafness |
| OC: | oral contraceptive |
| SAPHO: | synovitis, acne, pustulosis, hyperostosis, and osteitis |
| SCC: | squamous cell carcinoma |
| TLR: | Toll-like receptor |
| TNF: | tumor necrosis factor |

Hidradenitis suppurativa (HS), also known as acne inversa, is a chronic, recurrent, inflammatory disease, initially presenting as tender subcutaneous nodules. These lesions may spontaneously rupture or coalesce to form deep dermal, exquisitely painful abscesses.[1] The abscesses have typically been described as being deep and rounded without central necrosis or pointing (though this may sometimes occur), and they often exude a purulent drainage.[1,2] The inflammatory abscesses ultimately heal, producing fibrosis, dermal contractures, and induration of the skin.[3] The disease is insidious, typically developing in otherwise healthy postpubertal males and females.[4]

Another lesion typical in HS is the double comedone, a blackhead with multiple apertures that communicate below the skin.[5] These abnormal comedones have been reported on flexural surfaces of children, and may be the precursors of HS lesions.[6] The disease process is thought to begin with occlusion of hair follicles. The occluded follicles subsequently rupture and reepithelize, resulting in sinus tracts that can house foreign material and bacteria.[5] Over time, these tracts can coalesce into large regions of subcutaneous honeycombing.[7] They can potentially dissect into deep structures including muscle, fascia, lymph nodes, the urethra, and the

From the Department of Dermatology,[b] University of California—Davis School of Medicine.[a]
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Reprint requests: Daniel B. Eisen, MD, University of California—Davis School of Medicine, Department of Dermatology, 3301 C St, Ste 1400, Sacramento, CA 95816. E-mail: deisen@ucdavis.edu.
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bowels.[1,2,8] They release a serous to purulent malodorous discharge, resulting in both soiling of clothes and social embarrassment.[9,10] Ulceration of the skin has also been described with chronic HS.[3] The ulcers characteristically present with rolled, boggy, ragged edges and clean granulating bases.[3] Like most diseases, especially its analogue acne conglobata, the severity varies considerably.

Early symptoms and signs of HS are burning, pruritus, local warmth, hyperhidrosis, and pain.[3,11] HS may be divided into acute and chronic forms. Acute HS presents with a few deep-seated nodules, which may coalesce developing into cord-like red-to-bluish red structures.[3,12] Chronic HS is characterized by multiple abscesses, internetworking sinus tracts, foul exudate from draining sinuses, inflammation in the dermis, both atrophic and hypertrophic scars, ulceration, and infection which may extend deep into the fascia.[10,12]

The diagnosis of HS is made clinically, and biopsies are not routinely taken.[13] There is no confirmatory test for HS. Criteria for diagnosis vary widely but typically include chronicity, recurrence of disease, failure to completely clear with antibiotics, characteristic multifocal lesion distribution, sinus formation, scarring and dermal contracture, presence of double comedones, multiple open comedones, nodules, papules, tenderness of lesions, and malodorous discharge.[7]

## DEMOGRAPHICS

### Key points

- **HS tends to begin in the second or third decades of life**
- **Females are more likely to develop HS**
- **Recent studies suggest a prevalence of about 1%**

HS typically occurs after puberty, with average age of onset in the second or third decades of life. Onset after menopause is rare.[14,15] Isolated case reports have described HS in prepubertal subjects, but these are associated with premature adrenarche.[16-18] HS prevalence may diminish over time. A recent study showed that prevalence among those 55 years of age and older was significantly lower than in younger age groups (0.5% vs 1.4%).[19]

A large Danish study estimated 1-year prevalence at 1.0% based on subject recollection only, and point prevalence at 4.1% based on objective findings.[20] The point prevalence was based on a younger adult population and may, therefore, overestimate the true prevalence of HS.[7,20] A recent case control study based on a representative portion of the French population (n = 10,000) estimated prevalence at 1%.[19] Other studies have estimated various prevalence rates ranging from 0.00033% to 4%.[20-23]

HS appears to be significantly more common among females. HS researchers generally accept this, but many authors have argued that certain locations appear to have sexual predilection.[2,7,8,11,13,15,17,24-28] Perianal HS, for instance, seems to affect males more than females.[3]

Some authors have suggested a higher prevalence of HS among African Americans. This has not been substantiated by any studies examining racial predilection.[12,15,24,29-31]

## CLINICAL PRESENTATION

### Key points

- **HS most typically occurs in the axillary, inguinal, perianal, perineal, mammary, and inframammary regions**
- **The distribution pattern corresponds with the "milk line" distribution of apocrine-related mammary tissue in mammals**
- **The most commonly affected site is the axilla**
- **Perianal HS is associated with more debilitating outcomes**

HS is characterized initially by the presence of tender subcutaneous nodules. With time, the nodules may rupture, resulting in painful, deep dermal abcesses.[1] The lesions may be rounded, but, unlike furuncles, do not usually exhibit pointing. After rupture, the lesions often extrude a purulent, foul-smelling discharge.[1,2] As the disease process continues, fibrosis, dermal contractures, and induration of the skin occur. The presence of double comedones is typical of the disease. HS develops gradually in postpubertal males and females who are otherwise healthy.[13]

HS lesions occur most frequently in the intertriginous apocrine gland–bearing areas of the axillary, inguinal, perianal, perineal, mammary, inframammary, buttock, pubic, chest, scalp, retroauricular, and eyelid areas.[1,5] This distribution pattern corresponds for the most part with the "milk line" distribution of apocrine-related mammary tissue in mammals. The sites affected in HS correspond not only to the location of apocrine glands in the body but also to that of terminal hair follicles dependent on low androgen concentrations.[32] One study with 388 HS patients demonstrated axillary involvement in 278 patients (72%), perianal involvement in 125 patients (32%), groin involvement in 92 patients (24%), and mammary involvement in 32 patients (8%).[33,34] Similar rates were seen in a study by Barth et al.[35] Perineal HS is associated with a much higher rate of recurrence (74% in one study) and more