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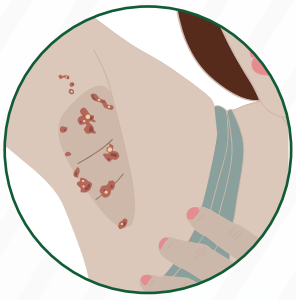
Hidradenitis Suppurativa

Understanding its epidemiology, pathophysiology, and clinical presentation



Hidradenitis suppurativa (HS), also known as acne inversa, is a chronic inflammatory skin disorder[1,2]

Manifests as painful lesions, abscesses, and scars

Primarily in intertriginous areas (axillary, groin, perianal, and perineal regions)

Prevalence and epidemiology of HS

Global estimates[2] 0.03% to 4.0%

In Australia[2] 0.67%

HS is more common among:

- Women
- Young people
- People with obesity
- Individuals who smoke
- People with low annual income

Risk factors of HS[1]

Genetic

- 33% to 40% have an affected first-degree relative
- Associated with mutations in γ-secretase complex, *DCD*, *PSTP1P1*, *SOX9*, and *KLF5*

Environmental

- Medication
- Cutaneous microbiome dysregulation
- Mechanical friction

Behavioural

- Smoking

Additional triggers

- Hormonal changes during menstruation and menopause
- Stress
- Excessive heat and perspiration
- Weight gain

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Clinical presentation of HS[1]

Symptoms preceding lesions

- Burning, pain, pruritus, warmth, and hyperhidrosis

Lesions

- Deep-seated nodules 0.5 to 2 cm (persisting for days to months)
- May have serosanguinous discharge
- Recurrent nodules, prone to rupture
- Tunnels and tracts that may ulcerate

Clinical classification of HS[1]

Hurley stage I

- Abscesses lack tunnels and scars

Hurley stage II

- Recurrent abscesses with tunnels and scars
- Single or multiple lesions widely separated

Hurley stage III

- Diffusely involves the skin
- Lesions with multiple interconnected sinus tracts
- Abscesses occupy large areas

HS-associated comorbidities[1,3]*

*Percentages in Australian patients[3]

- 52% Acne vulgaris or acne conglobata
- 42% Depression
- 16% Diabetes mellitus
- 45% Hyperlipidaemia
- 14% Hypertension
- 39% Insulin resistance
- 12% Keratosis pilaris
- 61% Obesity
- 27% Pilonidal sinus
- 16% Polycystic ovarian syndrome

Dissecting scalp cellulitis | Inflammatory bowel disease | Metabolic syndrome | Spondyloarthritis

Impact on quality of life[1,4]

Physical discomfort

- Pain
- Drainage
- Malodour
- Bleeding
- Scarring
- Difficulty moving limbs

Psychosocial impact

- Depression
- Social isolation (embarrassment and self-consciousness)
- Employment difficulties
- Sexual dysfunction
- Higher rate of suicide

Proper disease management and psychosocial support are essential for improving the quality of life of patients with late-stage HS[4]

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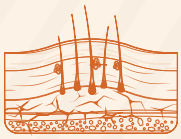
Pathophysiology of HS[1,5,6]

The exact causes and pathogenic mechanisms underlying HS development and progression are only partially understood[5]

Early HS[6]



- Infiltration of neutrophils, innate immune cells, and lymphocytes
- Follicular hyperkeratosis and plugging
- Sebum accumulation and secretion
- Keratin debris causing folliculitis
- Bacterial dysbiosis

Advanced HS[1,6]

- Rupture of inflamed follicles
- Advanced perifolliculitis
- Dermal inflammation
- Infiltration of immune cells

Chronic, uncontrolled inflammation driven by[5]:

- Innate immune cytokines
- B cells
- Neutrophil extracellular traps
- Complement activation
- Damaging autoantibodies

- Abscess formation
- Dermal tunnels with discharge
- Tissue destruction and scarring

HS diagnosis[1]

Clinical diagnostic criteria[1,2]

Types of lesions

Characteristic nodules, abscesses, tunnels, and scars

Location

Intertriginous areas

Chronicity

Histopathological features of HS include[1]:

- Follicular occlusion
- Follicular hyperkeratosis
- Infundibular follicular epithelial hyperplasia
- Epidermal psoriasiform hyperplasia

- Keratin plugging
- Perifolliculitis (predominantly lymphocytic)
- Plasmacytic infiltrate
- Pseudoepitheliomatous hyperplasia

Differential diagnosis of HS[1]

Conditions to be ruled out

- Follicular pyoderma (including folliculitis, furuncles, and carbuncles)
- Granuloma inguinale
- Nodulo-ulcerative syphilis
- Tuberculous abscess
- Actinomycosis
- Lymphogranuloma venereum
- Acne vulgaris
- Epidermoid, dermoid, pilonidal, or Bartholin's cysts
- Crohn's disease (particularly, with perianal involvement)

Conditions to be included

- Granulomatous diseases (Crohn's disease, sarcoidosis, mycobacterial infection)
- Chronic deep folliculitis
- Langerhans cell histiocytosis
- Folliculotropic mycosis fungoides
- Pyoderma gangrenosum
- Follicular rupture
- Abscess

Multiple other conditions with similar histopathology, including squamous cell carcinoma, need to be ruled out through biopsy during the HS diagnosis[1]

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Challenges to HS diagnosis

Symptom onset → HS diagnosis: 7.23 ± 2.81 years[2,7]

6.8% In Australia, diagnosis rates are low[2]

Reasons for delayed diagnosis[2,7]

- Challenges to correctly diagnose HS at early stages
- Decentralisation of care
- Clinicians' unfamiliarity with the disease
- Misdiagnosis by clinicians
- Patients delaying seeking medical care
- Poor access to dermatology care

Blood, microbiological, histological, and imaging analyses using ultrasonography and magnetic resonance imaging may help in the accurate and early diagnosis of HS

Interdisciplinary management of HS[1]

Treatment

- Antibiotics (topical, oral, and intravenous)
- Anti-inflammatory medications
- Antiandrogen agents
- Metformin
- Pain management

Biologic therapy

- Anti-TNF-α antibody (adalimumab)
- IL-17 inhibitors (secukinumab and bimekizumab)

Surgical therapy

- Laser therapy
- Punch debridement
- Deroofing
- Excision with skin grafting

Lifestyle modifications

- Maintaining a healthy weight
- Smoking cessation
- Avoiding tight clothing, harsh cleansers, and adhesive dressings

Psychosocial support

- Patient education, counselling, and support
- Important to reassure patients that the condition is non-contagious and not caused by poor hygiene

TNF: Tumour necrosis factor; IL: Interleukin

Key messages

- Increased awareness of the differential diagnosis of HS is necessary among the medical community to improve diagnosis, reduce delays in diagnosis, and provide timely treatment
- Chronic relapsing inflammation is the main pathogenic mechanism driving HS progression and an improved understanding of the complex interactions can help identify potential therapeutic targets
- Interdisciplinary management and support for the psychosocial aspects of HS are needed to improve the physical functioning and emotional well-being of patients with HS

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