International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

# Epidemiological and Clinical Analysis of Hydradenitis Suppurativa in Maharashtra: A Case Series Review

**Tushar Kinjalk[1], Aarushi Kinjalk[2], Nishindra Kinjalk[3]**

[1]Junior Resident, Department of General Surgery, Dr D Y Patil Hospital, Navi Mumbai

[2]Consultant in Dermatology, Pune

[3]Associate Professor, Department of Medicine, Narayan Medical College, Rohtas

**Abstract:** ***Hidradenitis suppurativa (HS) is a chronicinflammatory and debilitating skin condition. HS, also known as acne inversus, is characterized with lesions including deep – seated nodules, abscesses, sinuses, and scars. These are common in the intertriginous areas, and the areas rich in apocrine glands;usually in axilla, groin, perineal, gluteal and perianal areas. With the prevalence of 1% to 4% with painful course HS is often underdiagnosed or diagnosed late. Its brittle nature of response to the treatment modalities makes it a challenge to clinicians of different specialties. This paper reviews the available literature on HS. It analyzes five peculiar cases from the outpatient clinics across Maharashtra, with an aim to help understand this disease better, and help improve patient care.***

**Keywords:** Hidradenitis suppurativa, acne inversus, Hurley staging system, Verneuil's disease

## 1. Introduction

Hidradenitis suppurativa (HS) is a chronic, painful, follicular and occlusive disease of skin. It affects usually the folliculopilosebaceous units.[1] It is common in intertriginous areas, axilla, groin, perianal, perineal, genital, gluteal and inframammary areas.[2] The clinical course is variable, ranging from a mild disease characterized by the recurrent appearance of papules, pustules, and a few inflammatory nodules to the cases demonstrating deep, fluctuant abscesses, draining sinuses, and rope-like scars.[3]

Hidradenitis suppurativa (Greek *hidros* meaning sweat, and *aden* meaning glands) is also known as Verneuil's disease or acne inversus.[4] It is referred to ashidradenitis also.[5]

With probable genetic, environmental, hormonal, and psychosomatic influences, the cause of HS remains unknown. 35% to 40% of HS cases have a historyof HS in a first degree relative. Women are more prone to suffer from HS, and good number of cases are reported at puberty.[6]

**Pathophysiology**: A defective hair follicle that becomes occluded, ruptures spilling its contents into dermis. The spill contains keratin and microbes. Lymphocytic and neutrophilic response may lead to abscess formation and further damage to the pilosebaceous units and adjacent structures.[7] Another view is that abnormal antimicrobial peptides, abnormal secretion of apocrine glands and abnormal invaginations of the epidermis leads to the tract formation and deficiency in function of sebaceous glands.[8] Abnormal immune behaviour has also been implicated with the evident elevated levels of cytokines, tumor necrosis factor- alfa (TNF) and several interleukins (IL). Bacteria as such, are not the causative organisms; and usually an aspirate from the lesion is sterile to bacterial culture. However, bacterial infection and colonization during the course of the disease may worsen the condition. [9]

Two main observations point to a genetic background of HS. First, approximately one third of HS patients have at least one family member also suffering from HS, suggesting an inheritable genetic predisposition. Second, families with several mutations or changes in genes of the gamma-secretase family [including nicastrin (NCSTN), presenilin 1 (PSEN1), presenilin enhancer 2 (PSENEN) gene mutation] show an autosomal dominant inheritance of HS, indicating a pathogenic role of these genes in HS. The resulting alteration in γ-secretase function might result in HS by affecting downstream Notch signalling in the skin. However, these patients show a severe disease phenotype not entirely typical of "sporadic" HS, raising questions as to what degree this genetic background is representative for all cases of HS. Current evidence supports a genetic predisposition for HS development.[10]

The pain, drainage, malodour, and disfigurement associated with HS, all contribute to the remarkable psychosocial impact of the disease, with negative effects on the patients' quality of life.[11]

**Cause of Pain**: nociceptive pain likely to be caused by inflammation causing peripheralneuroplastic changes and central sensitization.

**Cause of malodour**: This is commonly due to sweat, bacteria and skin tissues that are inflamed; with or without infection.

**Duration of a flare up:** A typical painful nodule usually lasts for 7-15 days. Usual recovery time of a flare up: 4-8 weeks. Smaller wounds heal more quickly, and with less scarring.

**Predisposing factors**[13]

- Obesity
- Smoking

- Weather
- Tight fitting clothes
- Stress
- Diet
- Vit D deficiency

**Trigger factors**[14]

- Smoking
- Obesity
- Sweating
- Shaving affected areas
- Skin irritation or injury
- Tight fitting clothes
- Changes in hormone levels, including menstruation
- Heat, humid and excessive sweating
- Dairy

**Comorbidities (screening recommended)** [15]

- PCOS
- Spondylarthropathy
- Keratitis- icthyosis-deafness (KID) syndrome
- Down's syndrome
- Dowling – Degos- Disease
- Pachyonychia congenita
- Adamantiades – Behcet disease (ABD)
- SAPHO (synovitis, acne, palmoplantar pustulosis, hyperosteosis, osteitis) syndrome

While dealing with a suspected case of HS, it is important to consider the predisposing factors, trigger factors and the comorbidities.

## 2. Case Series

**Case 1**

26 Mnondiabetic.Presented with a solitary swelling in left axilla. No obvious trigger factors like trauma, radiation or instrumentation were reported.The swelling had been recurringfor 8 years. Previous flare up was one year ago, lasting for 4 months.

**On examination:**BMI 22.7 kg/m$^2$. Single swelling in the axilla witha discharging sinus. (Figure 1 and Figure 2).

**Lab reports**:

Complete blood count (CBC): Haemoglobin (Hb) 13.7g/dL, TLC 13080 / cmm, DLC P64 L30 E 1 M4 B01. There was a left shift in the neutrophils. Platelets were adequate. Red blood cells (RBCs) showed anisocytosis. Target cells were present.

Fasting plasma glucose (FPG) 96 mg/dL, glycated haemoglobin (HbA1c) 5.4%. Kidney function tests (KFT), liver function tests (LFT) and thyroid function tests (TFT) were normal.

HbsAg, HIV and HCV markers were non-reactive.

The purulent discharge from the lesion on culture, grew *Staphylococcus aureus*, sensitive to ciprofloxacin.

**Treatment:** Oral ciprofloxacin 500mg twice daily for five days, followed by oral clindamycin 300mg twice daily for three months.Good response to the treatment.

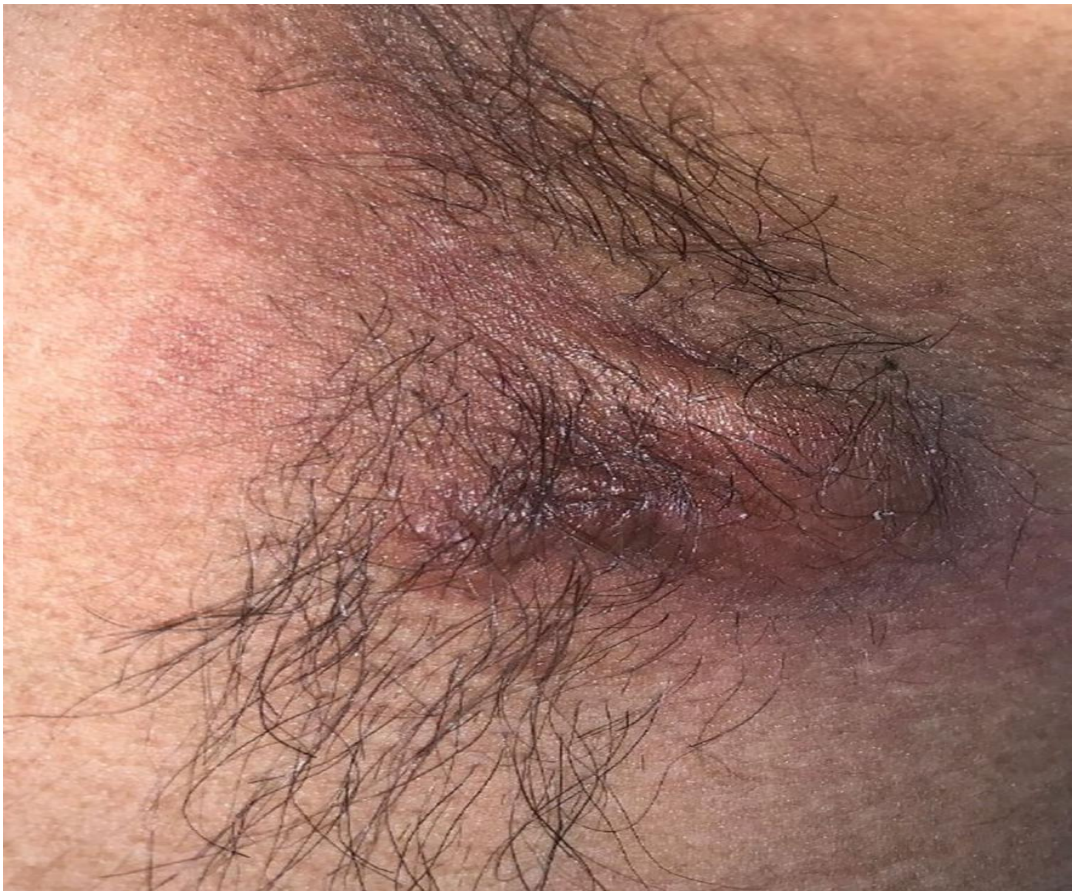

**Figure 1:** Swelling in the Axilla

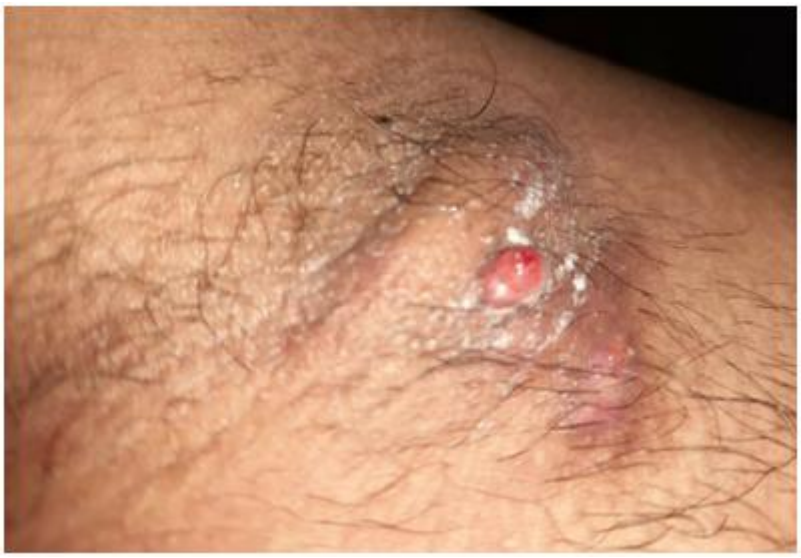

**Figure 2:** Discharging sinus from the lesion in axilla.

**Case 2**
31 Fnondiabetic. Presented with multiple swellings in both armpits. No reports of any trigger factors.
These swellings started appearing 10 Y ago, with frequent recurrence. Last flare up 8 months ago,lasting for 2 months.

**On examination:** BMI 22.7 kg/$m^2$. Multiple swellings in both axilla with discharging sinuses.

**Lab reports:**
CBC: Hb 14.4 g/dL, TLC 14040 /cmm, DLC P72 L24 E 1 M3. There was a left shift in the neutrophils. Platelets were adequate. RBCs showed anisocytosis. Target cells were present.
FPG 84 mg/dL, HbA1c 5.1%. KFT, LFT and TFT were normal.
HbsAg, HIV and HCV markers were non-reactive.
The purulent discharge from the lesion on culture, grew *Staphylococcus aureus*, sensitive to ciprofloxacin.

**Treatment**: Oral ciprofloxacin 500mg twice daily for five days, followed by oral clindamycin 300mg twice daily for three months. She showed good response to the treatment.

**Case 3**
22 M nonsmoker, nondiabetic. Referred to the skin OPD for evaluation of a congenital melanocytic lesion on the back, which was found on a routine health check up. A diagnosis of congenital nevus was made. Subsequent few months, hevisitedthe skinOPD for treatment of acne vulgaris. Isotretinoin was the mainstay of his acne management.

He laterdeveloped perineal folliculitis that failed to respond to topical antibiotics. Then he developed obvious signs of HS. No case of HS had been reported in his family earlier, and he had never been diagnosed with conditions known to be associated with HS.More specifically, he had no past medical history of arthritis or pyoderma gangrenosum.

**On examination:**
BMI 27.8 kg/m2. The physical exam showed inflammatory nodules and abscesses in the axilla. Similar inflammatory nodules were also found on his thighs, on the medial side. One swelling in the axilla with discharging sinus. (Image 1 and Image 2).

**Lab reports:**
CBC: Hb 14.6 g/dL, TLC 13700 /cmm, DLC P60 L35 E 1 M4. There was a left shift in the neutrophils. Platelets were adequate. RBCs showed anisocytosis. Target cells were present.
FPG 92 mg/dL, HbA1c 5.2%. KFT, LFT and TFT were normal.
HbsAg, HIV and HCV markers were non-reactive.
The purulent discharge from the lesion on culture, grew Staphylococcus aureus, sensitive to ciprofloxacin.
Treatment: Oral ciprofloxacin 500mg twice daily for five days, followed by oral clindamycin 300mg twice daily for three months. He+ showed good response to the treatment schedule.

**Case 4**
28 Fnon-smoker, nondiabetic.She presented to thesurgical OPD with a single swelling in the axilla. The swelling had been appearing on and off for a period of 5 years. The last flare up persisted for 2 months.

There was no history of trauma, radiation exposure and instrumentation, prior to present flare up.

**On examination:** BMI 23.3 kg/m2. Single swelling in the axilla discharging sinus.

**Lab reports:**
CBC: Hb 12,5 g/dL, TLC 13200/cmm, DLC P59 L35 E 1 M5. There was a left shift in the neutrophils. Platelets were adequate. RBCs showed anisocytosis. Target cells were present. PCV 32%.
FPG 84 mg/dL, HbA1c 5.1%. KFT, LFT and TFT were normal.
HbsAg, HIV and HCV markers were non-reactive.

The purulent discharge from the lesion on culture, grew Staphylococcus aureus, sensitive to ciprofloxacin.

**Treatment:** Oral ciprofloxacin 500mg twice daily for five days, followed by oral clindamycin 300mg twice daily for three months. She showed good response to the treatment.

**Case 5**
31 M nonsmoker, nondiabetic. Hepresented with a solitary lump in the left axilla. It was slightly tender, and pus exuded from several openings. No history of trauma, instrumentation, and radiation prior to this presentation.

**On examination:**
BMI 24.3 kg/m2. Physical examination showed single swelling in the axilla with pus discharge.

**Lab reports:**
CBC: Hb 13.2 g/dL, TLC 12800 /cmm, DLC P55 L39 E 2 M4. There was a left shift in the neutrophils. Platelets were adequate. RBCs showed anisocytosis. Target cells were present. PCV 34%.
FPG 84 mg/dL, HbA1c 5.1%. KFT, LFT and TFT were normal.
HbsAg, HIV and HCV markers were non-reactive.

The purulent discharge from the lesion on culture, grew *Staphylococcus aureus*, sensitive to ciprofloxacin.

**Treatment:**
Oral ciprofloxacin 500mg twice daily for five days, followed by oral clindamycin 300mg twice daily for three months. He was showing good response to the treatment.

## 3. Discussion

HS is a challenge to the clinicians. This condition, in different stages of presentation may be seen in the OPD of dermatology, surgery, medicine or gynaecology. Before contemplating the treatment schedule, it is important to assess the extent and severity of the condition, along with the comorbidities. The management is comprehensive, including life style modification, stress management, pharmacotherapy and surgical intervention.

**Assessment of HS**
Hurley Stages of HS**:** Based on the severity, HS is classified into three clinical stages.[16]

- Hurley stage I: A single bump with no sinus tracts.
- Hurley stage II: More than one bump but little tunnelling.
- Hurley stage III: Multiple bumps with a lot of sinus tracts and scars, involving an entire area of the body.

**Differential Diagnosis**
It is important to consider the clinical conditions which may mimic the features of HS.[17]

- Granuloma inguinale
- Follicular pyoderma
- Acne vulgaris
- Tubercular abscess
- Lymphogranuloma venereum (LGV)
- Crohn disease (perianal involvement)
- Noduloulcerative syphilis
- Epidermoid, dermoid, Bartholin, or pilonidal cysts

**Management**
Early diagnosis and early treatment is the key. Important ingredients of management, are as follows.

1) **Lifestyle modification:**Exercise, weight loss & dietary modification.Advisable to avoid sugary drinks, cakes, sweets, ice cream, processed food.
2) **Avoidance of trigger factors**
3) **Pharmacotherapy**
   a) **Antimicrobials:** Ciprofloxacin, clindamycin, tetracycline, doxycycline, rifampicin
   b) **Hormones:** Spironolactone, metformin, finasteride
   c) **Immunomodulation:** This is becoming popular for moderate-to-severe form of the disease. Targeting the tumor necrosis factor (TNF), interleukin 1 (IL-1), IL-12, and IL-23 has been considered as potential therapies.

Adalimumab is recommended as the first-line biologic therapy for moderate-to-severe HS, followed by Infliximab and anakinra as second- and third-line options, respectively.The recommended dose of adalimumab in HS is 160 mg on Week 1, 80 mg on Week 2 and then 40 mg weekly. Recent studies suggest that those patients who do not respond to this therapy within 12 weeks ( <25% improvement in inflammatory nodules and abscesses) should discontinue the drug. However, the patients with partial or good response should continue the therapy with periodic assessment.

Evidence is however lacking for other biologic therapies, and any clinical decision should be based on close monitoring of benefit and risk assessments. Most biologics are well-tolerated and show a favourable safety profile when used for a short period. However, long-term safety concerns, like infection risks, development of malignancy and demyelinating disorders should be kept in mind.

Risk-benefit assessment is vital in such cases. [18,19]

**Surgery**: Any plan for a surgical intervention must be discussed with the patient. An informed consent must be taken before contemplating a surgery.[20]

a) Local excision: Local excision of single lesions is only suggested in well-circumscribed, localized cases of Hurley I-II.
b) Surgical drainage: For painful abscesses, no medical therapy are usually effective. Surgical drainage might be indicatedhere to relieve pain.However, this should not be considered as the only treatment, because the recurrence in majority of patients, isinevitable.
c) Extensive excision and/ electrosurgical excision/ CO2 laser: These surgical interventions, with or without reconstruction, could be appropriate for chronic lesions to prevent recurrence.[21]
d) Step by step surgery: A new surgical approach has been described recently, called the step-by-step surgery. It consists of consecutively removing portions of HS skin with secondary intention healing. [22]

Wound healing, following surgery may be through primary closure, delayed primary closure, secondary intention, and with grafts, flaps or skin substitutes. Continuing medical

therapy in the perioperative period might be beneficial, and reduce risk of postoperative complications.[23]

**Stress management:** This is a crucial part of the therapy of HS. The reduction in the stress reduces the flare ups and helps in better treatment. Some modalities are, as follows.

a) Yoga
b) Mindfulness
c) Music therapy: In music therapy, precision of selection of music, is vital.

## 4. Conclusions

The study highlights the complex and chronic nature of Hidradenitis Suppurativa (HS), emphasizing the need for early diagnosis and comprehensive management approaches. It underlies the importance of individualized patient care and considers both medical and surgical interventions based on disease severity.

**Informed Consent**
The informed consent had been obtained from the patients to present this information and the picture.

**Conflict of Interests**
The authors have no conflicts of interest to declare.

**Funding Sources**
The authors have no funding sources to declare.

## References

[1] Miller IM, McAndrew RJ, Hamzavi I. Prevalence, Risk Factors, and Comorbidities of Hidradenitis Suppurativa. Dermatol Clin. 2016 Jan;34(1):7-16. [PubMed]
[2] Seivright J, Collier E, Grogan T, Shih T, Hogeling M, Shi VY, Hsiao JL. Pediatric hidradenitis suppurativa: epidemiology, disease presentation, and treatments. J Dermatolog Treat. 2022 Jun;33(4):2391-2393. [PMC free article] [PubMed] [Reference list]
[3] Kromann CB, Deckers IE, Esmann S, Boer J, Prens EP, Jemec GB. Risk factors, clinical course and long-term prognosis in hidradenitis suppurativa: a cross-sectional study. Br J Dermatol. 2014 Oct;171(4):819-24. [PubMed] [Reference list]
[4] Zouboulis VA, Zouboulis KC, Zouboulis CC. Hidradenitis Suppurativa and Comorbid Disorder Biomarkers, Druggable Genes, New Drugs and Drug Repurposing-A Molecular Meta-Analysis. Pharmaceutics. 2021 Dec 26;14(1) [PMC free article] [PubMed] [Reference list]
[5] Vinkel C., Thomsen S.F. Hidradenitis suppurativa: causes, features, and current treatments. J Clin Aesthet Dermatol. 2018;11(10):17–23. [PMC free article] [PubMed] [Google Scholar] [Ref list]
[6] Nazary M., van der Zee H.H., Prens E.P., Folkerts G., Boer J. Pathogenesis and pharmacotherapy of hidradenitis suppurativa. Eur J Pharmacol. 2011;672(1–3):1–8. [PubMed] [Google Scholar]
[7] Chu CB, Yang CC, Tsai SJ. Hidradenitis suppurativa: Disease pathophysiology and sex hormones. Chin J Physiol. 2021 Nov-Dec;64(6):257-265. [PubMed]
[8] Woodruff CM, Charlie AM, Leslie KS. Hidradenitis Suppurativa: A Guide for the Practicing Physician. Mayo Clin Proc. 2015 Dec;90(12):1679-93. [PubMed]
[9] von Laffert M, Stadie V, Wohlrab J, Marsch WC. Hidradenitis suppurativa/acne inversa: bilocated epithelial hyperplasia with very different sequelae. Br J Dermatol. 2011 Feb;164(2):367-71. [PubMed]
[10] van der Zee HH, Laman JD, Boer J, Prens EP. Hidradenitis suppurativa: viewpoint on clinical phenotyping, pathogenesis and novel treatments. Exp Dermatol. 2012 Oct;21(10):735-9. [PubMed]
[11] Fernandez JM, Thompson AM, Borgstrom M, Orenstein LAV, Hsiao JL, Shi VY. Pain management modalities for hidradenitis suppurativa: a patient survey. J Dermatolog Treat. 2022 May;33(3):1742-1745. [PubMed]
[12] Ramirez-GarciaLuna JL, Wang SC, Yangzom T, Piguet V, Kirby JS, Alavi A. Use of thermal imaging and a dedicated wound-imaging smartphone app as an adjunct to staging hidradenitis suppurativa. Br J Dermatol. 2022 Apr;186(4):723-726. [PubMed]
[13] Vossen ARJV, van Straalen KR, Swolfs EFH, van den Bosch JF, Ardon CB, van der Zee HH. Nicotine Dependency and Readiness to Quit Smoking among Patients with Hidradenitis Suppurativa. Dermatology. 2021;237(3):383-385. [PubMed]
[14] Ring HC, Theut Riis P, Zarchi K, Miller IM, Saunte DM, Jemec GB. Prodromal symptoms in hidradenitis suppurativa. Clin Exp Dermatol. 2017 Apr;42(3):261-265. [PubMed]
[15] Antia C, Alavi A, Alikhan A. Topical management and wound care approaches for hidradenitis suppurativa. Semin Cutan Med Surg. 2017 Jun;36(2):58-61. [PubMed] [Reference list]
[16] Hurley H. Axillary hyperhidrosis, apocrine bromhidrosis, hidradenitis suppurativa, and familial benign pemphigus: surgical approach. In: Roenigh R, Roenigh H, eds. Dermatologic surgery. New York: Marcel Dekker; 1989:729-739.
[17] Boer J, Nazary M. Long-term results of acitretin therapy for hidradenitis suppurativa. Is acne inversa also a misnomer? Br J Dermatol. 2011 Jan;164(1):170-5. [PubMed]
[18] 18. Soria A, Canoui-Poitrine F, Wolkenstein P, Poli F, Gabison G, Pouget F, Viallette C, Revuz J. Absence of efficacy of oral isotretinoin in hidradenitis suppurativa: a retrospective study based on patients' outcome assessment. Dermatology. 2009;218(2):134-5. [PubMed]
[19] Kimball AB, Okun MM, Williams DA, Gottlieb AB, Papp KA, Zouboulis CC, Armstrong AW, Kerdel F, Gold MH, Forman SB, Korman NJ, Giamarellos-Bourboulis EJ, Crowley JJ, Lynde C, Reguiai Z, Prens EP, Alwawi E, Mostafa NM, Pinsky B, Sundaram M, Gu Y, Carlson DM, Jemec GB. Two Phase 3 Trials of Adalimumab for Hidradenitis Suppurativa. N Engl J Med. 2016 Aug 04;375(5):422-34. [PubMed]
[20] Gulliver W, Zouboulis CC, Prens E, Jemec GB, Tzellos T. Evidence-based approach to the treatment of hidradenitis suppurativa/acne inversa, based on the European guidelines for hidradenitis suppurativa. Rev EndocrMetabDisord. 2016 Sep;17(3):343-351. [PMC free article] [PubMed] [Reference list]

[21] Tricarico PM, Zupin L, Ottaviani G, Rupel K, Celsi F, Genovese G, Boniotto M, Crovella S, Marzano AV. Photobiomodulation as potential novel third line tool for non-invasive treatment of hidradenitis suppurativa. G Ital Dermatol Venereol. 2020 Feb;155(1):88-98. [PubMed] [Reference list]
[22] Hazen PG, Hazen BP. Hidradenitis suppurativa: successful treatment using carbon dioxide laser excision and marsupialization. Dermatol Surg. 2010 Feb;36(2):208-13. [PubMed] [Reference list]
[23] Tricarico PM, Zupin L, Ottaviani G, Rupel K, Celsi F, Genovese G, Boniotto M, Crovella S, Marzano AV. Photobiomodulation as potential novel third line tool for non-invasive treatment of hidradenitis suppurativa. G Ital Dermatol Venereol. 2020 Feb;155(1):88-98. [PubMed]
[24] Matusiak Ł. Profound consequences of hidradenitis suppurativa: a review. Br J Dermatol. 2020 Dec;183(6):e171-e177. [PubMed] [Reference list]14. Ring HC, Theut Riis P, Zarchi K, Miller IM, Saunte DM, Jemec GB. Prodromal symptoms in hidradenitis suppurativa. Clin Exp Dermatol. 2017 Apr;42(3):261-265. [PubMed] [Reference list]
[25] Yuan JT, Naik HB. Complications of hidradenitis suppurativa. Semin Cutan Med Surg. 2017 Jun;36(2):79-85. [PubMed] [Reference list]
[26] Kinjalk N, Kinjalk M. Music Therapy Emerging Catalyst in the West: Are we ready to prescribe it. International Journal of Scientific Research. Volume-8 | Issue-12 | December - 2019 | DOI : 10.36106/ijsr