Caring for Pediatric Hidradenitis Suppurativa Patients in the Emergency Department

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Abstract: Hidradenitis suppurativa (HS) is a chronic and recurrent inflammatory skin condition resulting in the formation of nodules, sinus tracts, and abscesses in intertriginous areas. We provide recommendations for the management of children presenting to the emergency department with acute HS flares, based on a review of literature and insights from our own clinical experience. The purpose of the recommendations is to educate clinicians on specific considerations that should be made when caring for children with HS.

Kev Words: hidradenitis suppurativa, treatment, skin

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TARGET AUDIENCE

This continuing medical education material is designed primarily for nondermatology clinicians who will encounter pediatric patients presenting to the emergency department with acute flares of hidradenitis suppurativa or symptoms of undiagnosed hidradenitis suppurativa.

LEARNING OBJECTIVES

After completing this article, the reader should be better able to:

- 1. Explain the epidemiology and pathophysiology of pediatric hidradenitis suppurativa and identify associated comorbidities and complications;
- 2. Describe the presentation, examination, and evaluation of pediatric hidradenitis suppurativa in the emergency department;
- 3. Outline treatment recommendations for the acute management of pediatric hidradenitis suppurativa in the emergency room and assess long term treatment options.

idradenitis suppurativa (HS) is a chronic inflammatory skin condition characterized by recurrent, tender, inflamed nodules, or abscesses. Intertriginous areas such as the axillae, inguinal folds, and inframammary regions are most commonly affected. Deeper abscesses, fistulae, sinus tracts, and extensive scarring may occur as the condition progresses. Prevalence ranges from 1 to 4%; however, many cases of HS are misdiagnosed or undiagnosed, so the true prevalence may be higher than reported. Hidradenitis

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suppurativa is more common in African Americans, and females are twice as likely to be affected.2 Disease onset typically occurs after puberty, between the second and third decades of life.² Prepubescent onset of HS is infrequent and more commonly seen in males, as ovarian hormones are key players in postpubescent onset of HS.3

Hidradenitis suppurativa is uncommon in young children, with onset before 11 years of age occurring in only 2% of patients. Several differences should be noted with regard to childhood-onset HS. Hidradenitis suppurativa in children may not present with the classic features seen in adults and can have subtle manifestations on physical examination (Fig. 1). Hidradenitis suppurativa may progress to more severe disease, particularly in children who have an earlier age of disease onset.² Children with HS are also more likely to have associated endocrine or hormonal abnormalities. For example, early onset HS may be a sign of precocious puberty and has been reported in children with congenital adrenal hyperplasia, premature adrenarche, obesity, and polycystic ovarian syndrome.² Whereas only about one third of adults with HS have a family history of HS, about half of children have a positive family history.4

Emergency department (ED) and inpatient services are used more frequently by adult HS patients as compared with patients with other common dermatologic conditions.⁵ Although there have been no studies examining the utilization of inpatient and ED services by pediatric patients, our personal experience has found that ED visits from pediatric HS patients are likely to be just as common. For this reason, we feel that clinicians taking care of children in the ED should be knowledgeable about the condition and know how to acutely manage HS flares.

PATHOPHYSIOLOGY

Although the exact cause of HS remains unknown, evidence suggests that follicular abnormalities, immune dysregulation, and genetics all play a role in its development.⁶ A combination of innate follicular dysfunction, mechanical friction, and inflammation ultimately leads to occlusion of hair follicles in affected areas. Follicular occlusion of the pilosebaceous unit accompanied by bacterial overgrowth and follicular rupture triggers a robust immune response that causes inflammation and development of typical abscesses, sinus tracts, and scarring seen in HS (Fig. 2). Tender nodules can form when the occluded follicle ruptures, releasing sebum and debris to the surrounding dermis. Although additional studies are needed to further elucidate the specific cytokines involved in HS, it is hypothesized that the interleukin (IL)-23/TH17 pathway, particularly IL-17, along with tumor necrosis factor, IL-1β, and IL-10 are important players.⁶

COMORBIDITIES

Hidradenitis suppurativa is associated with a number of general medical conditions including cardiovascular disease, metabolic syndrome, thyroid disease, inflammatory bowel disease, polycystic ovarian syndrome, arthropathies, and other cutaneous conditions such as acne, pyoderma gangrenosum, hirsutism, and pilonidal cysts.

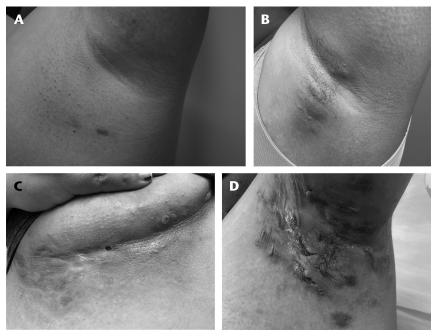


FIGURE 1. Early manifestations: (A) multiple open comedones with subtle atrophic scar in inferolateral axilla; (B) multiple subcutaneous nodules of the axilla. Late manifestations: (C) open draining sinus tract and scar in inframammary fold; (D) multiple connecting sinus tracts and bridging fibrosis of the axilla.

Psychiatric comorbidities are very common and can have a devastating impact on a child's quality of life. Hidradenitis suppurativa patients have increased rates of social isolation, depression, anxiety and suicide. 8 One study found that children with HS are nearly 3 times more likely to have at least 1 psychiatric diagnosis compared with the general population.9

Obesity is another comorbidity seen in children with HS. The prevalence of HS is higher in children with a high body mass

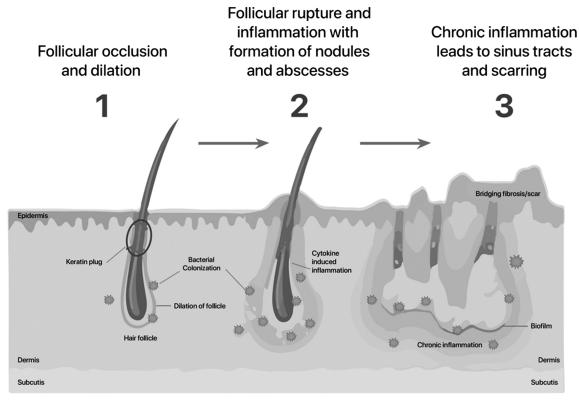


FIGURE 2. The sequence of events underlying HS pathophysiology.

index.⁸ One study demonstrated that children who returned to a healthy weight before 13 years of age could reduce their risk of developing the condition.¹⁰ Overweight children are also more likely to have a recalcitrant form of the disease, but weight loss may improve disease severity.¹¹ This highlights the importance of diet and exercise counseling and the impact healthy eating habits and physical activity can have on reducing disease burden.

Although the association between tobacco exposure in child-hood and the development of HS has not been studied, patients who smoke are more likely to have severe disease. Older children should be counseled on the negative health effects of tobacco and encouraged to abstain from smoking.

DIAGNOSIS

The diagnosis of HS is made clinically. The diagnostic criteria include the following:

- Characteristic lesions (ie, nodules, abscesses, papules, fistulas, and scars);
- Predilection of flexural sites (ie, inguinal folds, axillae, perianal and inframammary regions);
- 3. Lesion recurrence.⁶

Hidradenitis suppurativa in children commonly manifests as double comedones and tender subcutaneous nodules, which may drain malodorous discharge. Nodules may evolve into dermal abscesses, sinus tracts, fibrotic scars, and dermal contractures (Fig. 1). 12 Lesions are most common in the axillae but may present in other intertriginous areas. 13 Lesions are recurrent and can reappear at or near the site of the primary lesion. Clinicians should inquire about previous skin lesions, as this is an important diagnostic clue. It is important to note that the location, lesion type, and extent of disease may vary between patients, so clinicians should maintain a high index of suspicion for HS if the patient presents with recurrent inflammatory nodules in the intertriginous regions to ensure early intervention. The Hurley Staging system allows clinicians to classify HS based on severity: stage 1 (abscesses/nodules, minimal scarring, no sinus tracts), stage 2 (limited sinus tracts and scarring), and stage 3 (widespread sinus tracts and scarring).

Patients with HS have an average diagnostic delay of about 7 years. ¹⁴ The reason for the delay in diagnosis is multifactorial but is partially due to misdiagnosis by clinicians. One study showed that only about half of emergency medicine physicians diagnosed HS correctly and most commonly misdiagnosed the condition as furunculosis. Although distinguishing HS from furunculosis may at times be difficult, there are often diagnostic clues that may help clinicians differentiate between the 2 entities (Table 1). Hidradenitis suppurativa may also be misdiagnosed for other conditions including folliculitis, cellulitis, inflamed epidermal cysts, severe acne, Crohn's disease, infected Bartholin gland, and fungal infections.

COMPLICATIONS

Secondary Infection

Despite the fact that bacteria play a role in its pathogenesis, HS is not an infectious process. Cultures of HS lesions typically grow many bacterial species with no single dominant organism, which suggests that bacteria are secondary colonizers rather than etiologic agents. Although bacterial superinfection is uncommon, a secondary infection should be suspected in patients presenting with systemic symptoms such as fever, circumferential cellulitis, and malodorous and/or purulent discharge. A secondary infection may also be supported by history if a patient reports lesions distinct from their baseline flares. Although leukocytosis may be seen on a complete blood count in the setting of an infection, it should be noted that an elevated white blood cell count, particularly neutrophilia, may be present at baseline due to a state of chronic inflammation.¹⁵

Unless there is suspicion for a superinfection, we do not recommend obtaining a culture from HS lesions. Bacterial overgrowth generally occurs very deep in HS lesions, so a superficial swab of a draining abscess would likely result in growth of normal skin microflora and thus have limited utility. Incision and drainage (I&D) is not recommended to obtain a culture.

Complete Blood Count Abnormalities

As previously discussed, neutrophilia is common in HS patients as are other laboratory abnormalities, such as anemia and leukocytosis. Anemia of chronic disease is the most frequent underlying cause of anemia, and the degree of anemia seems to correlate with HS severity. White blood cell count and neutrophil count have been shown to be higher in Hurley stage 3 disease than Hurley stage 2, whereas lymphocyte count has been shown to be lower in Hurley stage 3 than Hurley stage 2 disease. Is

Contractures, Strictures, and Fistulas

Scarring and fibrosis are complications seen in all patients with severe HS. ¹⁶ In some patients, chronic scarring and fibrosis may lead to contractures, particularly in the axilla, resulting in reduced mobility. For those with severe inflammation, scarring and sinus tract formation involving the genital, perineal, or perianal area may lead to strictures and fistulas involving the urethra, bladder, or rectum.

Malignancy

Hidradenitis suppurativa patients have an increased risk of malignancy. One large retrospective study demonstrated a 50% increased risk of malignancy in individuals with HS. This study also demonstrated a nearly 5-fold increase in cutaneous squamous cell carcinoma. ¹⁷ The risk of squamous cell carcinoma in patients with

TABLE 1. Comparison of HS and	Furunculosis
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	Furunculosis	HS
History	Onset at any age, no familial association	Onset after puberty, 30% with strong family history
Site	Anywhere on skin	Intertriginous
Permanent scarring	Usually none	Yes
Sinus tract formation	None	Yes
Bacterial culture	+ (True infection)	+/- (Not a true infection)
Resolution with antibiotics	Yes	No (lesions may improve)

HS is likely associated with chronic inflammation, scarring, and lymphedema.

Lymphedema

Chronic lymphedema can occur secondary to scarring of lymph glands and obstruction of lymphatic drainage. It is not limited to the extremities and may also be seen in the genital areas.¹⁶

Psychosocial Burden

Hidradenitis suppurativa can have detrimental consequences on quality of life because of the negative effects on physical and psychosocial health. Physical symptoms, such as pain or malodorous discharge at affected sites, may lead to poor self-image and fear of stigmatization, leading to social isolation and depression. One study demonstrated that approximately 20% of children with HS have coexisting psychiatric conditions such as depression and anxiety. Children with HS should be screened for psychiatric disorders, and mental health services should be integrated into patient care if needed. Recommended patient resources include the HS Foundation and Hope for HS.

TREATMENTS

To date, there are no published treatment guidelines for children with HS. The majority of literature consists of case reports and small case series. The use of medical therapies is primarily based on these limited data and extrapolation from adult treatment guidelines. The following discussion of therapies is based on a review of the literature. Not all therapies are recommended for use in the ED setting. Of note, adalimumab is the only treatment approved by the US Food and Drug Administration for HS.

Topical Therapies

Topical treatments for HS include antiseptic cleansers, keratolytic agents, and antibiotics. The most commonly used topical treatment regimen for pediatric HS is clindamycin 1% lotion applied twice daily. 13 Clindamycin/tretinoin or azelaic acid 15% to 20% applied twice daily can be used as an alternative. 12 Benzoyl peroxide washes, which have anti-inflammatory, antibacterial, and comedolytic properties, can be used in conjunction with these treatments.

Systemic Antibiotics

Oral antibiotics are the mainstay of treatment for acute HS flares, and clindamycin is the most commonly used oral antibiotic in both children and adults. In addition, the use of combination therapy with clindamycin and rifampin in adults is supported by the literature. As clindamycin and rifampin are used in the

pediatric population for other indications, it is likely an efficacious treatment option for pediatric HS. Scheinfeld² used clindamycin (300 mg, twice daily) and rifampin (300 mg, twice daily) in 4 cases of pediatric HS with good results. Tetracyclines, specifically doxycycline, may be recommended for mild-to-moderate disease; however, their use should be limited to children 8 years or older because of potential of teeth discoloration and inhibition of bone growth in younger children.⁷ There is also some evidence that azithromycin may be helpful for flares in pediatric HS. Offidani et al¹² treated 8 children with acute flares with a combination of oral azithromycin, with topical clindamycin and oral zinc with favorable results. Zinc supplementation may be used in conjunction with antibiotics because it is an antioxidant with anti-inflammatory properties.

Hormonal Therapies

Finasteride is an antiandrogenic competitive inhibitor of type II 5a-reductase an enzyme found in hair follicles, which decreases the levels of dihydrotestosterone levels. A case series on 3 children with HS demonstrated that finasteride (5 mg) daily had beneficial effects with no major adverse events.² Low doses of spironolactone have also been shown to reduce lesion count and pain in adolescents and adults.7

Immunosuppressors

Although there have been no randomized controlled trials evaluating the effectiveness of biologics in children with HS, tumor necrosis factor α inhibitors are considered first-line treatment in adults with moderate to severe disease. The US Food and Drug Administration has approved the use of adalimumab in patients 12 years or older, with a dosing regimen of 40 mg every other week or 40 mg every week for obese or refractory adolescents. 18 A recent review of MEDLINE and EMBASE databases identified 12 pediatric HS patients treated with biologic treatment with good response. Seven patients experienced complete resolution of their HS, whereas 5 patients had partial resolution. 19 In a case series, 2 pediatric patients demonstrated a reduction in the number of affected areas and improved quality of life after treatment with adalimumab. 18 A case report described a 16-year-old adolescent girl who showed strong clinical and psychological improvement with adalimumab use for 4 years, with no adverse effects.¹⁸ Literature on infliximab or etanercept use in pediatric HS is scarce, but in a case series, 1 pediatric patient demonstrated subjective improvement with etanercept. 18 Although oral steroids and methotrexate are used in the adult literature, these medications are not typically used in pediatric HS unless the presentation is severe.

TABLE 2. Approach to the Emergency Department Management of HS in Children

Decommendations for Acute Treatment of HS in Padiatric Patients Who Present to the FD

Recommendations for Acute Treatment of II's in Fediatric Fatients who Fresent to the ED		
Topicals	Benzoyl peroxide wash $1-2\times$ daily Clindamycin 1% bid	
Oral antibiotics <8 years old	1st line: azithromycin $(5-10 \text{ mg/kg/d} \text{ once daily} \times 10-14 \text{ d } [\text{max } 500 \text{ mg/24 h}])$ or erythromycin $(30-50 \text{ mg/kg/d} \text{ divided } \text{tid} \times 10-14 \text{ d } [\text{max } 4 \text{ g/24 h}])$ 2nd line: cephalexin $(25-50 \text{ mg/kg/d} \text{ divided } \text{bid} \times 10-14 \text{ d } [\text{max } 4 \text{ g/24 h}])$	
Oral antibiotics ≥8 years old	1st line: doxycycline (2.2 mg/kg/dose bid \times 10–14 d [max 200 mg/24 h])* 2nd line: minocycline (1–2 mg/kg/dose bid \times 10–14 days [max 200 mg/24 h])	
Follow-up	2–4 wk with dermatology	

^{*}May also use first-line options for oral antibiotics <8 years old if patient cannot swallow pills. bid indicates twice daily; max, maximum; tid, 3 times daily.

Procedures

Although I&D is performed in nearly one third of adult HS ED visits, I&D should not be routinely performed.²⁰ Incision and drainage should only be considered for symptomatic relief from debilitating pain because it does not improve disease outcomes and can potentially result in scarring. Patients may experience pain from an open wound after I&D, which should be weighed against the average duration of a painful boil in HS, which is typically about 7 days.²¹ There are reports on the treatment of pediatric HS with botulinum toxin A and fractionated CO2 laser.² Intralesional triamcinolone acetonide (5-10 mg/mL) has also been used for acute flares in adults, especially resistant nodules and sinus tracts. Although no reports of this therapy are documented in children, it is a potential treatment option. ²² Anecdotally, laser hair removal has been used to address unwanted hair growth in areas affected with HS. Deroofing surgery, which involves using tissue sparing measures to transform tender lesions into cosmetically acceptable scars, has also been performed in children.

Wound Care

Maintaining good hygiene is important to prevent secondary infection and to suppress potential triggers for an immune response. Antiseptics, such as chlorhexidine, benzoyl peroxide, and zinc pyrithione, can be used to prevent colonization of HS lesions by pathogens. Adequate dressing for lesions depends on the location, extent, and morphology of the lesions. Additional factors to take into consideration include the characteristics of the drainage, cost and availability of product, and the need for antiseptics. Hydrocolloid dressings may be used on HS lesions with mild to moderate amounts of exudate. Lesions with large amounts of drainage should be dressed with extra absorbent dressings such as foams and alginates in addition to a secondary dressing. Silicone or petroleum jelly and a nonadherent dressing are recommended for lesions that have been traumatized to reduce friction. It is important to ensure that dressings fit the folds of the body to avoid additional trauma to the area. Lesions that have been excised should be dressed similarly.

Clothing

Strategic clothing selection can have dramatic effects on quality of life for patients. Clothing that decreases mechanical friction, humidity, heat, and microbial colonization is ideal. Clinicians should discuss clothing recommendations as part of initial counseling. Clothing made from cellulose-derived rayon fibers is recommended because it is moisture-wicking and has a high degree of intrinsic whiteness, which may be easier to clean. Clothing derived from bamboo fibers may also be recommended because it is softer than clothing made of cotton fibers, resulting in less mechanical friction. Bras without tight underwires or elastic bands, such as sports bras, camisole tanks, bra liners, and breast pads, can reduce mechanical stress and absorb any drainage from HS lesions in the inframammary and mammary regions.²³ Seamless "boy shorts," briefs, high-cut briefs, and/or "cheeky" underwear are recommended for women, whereas loose-fitting boxers or trunks are recommended for men. An absorbent pad or mesh underwear can be worn if there are draining lesions in the perineal, perianal, or inguinal regions. An abdominal liner may be beneficial for obese patients with a large pannus to absorb discharge and reduce mechanical friction.

Pain Control

Acute and chronic pain control in HS patients is an important consideration because pain can be severely debilitating and, in many instances, cause more distress than the presence of lesions alone. Pain is most common with deep inflammatory nodules. Topical analgesics, nonsteroidal anti-inflammatory drugs, and oral or intralesional steroids can be used to alleviate discomfort and inflammation. Opioids are rarely considered because there are many other safe and efficacious options.²⁴ In some instances, it may be beneficial to consult a pain management specialist.

RECOMMENDATIONS FOR ACUTE MANAGEMENT OF PEDIATRIC PATIENTS WITH HS IN THE ED

A patient who presents to the ED with a known diagnosis of HS should be continued on their prescribed therapy. If a patient is not on a topical treatment regimen at the time of presentation, we recommend starting topical clindamycin 1% with a benzoyl peroxide wash twice daily to affected areas. This is to be continued as long-term topical maintenance therapy.

We recommend specific oral antibiotics for patients presenting to the ED with an acute HS flare based on patient age. Antibiotics are given to help with the acute inflammation. For children younger than 8 years, we recommend azithromycin or erythromycin for 10 to 14 days. If both azithromycin and erythromycin are not available or have already been trialed, we recommend cephalexin for 10 to 14 days. For children 8 years or older, we recommend doxycycline 10 to 14 days. If a child has adverse effects on doxycycline, minocycline can be used as an alternative. Although clindamycin is the most commonly used antibiotic in the pediatric literature, we are not recommending it because of observed poor compliance given its poor palatability and high-volume dosing required. All patients should follow up with a dermatologist within 2 to 4 weeks of presentation to the ED. Table 2 summarizes our recommendation for the treatment of pediatric patients with HS in the ED setting.

CONCLUSIONS

Pediatric patients may present to the ED with acute flares of HS or symptoms of undiagnosed HS. Clinicians evaluating these patients should be able to recognize HS, understand the etiology of this condition, and be familiar with the recommendations for the acute management of HS.

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