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Letter to the Editor

Demographics and treatment trends in the largest cohort of patients with Down syndrome and hidradenitis suppurativa

Dear Editor,

Individuals with Down syndrome (DS) are at increased risk for follicular occlusion disorders, including hidradenitis suppurativa (HS).[1] The prevalence of HS within DS varies with estimates from 2.1% to 3.02%.[1,2] Herein, we present the largest cohort of patients with HS and DS with a focus on demographics and treatment.

A retrospective study was performed at the University of Virginia and Virginia Commonwealth University hospital systems to identify patients with HS and DS. Treatments were identified, and patients were confirmed to take medications through chart review. Patients were excluded if no treatments were reported.

Forty-three patients with HS and DS were identified; eight patients were not on medical therapy and were thus excluded. Of the eligible patients, 22 (51.2%) were male (Table 1). The median age of HS onset was 22.5 years (IQR 18–28), and the average body mass index (BMI) was 35.53 kg/m^2. Forty patients (93.0%) were nonsmokers, and the remaining three had no smoking status reported. Family history was inconsistently documented. Hurley stage I was most common (21 of 40, 52.5%), followed by stage II in 37.5% and stage III in 10.0%. The most common locations for HS were the groin (N = 29, 67.4%) and axillae (N = 28, 65.1%), followed by the buttocks, legs, and genital area. The most commonly used treatments were topical antibiotics (N = 37, 86.0%) and oral antibiotics (N = 33, 76.7%). Seven of 19 (36.8%) patients with moderate and severe HS were recommended to start adalimumab. Three patients successfully initiated the medication, whereas four patients declined due to caregiver concern about malignancy risk or patient inability to tolerate regular injections. Four patients underwent surgery, with two patients classified as Hurley stage III; one of these patients was also on adalimumab. Surgeries were caregiver-reported, and information on the type of procedure could not be obtained. The majority of patients tolerated medications well without adverse effects. Gastrointestinal upset was reported in four (12%) patients taking doxycycline, resulting in discontinuation.

We present the most extensive series of patients with DS and HS, emphasizing HS treatment. Our cohort was diverse, with a higher proportion of males than is typically seen in the general population.[3] The majority of patients in this cohort had early-stage HS, and typically, less aggressive treatment is needed for milder disease. However, a few patients, regardless of Hurley stage, were treated with biological therapy or surgery, which is consistent with the literature.[1,4] Notably, the median age of onset and average BMI in this cohort were higher compared to prior studies in the general HS population, reporting a median age of onset of 16 and 19 for women and men, respectively, and average BMI estimates ranging from 25.45 to 31.00.[1–4]

Given that there are FDA-approved treatments for moderate-to-severe HS, DS patients may be undertreated, especially those with Hurley stage II or III, representing potential care gaps in this population. DS-specific factors may play a role, including patient/caregiver hesitancy over using perceived "immunosuppressants" given the increased risk of leukemia in DS and concerns about patient tolerability of injections, infusions, or surgical procedures. Reassuringly, tumor necrosis factor-alpha inhibitors have not been shown to increase the risk of leukemia, and many patients with DS

Table 1 Summary of demographics and treatment for cases of hidradenitis suppurativa in Down syndrome patients

Sample size	43
Median age of onset (range)	22.5 (12–45)
Mean body mass index (kg/m^2)	35.53
Race/ethnicity, N =43 (%)	
White	19 (44.2)
African American or Black	20 (46.5)
Hispanic/Latino	4 (9.3)
Gender, N = 43 (%)	
Male	22 (51.2)
Female	21 (48.8)
Hurley stage, N = 40 (%)[a]	
I	21 (52.5)
II	15 (37.5)
III	4 (10.0)
Treatment, N = 43 (%)	
Topical antibiotics	37 (86.0)
Oral antibiotics	33 (76.7)
Doxycycline	22 (66.7)
Rifampin	4 (12.1)
Minocycline	4 (12.1)
Bactrim	2 (6.1)
Clindamycin	1 (3.0)
Biologics	3 (7.0)
Hormonal therapy	5 (11.6)
Spironolactone	4 (9.3)
Finasteride	1 (2.3)
Surgery	4 (9.3)

[a]This category has a different sample size, as some cases did not report the specified information.

treated at our institutions can tolerate subcutaneous injections at home.[5] Before starting any medication, practical treatment regimens and ensuring a DS patient's or caregiver's understanding of their treatment plan is essential.

The lack of consistent disease severity reporting and the retrospective nature of this study are limitations. In summary, this cohort demonstrates that patients with HS and DS may be undertreated, possibly because of misconceptions regarding the tolerability and safety of biologics and surgery. Notably, the safety and efficacy of biologics in the DS population require further study.

Data availability statement

The data that support the findings of this study are available upon request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

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References

1 Giovanardi G, Chiricozzi A, Bianchi L, de Simone C, Dini V, Franceschini C, et al. Hidradenitis suppurativa associated with down syndrome is characterized by early age at diagnosis. *Dermatology*. 2018;**234**(1–2):66–70.
2 Lam M, Lai C, Almuhanna N, Alhusayen R. Hidradenitis suppurativa and Down syndrome: a systematic review and meta-analysis. *Pediatr Dermatol*. 2020;**37**(6):1044–50. https://doi.org/10.1111/pde.14326
3 Naik HB, Paul M, Cohen SR, Alavi A, Suàrez-Fariñas M, Lowes MA. Distribution of self-reported hidradenitis suppurativa age at onset. *JAMA Dermatol*. 2019;**155**(8):971–3.
4 Hernández-Rodríguez JC, Osorio-Gómez GF, Ortiz-Álvarez J, Lebrón-Martín JA, Conejo-Mir J, Pereyra-Rodríguez JJ. Hidradenitis suppurativa and Down syndrome in a single-centre sample: a cross-sectional study. *Australas J Dermatol*. 2022;**63**(3):e231–7.
5 Haynes K, Beukelman T, Curtis JR, Newcomb C, Herrinton LJ, Graham DJ, et al. Tumor necrosis factor α inhibitor therapy and cancer risk in chronic immune-mediated diseases. *Arthritis Rheum*. 2013;**65**(1):48–58.