



Clinical characteristics of pediatric hidradenitis suppurativa: a cross-sectional multicenter study of 140 patients

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Abstract

Hidradenitis suppurativa (HS) rarely affects pediatric patients. The literature on pediatric HS patients is scarce. This is a cross-sectional study based on case note review or interviews and clinical examination of 140 pediatric patients undergoing secondary or tertiary level care. Patients were predominantly female (75.5%, $n = 105$) with a median age of 16. 39% reported 1st-degree relative with HS. Median BMI percentile was 88, and 11% were smokers ($n = 15$). Median modified Sartorius score was 8.5. Notable comorbidities found were acne (32.8%, $n = 45$), hirsutism (19.3%, $n = 27$), and pilonidal cysts (16.4%, $n = 23$). Resorcinol ($n = 27$) and clindamycin ($n = 25$) were the most frequently used topical treatments. Patients were treated with tetracycline ($n = 32$), or oral clindamycin and rifampicin in combination ($n = 29$). Surgical excision was performed in 18 patients, deroofting in five and incision in seven patients. Obesity seemed to be prominent in the pediatric population and correlated to parent BMI, suggesting a potential for preventive measures for the family. Disease management appeared to be similar to that of adult HS, bearing in mind that the younger the patient, the milder the disease in majority of cases.

Keywords Hidradenitis suppurativa · Children · Pediatric · Acne inversa

Introduction

Hidradenitis suppurativa (HS) is a chronic inflammatory skin disease. Patients recurrently develop painful nodules in the intertriginous regions. The nodules progress to abscesses that heal with scars, potentially forming cutaneous tunnels

predisposing to further inflammation [22, 43]. The disease has a negative impact on the life-course of patients which suggests that early identification and characterization of patients is important [7, 8, 11, 20, 52].

The prevalence of HS is 1–2% [23], of these approximately 2.2% have been estimated to be pediatric patients [13], making pediatric HS a rare condition, prevalence studies specifically targeting children report prevalences of 0.028% for American children, with a prevalence of 0.114% for children between 15 and 17 [17]. Incidence studies from the USA showed that 2.4% of new HS diagnosis over a 10-year period was for children aged 0–17 [14]. One European study indicates that 7.7% of HS patients' reports pre-pubertal onset [9], another study found that 19 of 235 HS patients (8.1%) reported onset before the age of 16 [3]. The literature on pediatric HS is based on cases, case series, and reviews of these. Common themes are either (1)

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an interesting comorbidity like HIV or Down's syndrome [16, 41], (2) premature adrenarche [31] or (3) a successful and unexpected treatment like botulinum toxin or finasteride [12, 42]. It may be speculated that the field of pediatric HS is influenced by publication bias, providing undue weight to the few cases published [34].

Several risk factors are suspected to influence the development of HS: smoking, obesity, and a genetic predisposition [22]. Smoking is associated with both the development and the severity of HS [15, 26, 46] and 41–92% of adult HS patients are reported to be smokers [5, 19, 26, 28, 35, 36, 45, 48, 55, 56]. Obesity affects self-evaluated health, severity of HS [25, 29, 45, 51], and reduces the chance of disease remission [28]. Genetic predisposition is suspected as 30–42% of HS patients report relatives with similar symptoms [9, 40].

This study aims to provide a clinical description of a large cohort of pediatric HS patients with a focus on common risk factors, clinical presentation, and treatments.

Materials and methods

This cross-sectional, explorative, and descriptive study is based on case note reviews ($n=20$), interviews ($n=120$), and clinical examination of pediatric patients (All, $n=140$) undergoing secondary or tertiary level care in the Netherlands, Poland, Sweden, Belgium, Spain, France, Italy, Switzerland, Denmark, Slovenia, Qatar, Egypt, Saudi Arabia, Turkey, Tunisia, and South Africa. Patients were included from 8th of April 2017 to 6th of February 2018.

Although it is debatable, patients < 18 years old were considered pediatric patients throughout the manuscript. Pediatric patients were screened consecutively for inclusion, and known pediatric HS patients were invited. All patients below 18 years of age, with diagnosed HS, were eligible to participate. Signed consent from the legal guardian was obtained, for all patients, before inclusion. The patients, their legal guardian, and the dermatologist completed the survey in concert. In total 57 dermatological centers were invited to participate in the study and 22 centers entered the study. The remaining centers declined either due to lack of time, lack of pediatric patients or failed to answer the invitation. In total 140 patients were recruited. The survey explored race, sex, age, age at disease onset, weight, height, smoking exposure, comorbidities including acne severity as suggested by Dréno et al. [10] and pain scores for the last week (Numeric rating scale 0–10). Patients with first-degree relatives with HS were considered genetically predisposed. The relatives were not examined, but the patient's assessment of their relatives' disease status was accepted, due to the unique nature of HS lesions. Body mass index (BMI) was assessed with sex, age, and country-specific growth curves and reported as the BMI percentile for the local population. BMI of the parents was

noted and reported as an average. Patients were examined clinically to determine modified Sartorius score [46] and to explore locations of active lesions (abscesses, inflamed nodules and draining fistula), other severity assessment systems were not used.

No severity assessment in HS is validated in the pediatric population. Sartorius score was chosen as severity measurement over Hurley stage as it is supposed to be superior to capture variation in mild cases of HS that are predominantly inflammatory, as was suspected to be the case in most pediatric patients.

Treatments used

The dermatologists were asked to provide information on treatments used and how the patient responded to treatment. The physician could report either Response, No response or Unknown response. This was a crude measurement of effect and not an actual measurement of efficacy.

Statistics

Categorical data are presented with frequency (percentage) and compared with a chi-squared test or Fisher exact test as appropriate. Continuous data are presented with medians (interquartile range–IQR) due to non-Gaussian distributions for all variables; comparisons were made with Mann–Whitney U tests. Spearman correlations were used. Binominal logistic regressions were used to assess possible predictors for having active lesions in the axilla, around the breast, on the buttocks, in the groin, in the pubic area, in the genital region, in the anal region or elsewhere. SPSS version 24.0 (IBM, New York, USA) was used for all statistics.

Results

Demographics

In total, 140 pediatric patients were included. Patients were predominantly female (75.5%, $n=105$). Smokers in the household were reported by 44.6% ($n=50$), while 11.1% ($n=15$) of the patients were smokers themselves. First degree relatives with HS were reported by 39.3% ($n=55$). Patients had a median age of 16 (IQR=3) and a median BMI percentile of 88 (IQR=23). (Table 1). Tanner development stages were skewed towards higher stage, with 48.9% of females and 71.0% of males being in stage V. (Table 1). Age and BMI percentile distributions were skewed towards higher age and higher BMI percentile. (Figs. 1 and 2).

Table 1 Demographics

Demographics	<i>n</i> = 140
Women, <i>n</i> (%)	105 (75.5)
Men, <i>n</i> (%)	35 (24.5)
Age in years, median years (IQR)	16 (3)
Age at disease onset, median years (IQR)	12.5 (4)
First degree relatives with HS, <i>n</i> (%)	55 (39.3)
BMI percentile, median BMI (kg/m ²) (IQR)	88 (23)
Average parent BMI (kg/m ²) (IQR)	25.9 (6.4)
Smokers, <i>n</i> (%)	15 (11.1)
Smoker in household, <i>n</i> (%)	50 (44.6)
Predominant race	
Caucasian, <i>n</i> (%)	88 (63)
Middle eastern, <i>n</i> (%)	28 (20)
Black, <i>n</i> (%)	15 (11)
Asian, <i>n</i> (%)	2 (1)
Other, <i>n</i> (%)	4 (3)
Tanner stage females (breasts)	
I	9 (10.2%)
II	2 (2.3%)
III	7 (8.0%)
IV	27 (30.7%)
V	43 (48.9%)
Tanner stage males (genitals)	
I	2 (6.5%)
II	0 (0%)
III	3 (9.7%)
IV	4 (12.9%)
V	22 (71.0%)

n number, *IQR* interquartile range, *BMI* body mass index, *HS* hidradenitis suppurativa

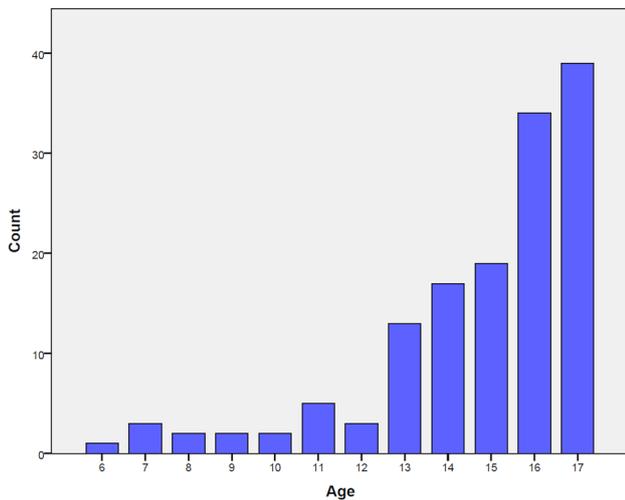


Fig. 1 Histogram of the age distribution in the cohort

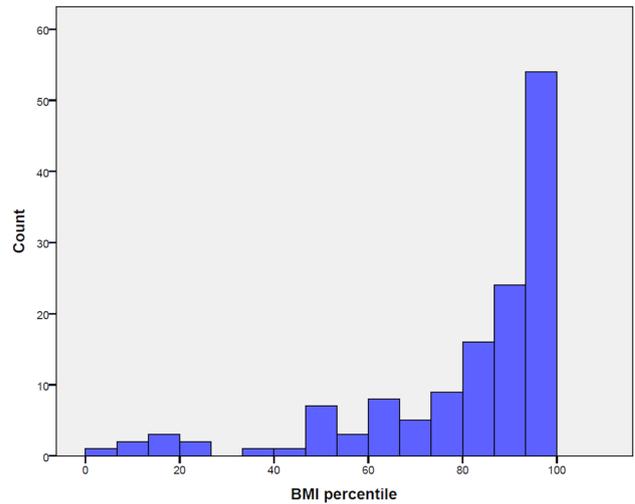


Fig. 2 Histogram of the body mass index percentile distribution in the cohort. *BMI* body mass index

Location of active lesions

Females had less axillary involvement than males (37.1% vs. 64.7%, *P* = 0.005), female patients with first-degree relatives with HS had less axillary involvement than female patients without first degree relatives (25% vs. 45.9%, *P* = 0.029) (Table 2.). Smoking, the presence of a smoker in the household, and BMI did not affect the location of active lesions. However, binomial regression showed that the odds ratio (OR) of having active axillary lesions were 1.30 (95% confidence interval (CI): 1.09–1.55, *P* = 0.003) per year increase in age. Age, BMI percentile, smoking or the presence of smoking in the household were not associated with the presence of active lesions in other locations. Typical lesions are shown in Figs. 3 and 4.

Severity

Patients had a median Sartorius score of 8.5 (IQR = 14) and reported a median pain score of 5 (IQR = 6) (Table 2). Patients with first-degree relatives with HS had statistically lower Sartorius scores than those who were not (median: 7 vs. 12, *P* = 0.006) and a non-significant lower pain score (median: 3 vs. 5, *P* = 0.236). There were no statistical differences in Sartorius score or pain score between male and female patients (median: 8.25 vs. 9, *P* = 0.643 and median: 6 vs. 5, *P* = 0.458, respectively). While BMI percentile was positively correlated to Sartorius score (Spearman’s rho: 0.242, *P* = 0.005), it was not significantly correlated to pain score (Spearman’s rho: 0.144, *P* = 0.204). Age correlated neither to Sartorius nor to pain scores (Spearman’s rho: -0.018, *P* = 0.844 and

Table 2 Location of active lesions

Location and severity	Total (<i>n</i> = 140)	Women (<i>n</i> = 105)	Men (<i>n</i> = 35)
Location			
Axillary region, <i>n</i> (%)	62 (44.3)	39 (37.1)	34 (64.7)
Breast region, <i>n</i> (%)	26 (18.6)	22 (21.0)	4 (11.8)
Buttocks region, <i>n</i> (%)	28 (20.0)	19 (18.1)	9 (26.5)
Groin region, <i>n</i> (%)	60 (42.6)	44 (41.9)	15 (44.1)
Pubic region, <i>n</i> (%)	27 (19.3)	21 (20.0)	5 (14.7)
Genital region, <i>n</i> (%)	10 (7.1)	6 (5.7)	3 (8.8)
Anal region, <i>n</i> (%)	12 (8.6)	8 (7.6)	4 (11.8)
Elsewhere, <i>n</i> (%)	15 (10.7)	9 (8.6)	5 (14.7)
Severity			
Median Sartorius Score, (IQR)	8.5 (14.0)	9 (13.0)	8.25 (31.5)
Median pain VAS Score last week, (IQR)	5 (6)	5 (7)	6 (8)

n number, *IQR* interquartile range, *NRS* (Numeric Rating Scale 0: no pain 10: maximal pain)



Fig. 3 A 17-year old man with solitary nodules and heavy scarring in the groin area

0.068, $P = 0.449$, respectively). Average parent BMI was correlated to their child's BMI percentile (Spearman's rho: 0.445, $P < 0.001$).

Comorbidities

The most common comorbidity found was acne, mild severity or higher was reported in 32.8% ($n = 45$), followed by hirsutism (19.3%, $n = 27$) predominantly in females (23/105 21.9%), and pilonidal cysts (16.4%, $n = 23$) predominantly in males (13/35, 37.1%).

Down's syndrome was observed in 7 patients (5%), precocious puberty in 5 patients (3.6%), and polycystic ovarian syndrome in 4 patients (2.9%). (Table 3). For complete information on all reported comorbidities see Supplementary Table 1.



Fig. 4 A 16-year old woman with solitary lesions and a surgical scar on her right upper thigh

Treatment response

Topical treatments

Resorcinol ($n = 27$) and clindamycin ($n = 25$) were the most frequently used topical treatments. A total of 36% (9/25) responded to topical clindamycin and 63% (17/27) to resorcinol (Table 4).

Table 3 Most common comorbidities

Comorbidities and acne severity	Total, <i>n</i> = 140	Women, <i>n</i> = 105	Men, <i>n</i> = 35
Hirsutism, <i>n</i> (%)	23 (16.4)	23 (21.9)	–
Hypertrichosis, <i>n</i> (%)	4 (2.9)	–	4 (11.8)
Pilonidal cysts, <i>n</i> (%)	23 (16.4)	10 (9.5)	13 (38.2)
Down's syndrome, <i>n</i> (%)	7 (5.0)	4 (3.8)	3 (8.8)
Precocious puberty, <i>n</i> (%)	5 (3.6)	3 (2.9)	2 (5.9)
Polycystic ovarian syndrome, <i>n</i> (%)	4 (2.9)	4 (3.8)	–
Acne severity			
Clear, <i>n</i> (%)	75 (53.6)	57 (54.3)	17 (50.0)
Almost clear, <i>n</i> (%)	17 (12.1)	12 (11.5)	5 (14.7)
Mild, <i>n</i> (%)	22 (15.7)	17 (16.3)	5 (14.7)
Moderate, <i>n</i> (%)	20 (14.3)	15 (14.3)	5 (14.7)
Severe, <i>n</i> (%)	3 (2.1)	1 (1.0)	2 (5.9)
Very severe, <i>n</i> (%)	1 (0.7)	1 (1.0)	0 (0)

Patients could have more than one comorbidity
n number

Table 4 Most common treatments and responses

Treatment	Response, <i>n</i> (%)	No response, <i>n</i> (%)	Unknown, <i>n</i> (%)
Topical treatments			
Clindamycin, (<i>n</i> = 25)	9 (36)	11 (44)	5 (20)
Resorcinol, (<i>n</i> = 27)	17 (63)	3 (11.1)	7 (25.9)
Oral antibiotics			
Tetracycline, (<i>n</i> = 32)	18 (56.3)	13 (40.6)	1 (3.1)
Doxycycline, (<i>n</i> = 15)	8 (53.3)	5 (33.3)	2 (13.3)
Lymecycline, (<i>n</i> = 4)	2 (50)	2 (50)	0 (0)
Minocycline, (<i>n</i> = 4)	2 (50)	2 (50)	0 (0)
Clindamycin and rifampicin, (<i>n</i> = 29)	19 (65.5)	8 (27.6)	2 (6.9)
Amoxicillin and clavulanic acid, (<i>n</i> = 13)	7 (53.8)	4 (30.8)	2 (15.4)
Pristinamycine, (<i>n</i> = 8)	3 (37.5)	4 (50)	1 (12.5)
Ceftriaxone and metronidazole, (<i>n</i> = 6)	4 (66.7)	2 (33.3)	0 (0)
Biologic treatments			
Adalimumab, (<i>n</i> = 4)	2 (50)	0 (0)	2 (50)
Infliximab, (<i>n</i> = 1)	0 (0)	1 (100)	0 (0)
Surgical treatments			
Excision, (<i>n</i> = 18)	14 (77.8)	0 (0)	4 (22.2)
Incision, (<i>n</i> = 7)	0 (0)	5 (71.4)	2 (28.6)
Deroofing, (<i>n</i> = 5)	5 (100)	0 (0)	0 (0)

n number

Oral medication

Patients responded in 18 of 32 cases (56.3%) to treatment with tetracycline, with a similar success rate for doxycycline, lymecycline, and minocycline (Table 4).

Oral clindamycin and rifampicin in combination had 19/29 (65.5%) responders, while the combination of amoxicillin and clavulanic acid had 7/13 (53.8%) responders (Table 4).

Biologic treatments were uncommon. Adalimumab had 2/4 (50%) responders. Infliximab had failed in a single patient (Table 4).

Surgical treatments

A surgical approach was taken in 30 patients out of 140 (21.4%). In details, the response rates were 14/18 (77.8%) for surgical excision, 5/5 (100%) for deroofing, and 0/7 (0%)

for incision and drainage (Table 4). For the full list of treatments employed and the response rates see Supplementary Table 2.

Sex, smoking and the reported first-degree relatives with HS did not influence response rates. However, patients with a BMI percentile over the median responded less to topical clindamycin than those with a BMI percentile below the median (22.2% vs. 50.6% responders, $P=0.008$).

Discussion

HS is a severe inflammatory skin disease with a profound effect on the patients, not only physically and psychologically, but also socially. Therefore, data on the rare pediatric cases are essential. This study describes suspected HS risk factors, clinical characteristics, and treatment responses in a cohort of 140 pediatric cases recruited from a geographically wide area.

It has been hypothesized that a strong genetic predisposition is linked to early onset of HS [9]. Here, 39% of the pediatric cases had a 1st degree relative with HS, which is in good accordance with adult cases [9]. Only 11.1% of cases were smokers (age range: 14–17 years). The low number of cases per center makes a comparison to the general population difficult, due to significant regional differences in smoking rates. In the adult HS populations, smoking is reported by 41–92% [5, 19, 26, 28, 36, 45, 48, 55, 56]. The lower number in the pediatric population most likely reflects young age, legal age-restrictions to buying cigarettes, and limited financial means. However, 44.6% of patients in our cohort were subjected to passive smoking in the household, which could have influenced their development of HS. The questionnaire did not explore if the household smokers were first-degree relatives; hence, possible synergies between the two risk factors, could not be explored in this study.

The pediatric patients were overweight with a median BMI percentile of 88. BMI percentile has not been explored for adult HS patients, but several studies have reported that HS patients as a group have a high BMI [22, 29, 43]. Interestingly, patient BMI percentiles were positively correlated with the average parent BMI. This implies that obese patients' home environment and family unit should be the target for nutritional intervention and not just the patient [2].

A previous report on 33 American pediatric patients reported higher rates of involvement of all regions [4]; they found that 84.8% of patients had axillary involvement compared to 44.3% in our study, they found 57.6% had lesions in the groin compared with 42.6% in this study. We hypothesize that difference could be partly accounted for by the ethnic composition of the two grounds. The cohort reported on by Braunberger et al. [4] consist of 57.1% African American and 25.0% Caucasian, whereas the present cohort consists

of 11% Black children and 63% Caucasian. Prevalence and incidence studies both show a propensity for HS [14, 17] in African American children, and this might translate to more wide-spread lesions as well.

A French study of 302 adult patients found 69.4% having axillary lesions [5]—we found 44.3%. Lesions under/around the female breast were found in 22.5% by Canoui-Poitrine et al. [5] and 21.0% in this study, while the corresponding figures for the buttocks were 27.2% vs. 20.0%. The only noticeable difference was in the rates of axillary lesions, but that could be explained by young age. We found that the OR for the presence of axillary lesions increased with 1.303 (95% CI: 1.094–1.551) per year.

Patients under 18 years that develop lesions have likely not yet reached their lifetime peak HS severity. This might explain why we found a lower median Sartorius severity score of 8.5, while comparable adult studies report a mean Sartorius score of 17 [5].

Pediatric comorbidities

Case reports have previously found atopic dermatitis [49], premature adrenarche [31], dissecting cellulitis of the scalp [27], acanthosis nigricans [33], and Down's syndrome with hypothyroidism [33] in one case each of pediatric HS. In a cohort of 380 children with Crohn's disease, seven of them also displayed signs of HS [38]. Interestingly, none of the previous cases reported hirsutism or pilonidal cysts, which we found in 19.3% and 16.4%, respectively. Hirsutism occurs in 5–10% of adult women [1] but has been associated with obesity, which could explain the high prevalence of our sample [54]. Pilonidal cysts have been reported in 30.2% of adult HS patients [5], but only in 1.3% in a Finnish registry study of a pediatric HS population [53], however, pilonidal cysts were not the target in this study making an underestimation likely. The prevalence of pilonidal cysts in the general population has been estimated at 0.7% [6], with increased incidence in obese patients [30].

Acne was reported by 32.8% (mild or higher) and was predominantly mild, but the prevalence was expectedly lower than reported in studies targeting acne prevalence [18]. The low number of severe cases precluded association analysis with regards to HS severity.

Down's syndrome was found in 5% of cases in our study and 2.1% in a previous study [16] suggesting an association between the two diseases. Given the causative extra chromosome in Down's syndrome, the high rate of Down's syndrome in our study likely reflects a propensity for HS in patients with Down's syndrome rather than a two-way association.

Precocious puberty [39] and polycystic ovarian syndrome [42] have also been reported in two and one cases each, and have led to several reviewers recommending endocrine

screening of pediatric HS patients [32, 34, 47]. Five cases (3.6%) of precocious puberty, which is usually seen in 0.2% of girls and less than 0.05% of boys [50] and four cases (2.9%) of polycystic ovarian syndrome, along with the high rates of hirsutism supports that the endocrine screening of pediatric patients should be done.

We found two cases of anxiety in 140 patients (Supplementary Table 2) corresponding to 1.4%, while Tiri et al. found that 9.2% of 153 pediatric HS cases had anxiety. Tiri et al. found that in total 15.7% had psychiatric disorders [53], while we only found the two cases of anxiety. The current study did not focus on psychiatric disorders in particular but on comorbidities in general, which could explain the low number. This could be explained by a higher rate of psychiatric disorders among adolescents in Finland [44].

Treatment options for pediatric HS

Efficacy trials for the treatment of HS are rare in the adult population and, to the best of our knowledge, non-existent for the pediatric population. Clinicians rely on extrapolation from treatment guidelines for adults, cases, case series, and personal experience [34].

This cross-sectional study can explore real-world treatment and its reported effects as a starting point for future studies. This study employed a very crude measurement for effect (response/no response/unknown). The treatment questions were included to explore real-life practice. The data are not suitable for any efficacy measurements or meta-analysis, especially as disease severity at the time of treatment initiation and the reasoning for initiating treatment is heavily influenced by recall bias and therefore omitted in this study.

The European S1 hidradenitis suppurativa guidelines recommend topical clindamycin as the first treatment option to be explored, followed by oral tetracycline and, possibly, the combination of oral clindamycin and rifampicin [57]. These and topical resorcinol, were generally the most used treatment options for pediatric patients. Sizable differences in response to monotherapy were found, with e.g. 36% responders to topical clindamycin and 63% responders to resorcinol. Concluding anything on the response rates presented here should be done with great care, due to the lack of information on what prompted the treatment, and the crudeness of the response/non-response effect measurement.

Pediatric HS patients also undergo surgery. Surgical excision and deroofting were reportedly useful with no non-responders; whereas simple incision was universally ineffective with no responders in seven cases [21]. This is in agreement with the experiences in adult HS.

Cases of successful finasteride treatment of pediatric HS have been described [24, 37, 42]. Finasteride was only tried in three patients in our cohort, with two non-responders and

one with unknown response (Supplementary Table 2), suggesting limited efficacy.

Strengths and limitations

The large sample size is the apparent strength of this study. In addition, the survey was answered with the help of the dermatologist. However, the study has several limitations; some data, like the age of disease onset, are obviously subject to recall bias. To increase the sample size, known pediatric patients were invited to the clinic for examination and to complete the survey, meaning that the study used both consecutive- and convenience-sampling, which was done to increase the dataset on this rare condition. Due to the skewed age distribution (Fig. 1) a comparison of the children (≤ 12 year old, 12.9% $n = 18$) to the adolescents (> 12 year old, 87.1% $n = 122$) has not been the focus of this paper as it would require more cases of patients less than 12 years old for valid conclusions. Comparisons of demographics, locations, co-morbidities, and severity can be seen in Supplementary Table 3. Interpreting descriptive data should be done with great care and a control group of healthy children age-matched children would have been beneficial.

Conclusion

Pediatric HS is rare. Increased BMI may play a role, bearing in mind that the family unit may be an appropriate target for support and education when handling this. Similarly, although fewer pediatric patients were smokers themselves, almost half of them were exposed to passive smoking within the family.

It is further suggested that pediatric HS patients should be examined for hirsutism, pilonidal cysts, and endocrine disorders as these are not uncommon. A total of 96 of the 140 patients (69%) have agreed to be contacted again in the future for follow-up examination, allowing for the possibility to follow this unique cohort over time. This provides an opportunity to examine the evolution of HS over time in greater detail.

Compliance with ethical standards

Conflict of interest Gregor B.E. Jemec: Honoraria from AbbVie, Leo pharma, Pierre-Fabre, and Novartis for participation on advisory boards. Grants from Abbvie, Novartis, Regeneron and Leo Pharma for participation as an investigator. Research grants from Abbvie, Leo Pharma and Novartis. Ditte Marie Saunte was paid as a consultant for advisory board meeting by AbbVie, Janssen and Sanofi. She received speaker's honoraria and/or received grants from the following companies: Bayer, Abbvie, Desitin, Pfizer, Galderma, Astellas, Novartis, Sanofi, and Leo Pharma. José C. Pascual: Honoraria from Abbvie for participation on advisory boards. Jorge Romani: Honoraria from Ab-

bVie, Leo pharma, Almirall and Novartis for participation on advisory boards. Lisa Weibel received honoraria for participation on advisory boards from Sanofi, Merz, and Pierre-Fabre; for participating as study investigator from Abbvie and Novartis; and speaker's honoraria from Merz, Pierre Fabre, and MEDA Pharma. Philippe Guillem was paid as a consultant for advisory board meeting by AbbVie. He received as speaker's honoraria from AbbVie, Convatech, and Cicaplus. Axel P Villani received honoraria for participation on advisory boards from Bailleul and Merck. Other Authors stated no conflict of interest.

References

- Azziz R, Carmina E, Sawaya ME (2000) Idiopathic hirsutism. *Endocr Rev* 21:347–362. <https://doi.org/10.1210/edrv.21.4.0401>
- Bates CR, Buscemi J, Nicholson LM, Cory M, Jagpal A, Bohner AM (2018) Links between the organization of the family home environment and child obesity: a systematic review. *Obes Rev*. <https://doi.org/10.1111/obr.12662> (An official journal of the International Association for the Study of Obesity)
- Bettoli V, Ricci M, Zauli S, Virgili A (2015) Hidradenitis suppurativa-acne inversa: a relevant dermatosis in paediatric patients. *Br J Dermatol* 173:1328–1330. <https://doi.org/10.1111/bjd.13951>
- Braunberger TL, Nicholson CL, Gold L, Nahhas AF, Jacobsen G, Parks-Miller A, Hamzavi IH (2018) Hidradenitis suppurativa in children: the Henry Ford experience. *Pediatr Dermatol* 35:370–373. <https://doi.org/10.1111/pde.13466>
- Canoui-Poitrine F, Revuz JE, Wolkenstein P, Viallette C, Gabison G, Pouget F, Poli F, Faye O, Bastuji-Garin S (2009) Clinical characteristics of a series of 302 French patients with hidradenitis suppurativa, with an analysis of factors associated with disease severity. *J Am Acad Dermatol* 61:51–57. <https://doi.org/10.1016/j.jaad.2009.02.013>
- de Parades V, Bouchard D, Janier M, Berger A (2013) Pilonidal sinus disease. *J Visc Surg* 150:237–247. <https://doi.org/10.1016/j.jvisc.2013.05.006>
- Deckers IE, Janse IC, van der Zee HH, Nijsten T, Boer J, Horvath B, Prens EP (2016) Hidradenitis suppurativa (HS) is associated with low socioeconomic status (SES): a cross-sectional reference study. *J Am Acad Dermatol* 75(755–759):e751. <https://doi.org/10.1016/j.jaad.2016.04.067>
- Deckers IE, Kimball AB (2016) The handicap of hidradenitis suppurativa. *Dermatol Clin* 34:17–22. <https://doi.org/10.1016/j.det.2015.07.003>
- Deckers IE, van der Zee HH, Boer J, Prens EP (2015) Correlation of early-onset hidradenitis suppurativa with stronger genetic susceptibility and more widespread involvement. *J Am Acad Dermatol* 72:485–488. <https://doi.org/10.1016/j.jaad.2014.11.017>
- Dreno B, Poli F, Pawin H, Beylot C, Faure M, Chivot M, Aufret N, Moyse D, Ballanger F, Revuz J (2011) Development and evaluation of a Global Acne Severity Scale (GEA Scale) suitable for France and Europe. *J Eur Acad Dermatol Venereol* 25:43–48. <https://doi.org/10.1111/j.1468-3083.2010.03685.x>
- Esmann S, Jemec GB (2011) Psychosocial impact of hidradenitis suppurativa: a qualitative study. *Acta Derm Venereol* 91:328–332. <https://doi.org/10.2340/00015555-1082>
- Feito-Rodriguez M, Sendagorta-Cudos E, Herranz-Pinto P, de Lucas-Laguna R (2009) Prepubertal hidradenitis suppurativa successfully treated with botulinum toxin A. *Dermatol Surg* 35:1300–1302. <https://doi.org/10.1111/j.1524-4725.2009.01231.x>
- Garg A, Kirby JS, Lavian J, Lin G, Strunk A (2017) Sex- and age-adjusted population analysis of prevalence estimates for hidradenitis suppurativa in the United States. *JAMA Dermatol* 153:760–764. <https://doi.org/10.1001/jamadermatol.2017.0201>
- Garg A, Lavian J, Lin G, Strunk A, Alloo A (2017) Incidence of hidradenitis suppurativa in the United States: a sex- and age-adjusted population analysis. *J Am Acad Dermatol* 77:118–122. <https://doi.org/10.1016/j.jaad.2017.02.005>
- Garg A, Papagermanos V, Midura M, Strunk A (2018) Incidence of hidradenitis suppurativa among tobacco smokers: a population-based retrospective analysis in the USA. *Br J Dermatol* 178:709–714. <https://doi.org/10.1111/bjd.15939>
- Garg A, Strunk A, Midura M, Papagermanos V, Pomerantz H (2018) Prevalence of hidradenitis suppurativa among patients with Down syndrome: a population-based cross-sectional analysis. *Br J Dermatol* 178:697–703. <https://doi.org/10.1111/bjd.15770>
- Garg A, Wertenteil S, Baltz R, Strunk A, Finelt N (2018) Prevalence estimates for hidradenitis suppurativa among children and adolescents in the United States: a gender- and age-adjusted population analysis. *J Invest Dermatol*. <https://doi.org/10.1016/j.jid.2018.04.001>
- Ghods SZ, Orawa H, Zouboulis CC (2009) Prevalence, severity, and severity risk factors of acne in high school pupils: a community-based study. *J Invest Dermatol* 129:2136–2141. <https://doi.org/10.1038/jid.2009.47>
- Happle R, König A (2011) Smoker's boils. *Dermatology* 222:282–284. <https://doi.org/10.1159/000327923>
- Ibler KS, Jemec GB (2013) Cumulative life course impairment in other chronic or recurrent dermatologic diseases. *Curr Probl Dermatol* 44:130–136. <https://doi.org/10.1159/000350056>
- Janse I, Bieniek A, Horvath B, Matusiak L (2016) Surgical procedures in hidradenitis suppurativa. *Dermatol Clin* 34:97–109. <https://doi.org/10.1016/j.det.2015.08.007>
- Jemec GB (2012) Clinical practice hidradenitis suppurativa. *N Engl J Med* 366:158–164. <https://doi.org/10.1056/NEJMc1014163>
- Jemec GB, Kimball AB (2015) Hidradenitis suppurativa: epidemiology and scope of the problem. *J Am Acad Dermatol* 73:S4–7. <https://doi.org/10.1016/j.jaad.2015.07.052>
- Joseph MA, Jayaseelan E, Ganapathi B, Stephen J (2005) Hidradenitis suppurativa treated with finasteride. *J Dermatolog Treat* 16:75–78. <https://doi.org/10.1080/09546630510031403>
- Kjaersgaard Andersen R, Theut Riis P, Jemec GBE (2018) Factors predicting the self-evaluated health of hidradenitis suppurativa patients recruited from an outpatient clinic. *J Eur Acad Dermatol Venereol* 32:313–317. <https://doi.org/10.1111/jdv.14511>
- König A, Lehmann C, Rempel R, Happle R (1999) Cigarette smoking as a triggering factor of hidradenitis suppurativa. *Dermatology* 198:261–264. <https://doi.org/10.1159/000018126>
- Koshelev MV, Garrison PA, Wright TS (2014) Concurrent hidradenitis suppurativa, inflammatory acne, dissecting cellulitis of the scalp, and pyoderma gangrenosum in a 16-year-old boy. *Pediatr Dermatol* 31:e20–21. <https://doi.org/10.1111/pde.12196>
- Kromann CB, Deckers IE, Esmann S, Boer J, Prens EP, Jemec GB (2014) Risk factors, clinical course and long-term prognosis in hidradenitis suppurativa: a cross-sectional study. *Br J Dermatol* 171:819–824. <https://doi.org/10.1111/bjd.13090>
- Kromann CB, Ibler KS, Kristiansen VB, Jemec GB (2014) The influence of body weight on the prevalence and severity of hidradenitis suppurativa. *Acta Derm Venereol* 94:553–557. <https://doi.org/10.2340/00015555-1800>
- Levinson T, Sela T, Chencinski S, Derazne E, Tzur D, Elad H, Kreiss Y (2016) Pilonidal sinus disease: a 10-year review reveals occupational risk factors and the superiority of the minimal surgery trephine technique. *Mil Med* 181:389–394. <https://doi.org/10.7205/MILMED-D-14-00729>
- Lewis F, Messenger AG, Wales JK (1993) Hidradenitis suppurativa as a presenting feature of premature adrenarche. *Br J Dermatol* 129:447–448

32. Liy-Wong C, Pope E, Lara-Corrales I (2015) Hidradenitis suppurativa in the pediatric population. *J Am Acad Dermatol* 73:S36–41. <https://doi.org/10.1016/j.jaad.2015.07.051>
33. Mengesha YM, Holcombe TC, Hansen RC (1999) Prepubertal hidradenitis suppurativa: two case reports and review of the literature. *Pediatr Dermatol* 16:292–296
34. Mikkelsen PR, Jemec GB (2014) Hidradenitis suppurativa in children and adolescents: a review of treatment options. *Paediatr Drugs* 16:483–489. <https://doi.org/10.1007/s40272-014-0091-3>
35. Miller IM, Ellervik C, Vinding GR, Zarchi K, Ibler KS, Knudsen KM, Jemec GB (2014) Association of metabolic syndrome and hidradenitis suppurativa. *JAMA Dermatol* 150:1273–1280. <https://doi.org/10.1001/jamadermatol.2014.1165>
36. Miller IM, McAndrew RJ, Hamzavi I (2016) Prevalence, risk factors, and comorbidities of hidradenitis suppurativa. *Dermatol Clin* 34:7–16. <https://doi.org/10.1016/j.det.2015.08.002>
37. Mota F, Machado S, Selores M (2017) Hidradenitis suppurativa in children treated with finasteride—a case series. *Pediatr Dermatol* 34:578–583. <https://doi.org/10.1111/pde.13216>
38. Natarajan B, Sauer C, Shehata B, Kugathasan S (2015) Hidradenitis suppurativa and pediatric Crohn disease. *J Pediatr Gastroenterol Nutr* 60:e29–30. <https://doi.org/10.1097/MPG.0000000000000185>
39. Palmer RA, Keefe M (2001) Early-onset hidradenitis suppurativa. *Clin Exp Dermatol* 26:501–503
40. Pink AE, Simpson MA, Desai N, Dafou D, Hills A, Mortimer P, Smith CH, Trembath RC, Barker JNW (2012) Mutations in the gamma-secretase genes NCSTN, PSENEN, and PSEN1 underlie rare forms of hidradenitis suppurativa (acne inversa). *J Invest Dermatol* 132:2459–2461. <https://doi.org/10.1038/jid.2012.162>
41. Prabhu G, Laddha P, Manglani M, Phiske M (2012) Hidradenitis suppurativa in a HIV-infected child. *J Postgrad Med* 58:207–209. <https://doi.org/10.4103/0022-3859.101403>
42. Randhawa HK, Hamilton J, Pope E (2013) Finasteride for the treatment of hidradenitis suppurativa in children and adolescents. *JAMA Dermatol* 149:732–735. <https://doi.org/10.1001/jamadermatol.2013.2874>
43. Revuz J (2009) Hidradenitis suppurativa. *J Eur Acad Dermatol Venereol* 23:985–998. <https://doi.org/10.1111/j.1468-3083.2009.03356.x>
44. Santalahti P, Aromaa M, Sourander A, Helenius H, Piha J (2005) Have there been changes in children's psychosomatic symptoms? A 10-year comparison from Finland. *Pediatrics* 115:e434–442. <https://doi.org/10.1542/peds.2004-1261>
45. Sartorius K, Emtestam L, Jemec GB, Lapins J (2009) Objective scoring of hidradenitis suppurativa reflecting the role of tobacco smoking and obesity. *Br J Dermatol* 161:831–839. <https://doi.org/10.1111/j.1365-2133.2009.09198.x>
46. Sartorius K, Lapins J, Emtestam L, Jemec GB (2003) Suggestions for uniform outcome variables when reporting treatment effects in hidradenitis suppurativa. *Br J Dermatol* 149:211–213
47. Scheinfeld N (2015) Hidradenitis suppurativa in prepubescent and pubescent children. *Clin Dermatol* 33:316–319. <https://doi.org/10.1016/j.clindermatol.2014.12.007>
48. Schrader AM, Deckers IE, van der Zee HH, Boer J, Prens EP (2014) Hidradenitis suppurativa: a retrospective study of 846 Dutch patients to identify factors associated with disease severity. *J Am Acad Dermatol* 71:460–467. <https://doi.org/10.1016/j.jaad.2014.04.001>
49. Stojkovic-Filipovic JM, Gajic-Veljic MD, Nikolic M (2015) Prepubertal onset of hidradenitis suppurativa in a girl: a case report and literature review. *Indian J Dermatol Venereol Leprol* 81:294–298. <https://doi.org/10.4103/0378-6323.152741>
50. Teilmann G, Pedersen CB, Jensen TK, Skakkebaek NE, Juul A (2005) Prevalence and incidence of precocious pubertal development in Denmark: an epidemiologic study based on national registries. *Pediatrics* 116:1323–1328. <https://doi.org/10.1542/peds.2005-0012>
51. Theut Riis P, Saunte DM, Benhadou F, Del Marmol V, Guillem P, El-Domyati M, Abdel-Wahab H, Antoniou C, Dessinioti C, Gurer MA, Beksac B, Szepietowski JC, Matusiak L, Emtestam L, Lapins J, Riad H, Doss N, Massa AF, Hamzavi I, Nicholson C, Dolenc-Voljc M, Kim KH, Ohn J, Zouboulis CC, Karagiannidis I, Mokos ZB, Durinec P, Jemec GBE (2018) Low and high body mass index in hidradenitis suppurativa patients—different subtypes? *J Eur Acad Dermatol Venereol* 32:307–312. <https://doi.org/10.1111/jdv.14599>
52. Theut Riis P, Thorlacius L, Knudsen List E, Jemec GBE (2017) A pilot study of unemployment in patients with hidradenitis suppurativa in Denmark. *Br J Dermatol* 176:1083–1085. <https://doi.org/10.1111/bjd.14922>
53. Tiri H, Jokelainen J, Timonen M, Tasanen K, Huilaja L (2018) Somatic and psychiatric comorbidities of hidradenitis suppurativa in children and adolescents. *J Am Acad Dermatol*. <https://doi.org/10.1016/j.jaad.2018.02.067>
54. Uzuncakmak TK, Akdeniz N, Karadag AS (2018) Cutaneous manifestations of obesity and the metabolic syndrome. *Clin Dermatol* 36:81–88. <https://doi.org/10.1016/j.clindermatol.2017.09.014>
55. Vazquez BG, Alikhan A, Weaver AL, Wetter DA, Davis MD (2013) Incidence of hidradenitis suppurativa and associated factors: a population-based study of Olmsted County, Minnesota. *J Invest Dermatol* 133:97–103. <https://doi.org/10.1038/jid.2012.255>
56. Vinding GR, Miller IM, Zarchi K, Ibler KS, Ellervik C, Jemec GB (2014) The prevalence of inverse recurrent suppuration: a population-based study of possible hidradenitis suppurativa. *Br J Dermatol* 170:884–889. <https://doi.org/10.1111/bjd.12787>
57. Zouboulis CC, Desai N, Emtestam L, Hunger RE, Ioannides D, Juhasz I, Lapins J, Matusiak L, Prens EP, Revuz J, Schneider-Burrus S, Szepietowski JC, van der Zee HH, Jemec GB (2015) European S1 guideline for the treatment of hidradenitis suppurativa/acne inversa. *J Eur Acad Dermatol Venereol* 29:619–644. <https://doi.org/10.1111/jdv.12966>

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