Letters

RESEARCH LETTER

Prevalence of Dermatitis Herpetiformis Within the iCureCeliac Patient-Powered Research Network—Patient Characteristics and Dietary Counseling

Dermatitis herpetiformis (DH) is a cutaneous manifestation of celiac disease (CD). More than 90% of patients with DH have an associated gluten-sensitive enteropathy,[1,2] yet only 20% of patients with DH exhibit classic gastrointestinal symptoms at time of initial diagnosis.[1] Dermatologists may be the first to diagnose patients with CD via their DH. A paucity of nationwide data exists on the demographic characteristics of patients with DH and CD. Using a patient-powered research network (PPRN), we sought to (1) describe the prevalence of DH, (2) characterize the demographics of patients with DH and underlying CD, and (3) assess the extent of gluten-free diet (GFD) counseling received by patients with and without DH.

Supplemental content

Methods | Patients with DH were identified using the Celiac Disease Foundation's survey-based iCureCeliac PPRN (5807 volunteers enrolled online between 2015 and 2019), which has been previously validated for use.[3] All patients reported a diagnosis of CD, refractory CD, or DH, as outlined in the study inclusion criteria in the eFigure in the Supplement. Among the self-reported DH cohort, 2 case definitions were applied: broad DH indicated all patients with self-reported DH, and strict DH indicated self-reported DH with diagnosis confirmed by results of skin biopsy. The strict DH definition was used for comparisons. Data analysis included prevalence with 95% CI for each DH definition. Demographics were compared via frequency counts, percentages, means, χ^2 tests, and *t* tests. The association between initial diagnosis (DH vs CD) and receipt of GFD counseling was assessed via percentages and logistic regression odds ratios with 95% CI. The institutional review board at the University of Pennsylvania waived the need for study approval and patient informed consent owing to use of deidentified data that had previously been volunteered, collected, and stored within the Celiac Disease Foundation's database.

Table 1. Prevalence of Patients With Dermatitis Herpetiformis by Broad and Strict Definitions

Dermatitis herpetiformis definition applied	Analytic sample, No. (%) [95% CI] (n = 3775)
Broad definition	300 (7.95) [7.08-8.81]
Strict definition	82 (2.17) [1.71-2.64]

Table 2. Demographic Characteristics and Comparison of GFD Counseling of Patients With DH and CD as Defined by Strict Definitions[a]

Characteristic	Frequency, No./No. (%)	
	Strict DH (n = 82)	Strict CD (n = 3360)
Age, mean (SD), y		
At CD diagnosis[b]	40.46 (15.70)	33.25 (16.45)
At survey submission[b]	46.67 (14.42)	36.67 (16.99)
Gender		
Female	63/82 (76.83)	2789/3349 (83.28)
Male	19/82 (23.17)	560/3349 (16.72)
Race[c]		
White	77/79 (97.47)	3150/3313 (95.08)
Hispanic, Latin American, or Spanish	1/79 (1.27)	86/3313 (2.60)
Other	1/79 (1.27)	25/3313 (0.75)
Asian	0	24/3313 (0.72)
Native Hawaiian or Pacific Islander	0	1/3313 (0.03)
American Indian or Alaskan Native	0	7/3313 (0.21)
Black or African American	0	20/3313 (0.60)
Received counseling on GFD at time of initial diagnosis		
No[d]	11/80 (13.75)	192/3285 (5.84)
Yes	69/80 (86.25)	3093/3285 (94.16)

Abbreviations: CD, celiac disease; DH, dermatitis herpetiformis; GFD, gluten-free diet; PPRN, patient-powered research network.

[a] For each question in the analytic PPRN, some responses were missing. The denominator listed represents the total responses received.

[b] The *t*-test value was $P < .001$.

[c] Only ethnicities with at least 1 person reported in the PPRN are included.

[d] Odds ratio adjusted for gender and age at diagnosis was 2.58 (95% CI, 1.34-4.99).

Results | The prevalence of DH in the iCureCeliac PPRN is outlined in **Table 1**, with 82 of 3775 patients (2.2%) meeting the strictest inclusion criteria. The demographic characteristics and odds of receiving GFD counseling in DH vs CD patients (by strictest definitions) are presented in **Table 2**. Both DH and CD groups were predominantly White (97.5% vs 95.1%; $P = .91$) and female (76.8% vs 83.3%; $P = .12$). The DH and CD groups were significantly different in mean age at CD diagnosis and age at survey response (Table 2). The DH group was older at diagnosis (40.5 vs 33.3 years; $P = .001$) and time of survey completion (46.7 vs 37.7 years; $P = .001$). Eleven of 82 (13.4%) patients with DH and 192 of 3360 (5.7%) patients with CD reported no dietary counseling at initial diagnosis. After adjusting for gender and age at diagnosis, patients with DH had twice the odds of not recalling counseling on a GFD compared with patients with CD but without DH (odds ratio, 2.58; 95% CI, 1.34-4.99).

Discussion | In this survey study of patients with CD participating in the PPRN iCureCeliac, we found the prevalence of DH ranged from 2.17% to 7.95% depending on disease definition. Prior studies, largely retrospective medical record reviews, have

demonstrated approximately 5% to 15% of patients with CD exhibit DH.[2,4] The majority of patients in the present study were White and female, which aligns with prior studies using this PPRN[3] and reports that CD is more prevalent in women of northern European descent.[1,4] To our knowledge, no DH data have been previously published from this PPRN. While 300 patients reported a diagnosis of DH, only 82 reported confirmation by results of skin biopsy.

Patients with CD carry an increased risk of enteropathy-associated T-cell lymphoma owing to long-term gluten exposure and intestinal inflammation[5] with evidence suggesting the risk may be mitigated by gluten restriction.[5] A GFD is especially important in the first 5 years following diagnosis because increased lymphoma mortality exists during this window.[6] Because a majority of patients with DH have underlying enteropathy,[1] and dermatologists may be the first to diagnose patients with CD, dermatologists should also be prepared to counsel on a GFD and/or arrange follow-up with a gastroenterologist or dietitian. Results of the present study suggest a potential practice gap because patients with DH exhibited decreased odds of recalling counseling on a GFD at time of diagnosis when compared with patients with CD but without DH.

Inherent limitations of this study include recall bias, especially when the diagnosis of CD preceded survey completion by many years, and lack of detailed, skin-specific data. For example, we were unable to assess age at DH diagnosis. Further prospective studies are needed to better understand the DH population, specifically if comorbidities and prognosis mirror those in patients with CD but without DH.

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