

Prevalence and incidence of palmoplantar pustulosis in Sweden: a population-based register study*

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Summary

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Conflicts of interest

M.S.E. is responsible for dermatology in the project management for the national guidelines for psoriasis at the Swedish Board of Health and Welfare. J.M.N. and S.L. have been involved in the health economic analyses of the national guidelines for psoriasis at the Swedish Board of Health and Welfare.

Data availability

The data that support the findings of this study are available from the Swedish National Board of Health and Welfare and the Swedish Tax Agency. Restrictions apply to the availability of these data, which were used under license for this study.

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Background Palmoplantar pustulosis (PPP) is a chronic relapsing skin condition characterized by sterile pustules on the palm and soles. Population-based estimates of PPP incidence and prevalence are limited.

Objectives To estimate the prevalence and incidence of PPP in the Swedish general population and to estimate the prevalence of psoriasis vulgaris among the population with PPP.

Methods The Swedish National Patient Register was used, covering all inpatient and outpatient nonprimary care for the Swedish population. We identified cases (2004–2015) with one International Classification of Diseases 10th Revision diagnostic code (base case) for PPP. The point prevalence estimates at the end of this period (31 December 2015) were obtained by linkage to the Swedish Total Population Register. In sensitivity analyses, we used alternative case definitions: (i) requiring two visits and (ii) requiring two visits, one of which was within dermatology or internal medicine.

Results The base case prevalence of PPP was estimated to be 147 per 100 000 (women 227, men 68) and the annual prevalence was estimated to 26 per 100 000 in 2015. Among the population of people with PPP, 17% were registered with a diagnostic code for psoriasis vulgaris. The incidence of PPP in 2015 was estimated to be 12.7 per 100 000 (women 18.7, men 6.6). The criteria used had an impact on the prevalence and incidence estimates: strict case 1 gave an overall prevalence of 72 per 100 000 and an incidence of 5.4 per 100 000.

Conclusions The results indicate that the population-based prevalence of PPP may be larger than previously estimated. However, the estimates were sensitive to the employed PPP case criteria. The findings enhance demands for studies using validated diagnostic algorithms potentially also including data from primary care.

What is already known about this topic?

- There are few published estimates of the prevalence and incidence of palmoplantar pustulosis (PPP).
- The previous studies are mainly based on short observation periods and often lack coverage of the general population.
- Previous studies, with use of different case definitions and prevalence measures, show large variations (range from 50 to 120 per 100 000) in the prevalence estimates of PPP.

What does this study add?

- We found a population-based point prevalence on 31 December 2015 of 147 per 100 000, an annual prevalence of 26 per 100 000, and an incidence estimate of 12.7 cases of PPP per 100 000 in 2015.
- The population of people with PPP may be larger than previously estimated.

- Our estimates were sensitive to the case definition of PPP, highlighting the need for validated diagnostic algorithms.
- Comparisons of incidence and prevalence across studies must be done with caution due to differences in methodology.

Palmoplantar pustulosis (PPP) is a chronic relapsing skin condition characterized by sterile pustules together with redness and scaling on the palm and soles, which erupt over time. Traditionally PPP has been classified as a subgroup of psoriasis, as it is often seen in combination with psoriasis vulgaris.^{1,2} Smoking, infection and certain drugs trigger or exacerbate the disease.³ Many people who have PPP experience reduced quality of life, and one study reported that PPP is associated with greater impairment of quality of life than moderate-to-severe psoriasis.⁴

Accurate population-based prevalence and incidence estimates are of importance in the understanding of the burden of disease. Furthermore, they constitute a backbone for epidemiological research. Studies from across the world have reported prevalence estimates of PPP ranging from 0.001% to 0.12%.^{1,5} The estimated proportion of patients with psoriasis vulgaris among those with PPP has varied between 14% and 60%.^{1,6} PPP is more frequent in women than in men, with reported male-to-female ratios of 0.21–0.52,⁵ and the median age at onset has been found to be between 45 and 65 years.^{2,6} Prevalence estimates of PPP using population-based administrative databases have recently been determined in Denmark and Japan.^{1,7}

In the current study we used longitudinal healthcare register data covering the total Swedish population (~9.9 million in 2015).⁸ The usual pathway into healthcare in Sweden is by a visit to a general practitioner. People in need of secondary care are then referred by the general practitioner to specialized care, but people can access secondary care directly under certain circumstances. The study objectives were to estimate the prevalence and incidence of physician-diagnosed PPP in a population-based healthcare register. Analyses were stratified by sex and age. Prevalence of psoriasis vulgaris in the PPP population was also estimated.

Patients and methods

National Patient Register

The Swedish National Patient Register (NPR) is a national individual-level data register covering all inpatient care since 1987 and all secondary outpatient care since 2001. The NPR contains diagnostic codes (International Classification of Diseases 10th Revision; ICD-10) and admission and discharge dates from both private and public caregivers. NPR does not cover primary care or healthcare provided by caregivers other than physicians. In 2019 the loss of primary diagnosis was

0.9% for inpatient care and 3.2% for outpatient care. The NPR has been described in detail elsewhere.⁹

Total Population Register

The Total Population Register (TPR) is the civil registration of vital events (e.g. births and deaths) of all Swedish inhabitants, administrated by the Swedish Tax Agency. The register is continuously updated and is used for a variety of purposes by healthcare providers and medical researchers.¹⁰ In TPR, all citizens are identified by their unique personal identification number (PIN). By law, all healthcare provided must be registered by the patient's PIN, which is automatically assigned to all residents. Information on the Swedish population size was collected from public statistics at Statistics Sweden.⁸

Source population and case definitions

During 2004–2015 we identified all cases with a primary or secondary diagnosis for psoriasis in the NPR (Table S1; see Supporting Information). The authors (S.L. and J.M.N.) had fully access to this data extraction. Out of these cases we selected those of all ages with a diagnosis of PPP (code L40.3).

There are no validation studies of PPP diagnostic codes in the NPR. Therefore, we used different case criteria to define cases of PPP. Primary care is usually the first step in addressing dermatology healthcare needs. However, the NPR does not hold information on primary care. Therefore, as patients with less severe PPP are managed mainly in primary care, we required only one visit with a diagnostic code of L40.3 in specialized care in the base case scenario. In a sensitivity analysis, we used a strict criterion, requiring registration in the NPR of two physician visits (strict criteria 1). This probably mirrors cases with more severe disease. In a third alternative, we required two physician visits of which at least one visit was to dermatology or internal medicine (strict criteria 2), as dermatology is sometimes incorporated into an internal medicine department in Sweden.

Among the base case PPP population, we identified those with a diagnostic code indicating psoriasis vulgaris using ICD-10 codes L40.0 (psoriasis vulgaris) and L40.9 (psoriasis, unspecified). The rationale for using two different codes is that there is reason to believe that both codes are used for the diagnosis of psoriasis vulgaris in clinical practice.

Prevalence estimates of physician-diagnosed palmoplantar pustulosis

The different case criteria were applied to identify patients diagnosed with PPP at any time during 1 January 2004 to 31 December 2015. By means of the individuals' PINs, data were linked from the NPR to the TPR to exclude those who were no longer alive or no longer residents in Sweden by the end of 2015. Hence, the point prevalence of physician-diagnosed PPP on 31 December 2015 was estimated by dividing the number of cases who met our inclusion criteria by the number of current Swedish residents by the end of December 2015. To facilitate comparisons with other studies, we also estimated the annual prevalence in 2015 using the base case in the main analysis. The number of unique persons who had a physician visit with a diagnosis of PPP in 2015 was divided by the total Swedish population for that year. In sensitivity analyses we used three additional case definitions (PPP within dermatology or internal medicine, PPP within dermatology only, and PPP as a primary diagnosis within dermatology or internal medicine) in the estimate of the annual prevalence in 2015 (Table S2; see Supporting Information).

Prevalence of psoriasis vulgaris in the palmoplantar pustulosis population

The denominator for the calculation of psoriasis vulgaris prevalence was the base case PPP population. The date of the first recorded diagnosis for PPP was defined as the index date. Patients with co-occurring psoriasis vulgaris were identified with at least one L40.0 or L40.9 diagnostic code within 365 days before and after the index date.

Incidence estimates of physician-diagnosed palmoplantar pustulosis

We estimated the incidence of PPP for the year 2015. We considered all persons who presented as incident cases according to our case definitions. Incident cases were defined as those not having any registrations of such diagnoses in the NPR in the previous 10 years (2004–2013). For the base case definition, incidence was defined as the first ever diagnosis of PPP in 2015. For the stricter case definitions we regarded the second visit with PPP as the 'incidence visit', meaning that we also included persons who had one visit with the diagnosis in 2014 and the second visit for PPP in 2015. The Swedish population on 31 December 2014 served as the basis for the denominator for both the base case and stricter case definitions. To compensate for prevalent cases (not at risk of disease) the denominator was reduced by the prevalence estimates obtained for age and sex strata.

Statistical analysis

All estimates were presented in total, and across sexes and age groups (10-year bands). The estimates were calculated using a binomial distribution with 95% confidence intervals (CIs).

Analyses were performed using Stata statistical software, version Stata/IC 14.2 (StataCorp, College Station, TX, USA).

Ethical approval

This study was conducted according to the Declaration of Helsinki and was approved by the regional ethical review board at Umeå University.

Results

Prevalence

During the 12-year observational period (2004–2015) we identified 153 733 cases of all ages with a primary or secondary diagnosis for psoriasis in inpatient or nonprimary outpatient care (Figure 1). Of those, 15 654 (10.2%) had received a PPP diagnosis, and 14 494 (9.4%) had a diagnosis for PPP (12 095, 7.9% as primary diagnosis) and were alive and living in Sweden at the end of 2015. Additional information about the case characteristics are presented in Table S3 (see Supporting Information).

Applying the base case definition, we identified 14 494 persons (76.9% women) with PPP who were still alive and residents in Sweden by the end of 2015, resulting in a prevalence of 147 cases of PPP per 100 000 in Sweden (Table 1). The mean age (SD) of the prevalent cases of PPP was 60.6 (14.7) years for women and 57.9 (16.9) years for men. Women had a higher prevalence of PPP than men, with a ratio of 3.3 (women 227, men 68 per 100 000). The annual prevalence in 2015 was estimated to be 26 cases of PPP per 100 000, with a similar female-to-male ratio of 3 (women 39, men 12 per 100 000).

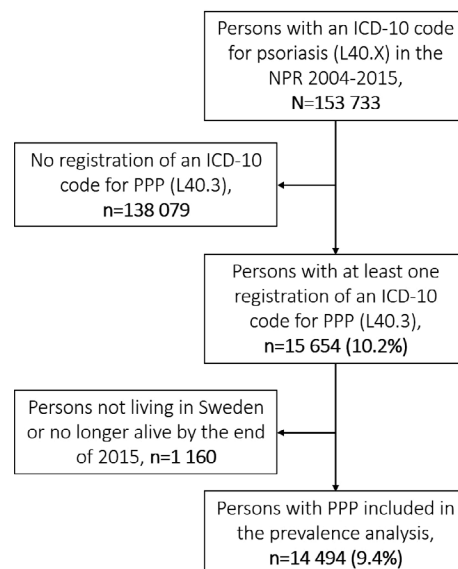


Figure 1 Flowchart of inclusion and exclusion in the palmoplantar pustulosis (PPP) study population. ICD-10, International Classification of Diseases, 10th Revision; NPR, National Patient Register.

Table 1 Prevalence estimates of palmoplantar pustulosis (PPP) per 100 000 women and men in Sweden, 2015

Prevalent PPP, 31 December 2015	Women	Men	Total
Cases in NPR (base case)	11 145	3349	14 494
Cases in NPR (strict case 1)	5665	1472	7137
Cases in NPR (strict case 2)	5098	1303	6401
Swedish population, 31 December 2015	4 920 051	4 930 966	9 851 017
Age (years), mean (SD) ^a	60.6 (14.0)	57.9 (16.9)	60.6 (14.7)
Base case prevalence, n/100 000 (95% CI)	227 (222–231)	68 (66–70)	147 (145–150)
Strict case 1 prevalence, n/100 000 (95% CI)	115 (112–118)	30 (28–31)	72 (71–74)
Strict case 2 prevalence, n/100 000 (95% CI)	104 (101–107)	26 (25–28)	65 (63–67)
Annual prevalence, n/100 000 (95% CI) ^a	39 (38–41)	12 (11–13)	26 (25–27)
Prevalence of PV in the PPP population, % (95% CI) ^a	16.1 (15.3–16.8)	21.2 (19.7–22.8)	17.2 (16.6–17.9)

CI, confidence interval; NPR, National Patient Register; PV, psoriasis vulgaris. ^aBase case criteria population.

The point prevalence estimate was substantially reduced from the base case scenario (from 147 to 72 per 100 000) by requiring two visits with a diagnosis of PPP (strict case 1). The prevalence changed to 65 per 100 000 using strict case 2. The annual prevalence in 2015 change to 22.7 when requiring a PPP diagnosis in a dermatology or internal medicine department and to 19.8 when also requiring the diagnosis to be primary (Table S2; see Supporting Information).

Among the overall base case population, 17.2% had a registered diagnostic code for psoriasis vulgaris within 365 days before or after the index date. The proportion of persons with a psoriasis vulgaris diagnostic code was significantly higher for men (21.2%, 95% CI 19.7–22.8) than for women (16.1%, 95% CI 15.3–16.8).

The point prevalence of PPP at the end of 2015 was higher for women than for men across all age bands (Figure 2; and Table S4; see Supporting Information). The base case prevalence peaked at age 60–69 years for women (626 per 100 000) and at age 60–79 years for men.

Incidence

Using the base case definition (first ever visit in 2015) we identified 1233 persons (79.8% women), resulting in an

overall incidence of 12.7 cases of PPP per 100 000 (Table 2). The mean age (SD) of the incident cases was 56.4 (15.9) years, with no sex differences. The incidence per 100 000 was 18.7 (95% CI 17.5–20.0) in women and 6.6 (95% CI 5.9–7.4) in men.

The incidence decreased by more than half (from 12.7 to 5.4 per 100 000) when the strict criteria 1 were used (two first ever visits in 2015 or first ever visit in 2014 and second in 2015). The incidence changed to 5.1 per 100 000 with strict criteria 2.

For men, the incidence peaked in the 70–79-year age group with 14.6 per 100 000, while the incidence peak for women was in the 50–59-year age group with 47.7 per 100 000 (Figure 3; and Table S5; see Supporting Information). However, the CIs overlapped in the age groups 50–79 years for both men and women.

Discussion

This nationwide register-based study provides novel information on the prevalence and incidence of PPP. We estimated the point prevalence (PPP diagnosis 2004–2015, alive in Sweden at the end of 2015) of PPP as 147 cases per 100 000, and the annual prevalence as 26 per 100 000 in 2015. The

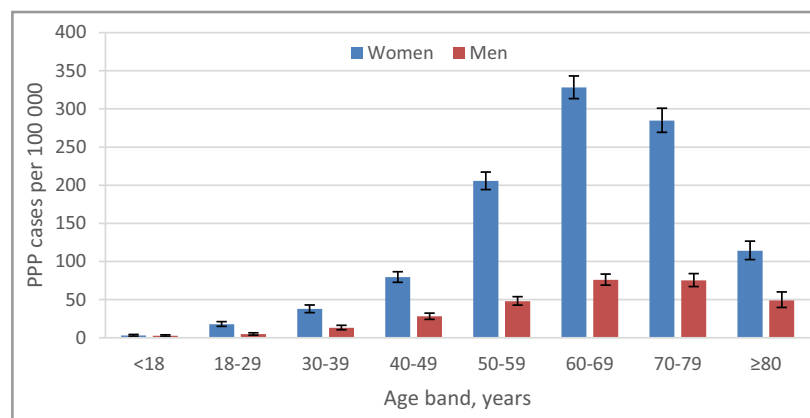
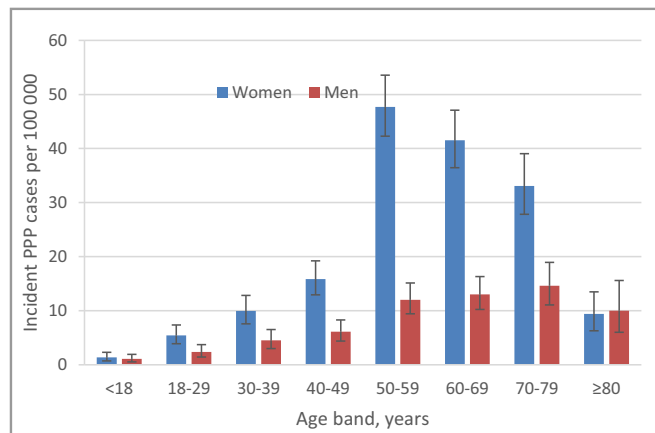


Figure 2 Prevalence estimates and 95% confidence intervals for palmoplantar pustulosis (PPP) in Sweden at the end of 2015, stratified by age and sex. The base case definition was used.

Table 2 Incidence estimates of palmoplantar pustulosis (PPP) per 100 000 women and men in Sweden, 2015

Incident PPP, 2015	Women	Men	Total
New cases in NPR (base case)	910	323	1 233
New cases in NPR (strict case 1)	392	135	527
New cases in NPR (strict case 2)	368	126	492
Swedish population without PPP, 31 December 2014	4 869 842	4 870 903	9 740 745
Age (years), mean (SD) ^a	56.7 (15.0)	55.5 (18.3)	56.4 (15.9)
Base case incidence, n/100 000 (95% CI)	18.7 (17.5–20.0)	6.6 (5.9–7.4)	12.7 (12.0–13.4)
Strict case 1 incidence, n/100 000 (95% CI)	8.1 (7.2–8.9)	2.8 (2.3–3.3)	5.4 (5.0–5.9)
Strict case 2 incidence, n/100 000 (95% CI)	7.6 (6.8–8.4)	2.6 (2.2–3.1)	5.1 (4.6–5.5)

CI, confidence interval; NPR, National Patient Register; PV, psoriasis vulgaris. ^aBase case criteria population.

**Figure 3** Incidence estimates and 95% confidence intervals for palmoplantar pustulosis (PPP) in Sweden at the end of 2015, stratified by age and sex. The base case definition was used.

advantage of retrospectively identifying all cases diagnosed during 2004–2015 for the calculation of point prevalence at the end of 2015 is that prevalent cases are more comprehensively captured than is possible in a narrower timeframe. Psoriatic conditions have a relapsing and remitting nature, and therefore persons with PPP may not visit specialized care on an annual basis.¹¹ The incidence was estimated to be 12.7 per 100 000 in 2015. The estimates were sensitive to the criteria used: both the prevalence and incidence decreased by over a factor of two (from 147 to 72 per 100 000 and from 12.7 to 5.4 per 100 000, respectively) when using strict criteria.

Given the relapsing and remitting course of PPP and as there are no validation studies of the diagnosis of PPP in NPR we estimated prevalence and incidence using different case criteria. We found a large difference between the estimates using the base criteria compared with the stricter criteria. One interpretation of this difference is that most patients with psoriasis and PPP seek initial assessment in primary care, where primary care physicians diagnose and treat these diseases. In the case of psoriasis vulgaris, patients with mild disease requiring topical treatment only are managed mainly in primary care, while patients with moderate-to-severe psoriasis are managed mainly in specialist care. We anticipated a similar pattern for PPP. However, even mild cases of PPP are

occasionally referred to specialized dermatology care, for example to verify the diagnosis, due to a disappointing treatment response, or just due to the chronicity of the disease, which can frustrate primary care physicians and patients alike.¹² Thus, while mild cases of PPP are likely insufficiently captured in the NPR, which is based on specialized care, relying on only one diagnostic code likely more closely approximates the real prevalence, under the assumption that at least a proportion of patients with PPP primarily managed in primary care have had at least one specialist visit.

Other factors may have contributed to the differences between the case definitions. Firstly, there is a shorter time to receive a second diagnosis for those included with a first diagnosis near the end of the observational period. However, this likely explains only a minor part of the differences as, out of those with two or more diagnoses of PPP, 61% received the first and second diagnoses in the same year (Table S3). The median number of days between the first and second PPP diagnoses was 89 (Table S3). Secondly, the base criteria may include persons where the first PPP diagnosis turned out to be another condition or disease (i.e. misclassification). Thirdly, the discrepancies may reflect the access to specialist care. Access to specialized dermatological care may differ due to factors such as availability of dermatologists.¹³

There is a dearth of studies on the epidemiology of PPP, and existing studies often lack an adequate representation of the general population. This is the largest study in recent years to comprehensively estimate a population-based point prevalence of physician-diagnosed PPP based on diagnoses made over an extensive (12-year) timeframe, and to our knowledge, the first population-based study of PPP incidence.

A Swedish dermatology department-based study from 1971, with 85 outpatient cases clinically examined in 1964–65, reported a PPP period prevalence of 50 per 100 000 with a male-to-female ratio of 0.22 and onset age of 40–59 years.¹⁴ In our study the point prevalence was higher irrespectively of the case definition used, and the male-to-female ratio (base criteria) was marginally higher, at 0.26. The mean age for incident cases in 2015 was 57 years, which was within the age onset range reported by Hellgren and Mobacken.¹⁴ The lower prevalence observed in their study may be explained by a shorter inclusion time (2 years), only inclusion of outpatient cases, and case definition by clinical examination.

We found three national population-based studies, based on claims data, estimating the annual prevalence of PPP, showing prevalence in Germany (2005),¹⁵ Japan (2010–11)⁷ and Korea (2015)¹⁶ of 90, 120 and 52 cases per 100 000, respectively. In a recent multinational study, the annual PPP prevalences in the USA (2015–16), Denmark (2015–16) and Germany (2014) were estimated to be 9, 5 and 80 cases per 100 000, respectively.¹ Healthcare insurance claims data (USA and Germany) and data from the Danish NPR were used.

Compared with the overall annual prevalence range from five cases in Denmark to 120 cases in Japan, the annual prevalence estimate of 26 cases per 100 000 in 2015 in the present study was in the lower range. The difference in the annual prevalence between Sweden and Denmark was somewhat surprising as the data sources are quite similar; they are both population-based registers covering inpatient and outpatient nonprimary care. One factor that might have contributed to the difference is that in Denmark the PPP diagnosis was verified by a dermatologist, which was not the base case for Sweden. Restricting our population to the Danish criteria, requiring a dermatology-verified diagnosis, the annual prevalence was reduced from 26 (base case) to 22.1 cases per 100 000 population. In an additional analysis requiring a dermatology- or internal medicine-verified primary PPP diagnosis the prevalence decreased further to 19.8 cases per 100 000 (Table S2), which is still higher than the estimates published for Denmark. There may also be differences in the healthcare visit patterns and differences in what clinicians define as PPP.

Overall, it is difficult to discern any clear pattern as to why there is such a wide variation in the prevalence between the studies above. It is thus also difficult to say anything about the external validity of our study results. Variations may be explained by differences in methodology such as case definitions, type of prevalence, target populations, and sex and age patterns, but also by use of unvalidated case definitions. The lack of generally accepted diagnostic and classification criteria for PPP, and lack of clarity whether PPP is a distinct entity or

a subgroup of psoriasis,^{17,18} may underpin variations in how this disease is diagnosed in clinical practice. Ongoing work to achieve consensus on the phenotypes for the disease group of pustular psoriasis¹⁹ is consequently of particular value.

We found that PPP was more common in women than in men (male-to-female ratio of 0.26), which is in line with estimates from other studies (range 0.21–0.53).^{2,7,14,20} Psoriasis vulgaris occurred in 17% (n = 2500) of the PPP population. Most patients (63%) were diagnosed with only code L40.0, followed by only L40.9 (27%). Our result corroborates findings reporting a psoriasis vulgaris occurrence of around 15%.^{1,3,21,22} However, some studies have reported higher prevalence (range 36–60%) of psoriasis vulgaris in PPP populations.^{1,2}

This study presents estimates of PPP prevalence and incidence within the context of a large population-based register. Of particular value was the use of the NPR on inpatient and outpatient specialist care visits covering the whole Swedish population for an extensive period. Using this large source with routinely collected data in clinical practice, prevalence and incidence could be estimated by subgroups of interest. In addition, we employed different case definitions to increase the utility for comparison with future studies.

There are some limitations to be considered. Firstly, as there is no standard case definition for the diagnosis of PPP in Sweden, our case definitions were based on coded diagnoses of PPP and not on classification criteria or validation through medical record review, and thus are potentially subject to misclassification bias. Furthermore, the study design did not allow analysis of potential misclassification due to drug-induced dermatological reactions similar to PPP skin symptoms. However, to mitigate misclassification bias, we used both sensitive and more specific case definitions. Secondly, our sample excluded people with PPP not seeking healthcare and those only seen in primary care. However, a psoriasis study shows that including primary care data will result in cases added, but it may happen at the expense of a larger degree of misclassification.²³

In conclusion, we studied the occurrence of PPP to a limited extent. We estimated the Swedish nationwide population-based PPP point prevalence at the end of 2015 to be 147 cases per 100 000 and the annual prevalence in 2015 to be 26 cases per 100 000. We demonstrate that the PPP population may be larger than previous estimated in 1-year prevalence studies. The estimates were sensitive to which case criteria definition was used; our findings thus highlight the need for studies using validated diagnostic algorithms potentially also including data from primary care. Treatment options for PPP are currently limited, but future therapies could increase the demand for research in PPP for further understanding of the prevalence and incidence of the disease.

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Supporting Information

Additional Supporting Information may be found in the online version of this article at the publisher's website:

Table S1. International Classification of Diseases 10th revision codes.

Table S2. Annual prevalence of palmoplantar pustulosis (PPP) in 2015 across different criteria for PPP.

Table S3. Additional characteristics of the case population.

Table S4. Point prevalence of palmoplantar pustulosis by age and sex.

Table S5. Incidence of palmoplantar pustulosis by age and sex.

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