# Prevalence of Cold Dermatoses in Kashmir Valley: A Cross-Sectional Study from North India

#### Abstract

Background: Often quoted as "heaven on earth," Kashmir forms one of the two divisions of the Union territory of Jammu and Kashmir. A high-altitude region with abundant precipitation and snowfall, the people of Kashmir experience peculiar dermatoses not commonly seen in the majorly tropical subcontinent of India. In this study, we focussed on cold dermatoses as a comprehensive cluster and attempted to study them as a group. Aims: To determine the prevalence of cold dermatoses in Kashmir valley and study their epidemiological characteristics. Methods: This observational, cross-sectional community-based study was conducted on native Kashmiri population in three districts of the valley, exclusively during the winter season of the year 2016–17 and 2017–18. The data were tabulated and analyzed with Chi-square test for discrete variables and *t*-test for continuous variables, using OpenEpi. A P value of less than 0.05 was taken as significant. Results: The study included a total of 1200 cases with 602 males and 598 females. Perniosis was most commonly encountered dermatoses in our study with a prevalence of 12.2%. Frostbite had a prevalence of 0.83%. Raynaud's phenomenon and asteatotic eczema were seen in 1.5% and 1.67% of the population, respectively. Cold panniculitis, cold urticaria, and livedo reticularis were each seen in 0.08% of the population. Conclusions: Cold dermatoses form an important source of morbidity among the native population of Kashmir. These can be easily prevented by ensuring adequate protection against cold. Creating awareness regarding these disorders and probable association with connective tissue disorders is also imperative.

**Keywords:** Asteatotic eczema, cold urticaria, cold weather injuries, Kashmir, Raynaud's phenomenon

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## Introduction

Kashmir is one of the administrative of Jammu and divisions Kashmir situated at an average height of 1850 m (6070 feet) above sea level. The main Kashmir valley is 100 km (62 miles) wide and 15,520.3 km<sup>2</sup> (5992.4 square miles) in area.<sup>[1]</sup> The climate of Kashmir varies greatly owing to its rugged topography. The typical climatic conditions in Kashmir, sudden and unexpected changes in weather, subzero temperatures in winter, and abundant precipitation predispose people of the valley to peculiar dermatological problems.

Exposure to cold environment for prolonged periods can have a multitude of adverse effects on the skin. When exposed to low temperature, blood vessels in skin undergo vasoconstriction and viscosity of blood increases, resulting in a decrease in blood supply and oxygen delivery to the skin.

Vasoconstriction of arterioles and veins occurs by a direct mechanism mediated in part by the endothelial synthesis of vasoconstrictor peptide - Endothelin-1.<sup>[2]</sup> Persistent exposure to cold leads to "Hunting reaction of Lewis," characterized by repeated cycles of vasodilatation following periods of vasoconstriction. However, when the core temperature is under threat. Hunting reaction stops and vasoconstriction persists.<sup>[3]</sup> Increased viscosity of blood, the shift of oxyhemoglobin dissociation curve to the left, diminished conduction velocity in cutaneous nerves, and change in platelet adhesiveness<sup>[4]</sup> are other physiological effects of cold.

Owing to the paucity of prevalence studies for cold dermatoses, this study offers a novel look at the trends of cold dermatoses in Kashmir valley. To our knowledge, a similar study has not yet been conducted

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anywhere in India or even abroad till date. All the studies that we came across, during an extensive review of literature, focused on a single disorder and never on this entire group of disorders, and they were either conducted in a hospital setup or on some fixed cohort groups such as workers in an establishment, soldiers, figure skaters, mountaineers, etc., and never in a community setup.<sup>[5-19]</sup> In this one of a kind study, we focussed on cold dermatoses as a comprehensive cluster and attempted to study these disorders as a group.

## **Methods**

The study was approved by the institutional ethics committee and informed consent was obtained from all participants.

This study was an observational, cross-sectional community-based study conducted on native Kashmiri population in three districts of the valley, exclusively during the winter season (December, January, and February) of the year 2016-17 and 2017–18. The sample size was decided as 1200 after consultation with a statistician and using OpenEpi software.

To avoid any selection bias, the selection of focus spots for the study was done randomly from Kashmir valley, by multistage cluster sampling.

Demographic details, clinical data (including patient's age, sex, weight, height, lesion type, its duration, and history of evolution and progression) and relevant details of systemic and mucocutaneous examination were noted and recorded on specially designed proformas.

Data were tabulated and analyzed with Chi-square test for discrete variables and *t*-test for continuous variables using OpenEpi. A P value of less than 0.05 was taken as significant.

## **Inclusion criteria**

All native Kashmiris (all ages, both sex groups, all tribes and religious groups) who gave consent for participation in the study were included.

## **Exclusion criteria**

Non-native workers, tourists, and soldiers and people who did not consent to be a part of the study were excluded.

## Results

The study included a total of 1200 cases comprising of 602 males and 598 females [Table 1].

Perniosis was the most commonly encountered cold dermatoses in our study, seen in 146 cases with a prevalence of 12.2% (95% CI, 10.4–14.1%, n = 146/1200).

Out of the 146 cases of perniosis, 84 were females and the remaining 62 were males. The prevalence in females

was found to be 14% (n = 84/598) while that in males was 10.3% (n = 62/602). The difference was statistically significant with a *P* value of 0.047.

The prevalence of perniosis in a study population decreased progressively with age. It was most common in the age group of 0-10 years with an age-specific prevalence of 20.1% [Table 2].

A family history of similar complaints in a first degree relative was present in 86 (58.9%) cases as opposed to 60 (41.1%) cases who did not report any such history. The correlation was found to be statistically significant (*P*-value = 0.0000001).

Around 22 cases (15.07%) reported the occurrence of perniosis for the first time while the remaining 124 cases (84.93%) had experienced one or more recurrences in the previous winter with complete remission during the summer season.

Nearly 98 patients (67.12%) had their first episode of perniosis during the second or third year of life, an additional 34 patients (23.29%) experienced the same before 18 years of age while only 14 patients (9.59%) had their first episode during adulthood. Thus, more than 90% of the patients had their first episode during childhood and the occurrence was found to be statistically significant (*P*-value = 0.0000001).

The sites of involvement and common presentations of perniosis seen in our study are documented in

Table 1: Age distribution of study population				
Age group	Number	Percentage		
0-10 years	268	22.3% ( <i>n</i> =268/1200)		
11-20 years	262	21.8% ( <i>n</i> =262/1200)		
21-30 years	202	16.8% ( <i>n</i> =202/1200)		
31-40 years	184	15.3% ( <i>n</i> =184/1200)		
41-50 years	122	10.2% ( <i>n</i> =122/1200)		
51-60 years	90	7.5% ( <i>n</i> =90/1200)		
61-70 years	50	4.2% ( <i>n</i> =50/1200)		
71-80 years	20	1.7% ( <i>n</i> =20/1200)		
81-90	2	0.2% ( <i>n</i> =2/1200)		
Total	1200	100%		

Table 2: Age-specific prevalence of perniosis		
Age group	Prevalence	
0-10 years	20.1% ( <i>n</i> =54/268)	
11-20 years	16% ( <i>n</i> =42/262)	
21-30 years	12.9% ( <i>n</i> =26/202)	
31-40 years	6.5% ( <i>n</i> =12/184)	
41-50 years	4.9% ( <i>n</i> =6/122)	
51-60 years	4.4% ( <i>n</i> =4/90)	
61-70 years	4% ( <i>n</i> =2/50)	
71-80 years	0 ( <i>n</i> =0/18)	
81-90 years	0 ( <i>n</i> =0/2)	

Tables 3 and 4 respectively and relevant photographs in Figures 1-6.

The mean body mass index (BMI) of adults affected with perniosis was determined to be  $22.6 \pm 2.3 \text{ kg/m}^2$  while that of the remaining adult population without perniosis was calculated as  $24.7 \pm 2.8 \text{ kg/m}^2$ . This difference in BMI of cases versus unaffected adults was found to be highly statistically significant using *t*-test (*P*-value = 0.0000001).

BMI of children with perniosis was compared with the standard population using WHO BMI charts. The children were broadly divided into two groups, those with BMI less than  $50^{\text{th}}$  percentile and those with BMI more than  $50^{\text{th}}$  percentile as determined from the charts. These excluded two very young children from the perniosis group and 26 infants and very young children from the unaffected group whose BMI could not be determined due to feasibility issues. It was found that children with perniosis had a higher likelihood of having BMI less than the  $50^{\text{th}}$  centile as compared to those not affected by perniosis (*P*-value = 0.0000001).

Frostbite was observed in 10 cases with a prevalence of 0.83% (95% CI, 0.45–1.53%, n = 10/1200). Out of the 10 cases, 6 were males and 4 were females.

Majority of cases were seen in children under the age of 18 years (9 out of 10) and only 1 case was documented in a 50-year-old female. The correlation with age was found to be statistically significant (P-value = 0.0005).

Site of involvement were toes in all 10 cases (100%, n = 10/10). Other sites of involvement such as fingers, the tip of the nose and so on were not seen in any case.

Out of 10 cases, 9 had first degree frostbite (superficial frostbite) and all these cases were seen in children younger than 18 years while 1 case had third

Table 3: Site of involvement in perniosis cases				
Site of lesion	No. of patients	Percentage		
Feet only	84	57.5% (n=84/146)		
Hands only	20	13.7% (n=20/146)		
Both	42	28.8% (n=42/146)		
Ears (in addition to hands and feet)	2	1.4% ( <i>n</i> =2/146)		
Cheeks (in addition to hands and feet)	1	0.7% ( <i>n</i> =1/146)		

Table 4: Morphology of lesions in perniosis cases					
Type of lesion	No. of patients	Percentage			
Erythematous cyanotic swelling	120	82.2% (n=120/146)			
Vesicular/Bullous lesions	19	13.0% ( <i>n</i> =19/146)			
Ulcerative lesions	17	11.6% ( <i>n</i> =17/146)			
Targetoid lesions	10	6.8% ( <i>n</i> =10/146)			
Maculopapularlesions/plaques	5	3.4% ( <i>n</i> =5/146)			

degree frostbite (deep frostbite) seen in the 50-year-old woman [Figures 7 and 8].

The prevalence of Raynaud's phenomenon was documented as 1.5% (95% CI, 0.9–2.4%, n = 18/1200) when the history of cold digits along with biphasic/triphasic discoloration was invoked as inclusion criteria and this was taken as the measure of prevalence in accordance with international



Figure 1: Perniosis - bullous lesions (black arrow)



Figure 2: Perniosis presenting as erythematous cyanotic swelling



Figure 3: Perniosis - erythematous lesions with overlying flaccid bullae



Figure 4: Perniosis - edema and cyanotic discoloration of hands



Figure 6: Perniosis - erythema multiforme like lesions

consensus criteria.<sup>[20]</sup> Out of the 18 cases, 16 were females and 2 were males. The gender-specific prevalence was 2.68% in females (n = 16/598) and 0.33% in males (n = 2/602) with a female to male ratio of 8:1, which was found to be statistically significant (*P*-value = 0.0008).

The age of the patients ranged from 15-32 years with a mean age of  $24.2 \pm 4.8$  years. Most common age group affected was 21-30 years recording 12 cases (66.7%, n = 12/18). Around 4 cases (22.2%, n = 4/18) were in the age group of 11-20 years and 2 cases (11.1%, n = 2/18) in the 31-40 year age group.

The duration of disease ranged from a few months to 6 years with a mean disease duration of  $2.8 \pm 1.9$  years. Thus, the onset of the disease had a different pattern, occurring most commonly in the second decade of life (i.e., 11–20 years). Out of the 18 cases, 10 (55.6%, n = 10/18) had experienced their first episode during the second decade of life (11–20 years) while the remaining 8 (44.4%, n = 8/18) recollected their first episode to the third decade of life (21–30 years).

Patients reported affliction of all fingers and never as an isolated phenomenon in one or two fingers. The disorder predominantly affected the fingers in all patients [Figure 9] and additional involvement of toes was seen in 4 cases



Figure 5: Perniosis - ear involvement



Figure 7: Superficial frostbite - waxy appearance of affected toe (black arrow)

only. Extremities such as the nose, tips of ears, and so on were not involved in any case.

Raynaud's phenomenon, severe enough to cause ulceration or gangrene was not seen in any case. A link with stressors (other than cold) such as smoking, menstrual cycles, emotional stress, drugs, etc., could not be established. Symptoms related to an underlying connective tissue disorder such as photosensitivity, oral/nasal ulcers, binding down of skin, etc., were not present in any case.

Asteatotic eczema was seen in 20 patients giving a prevalence of 1.67% (95% CI, 1.08–2.56%, n = 20/1200). Out of the 20 cases, 10 (50%) were females and 10 (50%) were males.

The age of these cases ranged from 35–74 years, with a median age of 61 years. Prevalence of asteatotic eczema



Figure 8: Deep frostbite - ulcerated lesions

was found to increase with age till the age of 70 years and then showed a dip in the 71–80 years age group [Table 5].

Majority of cases were in the old age group of 65 years and above (50%, n = 10/20). The middle age group of 45–64 years contributed an additional 8 cases (40%, n = 8/20). Thus, a vast majority of cases (18 out of 20) were contributed by middle-aged or old population and only 2 cases (10%, n = 2/20) could be documented in the young population (18–45 years) but none in children aged <18 years. The association of asteatotic eczema with old age was found to be statistically significant (*P*-value = 0.0000001).

Involvement of shins was seen in all cases (100%, n = 20/20). Additional involvement of the forearms was documented in three cases (15%, n = 3/20). Other sites of involvement such as the abdomen, thighs, back, etc., were not seen in any case [Figure 10].

Cold panniculitis, cold urticaria, and livedo reticularis were each seen in one case with a prevalence of 0.08% (95% CI, 0.01–0.47%, n = 1/1200) [Figure 11].

No case of cryoglobulinemia or acrocyanosis was encountered.

#### Discussion

Perniosis was the most common cold dermatoses we came across in this study. The prevalence of perniosis in our study (12.2%) was consistent with textbook citations which ranged from 7% to 12.2%. Singh *et al.* noted a prevalence of 5.7% which was significantly lower as compared to our study.<sup>[12]</sup> However, their study was conducted in a hospital setup and milder cases of perniosis were bound to be missed as they were less likely to attend the hospital for the same. Besides, their study period included all the seasons while our study was conducted exclusively during the winter season. This explains the higher incidence of perniosis in our study. Only a few studies have been conducted on this topic in tropical countries. Al-Nuaimy *et al.* studied perniosis in Iraq and reported that perniosis



Figure 9: Raynaud's phenomenon – Cold-mediated vasoconstriction (black arrow)

Table 5: Age-specific prevalence of asteatotic eczema					
Number	Total number	Age-specific			
of cases	of individuals	prevalence			
2	184	1.1% ( <i>n</i> =2/184)			
2	122	1.6% ( <i>n</i> =2/122)			
6	90	6.7% ( <i>n</i> =6/90)			
8	50	16% ( <i>n</i> =8/50)			
2	20	10% ( <i>n</i> =2/20)			
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is a common problem in their sub-tropical country owing to the diurnal and seasonal variation in temperature, though they did not comment regarding its prevalence in the general population.<sup>[21]</sup> The Netherlands Institute for Health Services Research (NIVEL) reports the prevalence to vary between 0.9 per 1,000 and 1.7 per 1,000.<sup>[22]</sup> This was much lower than our findings as expected owing to the temperate climate in the Netherlands which does not have extremes of temperature variability.

Perniosis or chilblains are said to occur more commonly in females<sup>[12]</sup> and the same was corroborated in this study with a statistically significant difference between male and female gender (*P*-value = 0.047). This can be attributed to genetic and hormonal differences between the two groups.

More than 90% of cases had onset during childhood before the age of 18 years. It was observed that 67.1% of cases experienced their first episode during the second or third year of life but never during infancy. This pointed to the fact that people who have an abnormal sensitivity to cold possess it right from birth. Absence of perniosis during the first year of life could be attributed to the fact that the infant is largely confined indoors along with adequate padding and protective clothing to prevent the occurrence of any cold-related injuries. The fact that the child could move about independently after the first year of life correlated well with the occurrence of perniosis during the second or third year of life. This would lead to increased exposure to cold and dampening of protective clothing, conditions which are apt for the development of perniosis.

The involvement of feet was expected to be more common as feet bear the major brunt of cold during the winter



Figure 10: Asteatotic eczema - Crazy pavement appearance

season. Not only are the feet constantly exposed to snow that covers the roads of Kashmir valley in winters but also more likely to become dampened due to moisture and water, producing conducive circumstances for the development of perniosis. These findings were in concordance with those of Singh *et al.* and Al-Nuaimy *et al.*<sup>[12,21]</sup>

BMI in adults, as well as children affected with perniosis, was found to be significantly lower as compared to the rest of the population. Vasoconstriction has a protective effect as it prevents the core body temperature from falling too much and thus, has a role in survival under adverse cold conditions. It can thus be postulated that any factor which provides better protection to the core body temperature would lead to a lesser need of vasoconstriction in response to cold and thus, a lesser risk of cold-related adverse events. The presence of subcutaneous fat is a protective factor for the core body temperature and hence, patients with a higher BMI should be at a lower risk of developing perniosis. The same was validated in this study.

Frostbite was encountered in 0.83% of the population. The findings were comparable to studies conducted by Lehmuskallio *et al.* who recorded the incidence of frostbite at 1.8 per 1000 conscripts per year (0.18%) and Ervasti *et al.* who recorded the annual occurrence at 2.2%.<sup>[14,23]</sup>

Frostbite was seen most commonly in children. It can be proposed that children are at higher risk of developing frostbite as they are less likely to adhere to protective measures like adequate clothing and more likely to engage in recreational activities like playing with snow.

The prevalence of Raynaud's phenomenon (RP) shows great variation in various studies and textbook citations, ranging from a few to more than 20%.<sup>[17,18]</sup> In this study, we found the prevalence of Raynaud's phenomenon to be 1.5%. Our findings were comparable to those of Bartelink *et al.* when they used strict criteria of at least a biphasic discoloration similar to ours.<sup>[17]</sup> However, when they used cold digits with monophasic white discoloration



Figure 11: Cold panniculitis - bilaterally symmetrical erythematous indurated plaques over the cheeks

as a criterion, the prevalence increased to 5.4% in males and 7.5% in females which further jumped to 10.4% and 21.2% when monophasic white or blue discoloration was used as a criterion. In another study by Voulgari et al. in Greece, the prevalence was found to be 5.2% which was higher than that seen by us. Besides geographical and climatological dissimilarities and variations in structures of studied populations and genetic variability, differences in criteria defining RP may largely be responsible for the wide variation in cited prevalence in different studies. In addition, most of the studies for determining the prevalence of Raynaud's have been questionnaire-based and did not have any objective criteria for defining RP, resulting in large variations in the results obtained in different studies. We used the three-step diagnostic approach of international consensus criteria<sup>[20]</sup> to diagnose Raynaud's phenomenon and hence, at least a biphasic discoloration was mandatory for the inclusion of any case.

Gender-specific prevalence was noted to be 2.68% in females and 0.33% in males. Our findings closely resembled those of Bartelink *et al.* who noted the prevalence of RP as 0.5% in males and 2.9% in females.<sup>[17]</sup> The difference suggests that hormonal factors may be a contributing factor to the development of the disorder with estrogen potentiating  $\alpha$ -adrenergic mediated vasoconstriction.

Diagnosis of RP is primarily clinical but further distinction into primary and secondary RP can be made based on clinical tests and laboratory investigations. Bilaterally symmetrical involvement, absence of ulcerative or necrotic lesions, normal peripheral pulses, and early age of onset pointed towards the possibility of primary RP in the cases that we came across in our study.

The prevalence of asteatotic eczema in our study was found to be much less than that of textbook citations and previous studies.<sup>[19,24]</sup> This was expected as those studies were conducted exclusively in geriatric populations. As ours was a community-based study with the study population comprising of individuals from all ages, the prevalence was expected to be much lower. The age-related changes in the skin which predispose to xerosis are expected to become more pronounced with increasing age. This finding was validated in our study as well. The age-specific prevalence of AE increased steadily with increasing age, reaching a peak in the seventh decade of life (16%). Iqbal recorded similar findings with the majority of cases occurring in the 65–74 year age group.<sup>[19]</sup>

Cold panniculitis, cold urticaria, and livedoreticularis were recorded in one case each with a prevalence of 0.08% each. This was again in concordance with textbook citations.<sup>[24]</sup>

#### Conclusions

Cold dermatoses form an important source of morbidity among the native population of Kashmir. These can be easily prevented by ensuring adequate protection against cold. Creating awareness regarding these disorders and probable association with connective tissue disorders is also imperative.

## Limitations

This was a clinical study and no investigations were done to corroborate the diagnosis, owing to non-feasibility of the same in a community setup. Cold urticaria, cold panniculitis, and livedo reticularis are rare disorders with a very low prevalence and the sample size in our study were quite small to make any concrete inferences regarding the exact prevalence and pattern of these disorders.

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#### **Conflicts of interest**

There are no conflicts of interest.

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