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# Keratosis Pilaris: A Common Follicular Hyperkeratosis

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*Keratosis pilaris (KP) is a common inherited disorder of follicular hyperkeratosis. It is characterized by small, folliculocentric keratotic papules that may have surrounding erythema. The small papules impart a stippled appearance to the skin resembling gooseflesh. The disorder most commonly affects the extensor aspects of the upper arms, upper legs, and buttocks. Patients with KP usually are asymptomatic, with complaints limited to cosmetic appearance or mild pruritus. When diagnosing KP, the clinician should be aware that a number of diseases are associated with KP such as keratosis pilaris atrophicans, erythromelanosus follicularis faciei et colli, and ichthyosis vulgaris. Treatment options vary, focusing on avoiding skin dryness, using emollients, and adding keratolytic agents or topical steroids when necessary.*

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**K**eratosis pilaris (KP) is a hyperkeratotic disorder that manifests as grouped folliculocentric keratotic papules, with a variable degree of perifollicular erythema. This benign condition is marked by a characteristic distribution over the extensor aspects of the arms and thighs and on the buttocks.<sup>1</sup>

## Epidemiology

Keratosis pilaris is a common disorder that often is an incidental finding on physical examination. Many patients with KP are unaware they are affected. A 1985 survey noted a prevalence of 44% in

155 otherwise unaffected patients.<sup>2</sup> In the adolescent population, its prevalence is postulated to be at least 50%; it is more common in adolescent females than males, seen in up to 80% of adolescent females.<sup>3</sup> The disorder is inherited in an autosomal dominant fashion with variable penetrance; no specific gene has been identified. In a study of 49 evaluated patients, there was a positive family history of KP in 19 patients (39%), while 27 patients (55%) had no family history of the disorder.<sup>4</sup>

## Clinical Features

The keratotic follicular papules of KP most commonly are grouped on the extensor aspects of the upper arms (Figure), upper legs, and buttocks.<sup>4</sup> Other affected locations may include the face and the trunk.<sup>5</sup> The small papules impart a stippled appearance to the skin resembling gooseflesh. This gooseflesh is differentiated from cutis anserina, a piloerector response to sympathetic stimuli. Individual papules are acuminate; approximately 1 mm in size; and often contain a fine-coiled, brittle hair.

Keratosis pilaris has varying degrees of perifollicular erythema determined by the extent of inflammation. When the surrounding erythema is marked, the term *keratosis pilaris rubra* may be used.<sup>5</sup> This erythematous component also can be observed without the follicular plugging. The papules of KP also can be grayish white without erythema and have been termed *keratosis pilaris alba*.<sup>4</sup> Patients with KP generally are asymptomatic but occasionally may be pruritic. Other common complaints include poor cosmetic appearance and persistent rough-textured skin, which may cause psychological distress for the patient. The severity of KP had seasonal variability in a 1994 survey evaluating 49 patients, as approximately half had improvement in the summer and approximately half had exacerbation in the winter.<sup>4</sup> The survey also determined that the condition generally appears in childhood, with age at onset

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being the first decade for half of patients. The condition tends to improve with age or remain unchanged; few patients experience worsening with age.<sup>4</sup> Although the causes of KP have yet to be identified, a hormonal influence may be involved, considering the high prevalence and intensity during adolescence.<sup>6</sup> In a study of 78 hirsute, obese, premenopausal women, increased incidence of KP was associated with hyperandrogenism.<sup>7</sup> In a report of 5 patients, the onset or severity of KP was related to hormonal changes of pregnancy.<sup>8</sup>

### Histopathology

The papules of KP consist of excess keratin in the follicular orifices, creating horny plugs that dilate the follicle's infundibulum. A superficial, mild, perivascular lymphohistiocytic infiltrate is noted in the upper dermis and in the perifollicular areas.<sup>3,9</sup> The epidermis demonstrates mild hyperkeratosis, hypogranulosis, and follicular plugging. The stratum corneum may exhibit focal parakeratosis, but the parakeratotic cells are not retained within the follicle. The keratotic plug extends deep into the hair follicle and may result in atrophy of the follicular walls, sebaceous glands, and arrectores pilorum. Specifically, the plug consists of horny lamellae and often entraps 1 or more coiled brittle hairs.<sup>10,11</sup>

### Associations

Keratosis pilaris is observed in association with a variety of conditions. A correlation with atopic dermatitis has been described<sup>12</sup>; however, studies have concluded that KP has no diagnostic significance for atopic dermatitis.<sup>2,13</sup> In fact, KP occurs considerably more frequently in patients with ichthyosis vulgaris without eczema than in patients with atopic dermatitis.<sup>2,13</sup> Other conditions associated with KP include keratosis pilaris atrophicans, erythromelanosus follicularis faciei et colli, and ichthyosis vulgaris.

*Keratosis pilaris atrophicans* is a term used to describe a set of related disorders that are characterized by KP followed by atrophy. Ulerythema ophryogenes is an example and is characterized by inflammatory keratotic facial papules in children that result in scarring and alopecia. The conditions differ based on the severity of inflammation and the distribution of affected areas, usually involving the face and scalp. Keratosis pilaris of the extremities is an associated finding.<sup>14-16</sup> Although these disorders within the keratosis pilaris atrophicans category have considerable overlap, studies have shown that they have substantial clinical and genetic variation.<sup>14,17</sup>

Erythromelanosus follicularis faciei et colli is characterized by erythema, hyperpigmentation,



Keratosis pilaris of the upper arm in a 25-year-old woman.

and follicular papules. These papules are grouped within well-demarcated, reddish brown areas that impart a granular texture to the skin. This rare disease also may be seen with KP of the upper extremities and trunk.<sup>18,19</sup>

Ichthyosis vulgaris is a disorder of abnormal keratinization that is inherited in an autosomal dominant fashion. Rarely, an acquired form of the disease occurs in association with systemic disease.<sup>20</sup> The skin appears dry and scaly. The affected areas generally are the extensor aspects of the upper and lower extremities with sparing of the flexural creases. Hereditary ichthyosis vulgaris frequently is associated with KP.<sup>2,21,22</sup> Other conditions that have been reported to correlate with KP are vitamin B<sub>12</sub> and vitamin A deficiencies,<sup>23,24</sup> hypothyroidism, Cushing disease, and corticotropin administration.<sup>9,25</sup>

### Differential Diagnosis

Various disorders may appear similar to KP, such as lichen spinulosus, pityriasis rubra pilaris, and phrynoderma. In lichen spinulosus, which is generally limited to the pediatric population, small follicular papules with keratotic spines group into large patches. These papules symmetrically affect the trunk and extremities and tend to remit spontaneously.<sup>26</sup>

Pityriasis rubra pilaris is characterized by follicular keratoses, erythroderma, and palmoplantar

keratoderma. The eruption generally begins with scaly macules on the head, neck, or upper trunk. In the subsequent weeks, numerous macules associated with erythematous perifollicular papules appear and proceed to affect the extremities.<sup>27</sup> Similarly, phrynoderma also consists of abnormal follicular hyperkeratosis, but the papules first appear on the extensor surfaces of the extremities, shoulders, and buttocks. Although phrynoderma may be caused by vitamin A deficiency, multiple etiologies, such as other nutrient deficiencies and general malnutrition, also may be involved.<sup>28</sup> Other causes of follicular keratoses include Darier disease and Kyrle disease; however, their clinical presentations usually are remarkably different from KP.

### Treatment

Keratosis pilaris often improves with increasing age (mean age of improvement, 16 years), though a few patients may have worsening symptoms with time.<sup>4</sup> Although no absolute cure is available, measures can be taken to decrease symptoms. Patients should prevent excessive skin dryness by using mild soaps, avoiding long hot baths, and optimizing home humidity.<sup>29</sup>

Although an emollient cream can alleviate mild KP, more extensive involvement may require a keratolytic agent such as lactic acid, salicylic acid, and urea cream. A preparation of salicylic acid 2% in urea cream 20% is an effective combination.<sup>30,31</sup> Topical tretinoin therapy also may be administered, especially when other treatments have been inadequate. However, retinoids have varying degrees of success.<sup>32,33</sup> Notably, calcipotriol ointment has been shown to be ineffective in KP.<sup>34</sup>

If KP lesions are marked by substantial inflammation, mild topical steroids may be beneficial. Preparations such as triamcinolone acetonide 0.1% or desonide 0.05% creams can be applied until inflammation improves, usually within 7 days. The patient should then discontinue steroids and manage KP with skin-softening therapies.<sup>30</sup>

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