Keratosis Pilaris

ABSTRACT

Keratosis pilaris (KP) is a common, benign skin condition that primarily affects children and adolescents. It is characterized by small follicular papules resembling "chicken skin" or goosebumps, most often on extensor surfaces of the proximal upper and lower extremities. The exact cause of KP is unclear, however it is believed to follow an autosomal dominant inheritance pattern and may be associated with a mutation in filaggrin, a protein involved in maintaining the skin barrier. This ultimately results in excessive keratin production and deposition surrounding hair follicles. Diagnosis is typically based on the patient's history and clinical presentation. KP is most often asymptomatic and tends to improve with age. However, treatment is aimed at enhancing cosmetic appearance or relieving mild pruritus. Management includes optimizing skin moisturization and using topical keratolytic agents, though other options such as topical retinoids and laser therapy may also be effective.

KEYWORDS: Keratosis pilaris, keratolytic agents, asymptomatic, benign

Introduction

Keratosis pilaris (KP) is a common and benign dermatological condition considered a normal variant of skin. It typically presents as numerous small papules resembling "chicken skin" or goosebumps, primarily on the extensor surfaces of the proximal extremities. There are several variants of KP, including *keratosis pilaris rubra*, characterized by marked erythema surrounding hair follicles, *keratosis pilaris alba*, featuring grayish-white lesions without erythema, and *keratosis pilaris atrophicans faciei* (also known as *ulerythema ophryogenes*), which primarily affects the lateral eyebrows and cheeks and is often associated with scarring or alopecia.[1,2] While there is no definitive cure or means for prevention, KP typically improves spontaneously with age and may become less noticeable with moisturization.[3,4] It typically presents in children and adolescents, with most patients seeking treatment for cosmetic concerns.[4] It is most often asymptomatic, however some patients report having mild pruritus. Management aims to relieve symptoms and improve the appearance of lesions.

Bianca Te, BSc, *Faculty of Medicine, University of British Columbia, Vancouver, BC, Canada*
Joseph Lam, MD FRCPC, *Department of Pediatrics, Department of Dermatology and Skin Sciences, University of British Columbia, Vancouver, British Columbia, Canada*

Epidemiology

KP affects approximately 50% to 80% of adolescents and 40% of adults, typically appearing during the first decade of life and improving with age.[4] The condition affects males and females at comparable rates, with some studies suggesting a slightly higher prevalence in females.[4] People of all ethnicities are affected and studies have not found any significant association with ethnicity.[5] KP tends to worsen in the winter when the weather becomes drier.[4]

Atopic conditions such as asthma or atopic dermatitis, ichthyosis vulgaris, and xerosis are associated with KP.[4,5,6] It has also been linked to other conditions such as obesity, type 1 diabetes mellitus, Down Syndrome, Noonan syndrome, cardio-facio-cutaneous syndrome, and ectodermal dysplasias.[4,5,7,8,9]

Pathophysiology

The exact pathogenesis of KP remains unclear. It is believed to have an autosomal dominant inheritance pattern with variable penetrance.[3] While no specific gene has been definitively linked to KP, research suggests that many affected individuals carry mutations in filaggrin, an important protein involved in maintaining the skin barrier.[2,10] Other studies suggest an association with mutations in the Ras signaling cascade and *ABCA12* gene.[11]

KP arises from excessive keratin production, which accumulates around hair follicles and traps the hair beneath the skin's surface.[11] This creates the characteristic appearance of small raised bumps on the skin with varying degrees of surrounding redness. Some studies attribute this to a lack of sebaceous glands, which disrupts hair shaft and epithelial barrier function.[10] Others propose that a defective hair shaft ruptures the follicular epithelium, triggering inflammation and abnormal follicular keratinization.[12]

Differential diagnosis

- Acne vulgaris
- Atopic dermatitis
- Darier disease
- Eruptive vellus hair cysts
- Follicular lichen planus
- Folliculitis
- Food allergy
- Ichthyosis follicularis
- Kyrle disease
- Lichen nitidus
- Lichen spinulosus
- Medications side effects (e.g., from cyclosporine, the B-Raf inhibitors such as dabrafenib and vemurafenib, and tyrosine kinase inhibitors such as nilotinib, dasatinib, ponatinib, erlotinib, and sorafenib)[9]
- Milia
- Phrynoderma due to nutritional deficiencies
- Pityriasis rubra pilaris
- Rosacea

History

To accurately diagnose KP, it is important to begin with a comprehensive history focusing on the onset, appearance, location, and associated symptoms.[3] Most patients seek care due to cosmetic concerns, specifically the characteristic "chicken skin" or "goosebump" appearance and sandpaper-like texture.[3,13] Patients are often otherwise healthy as KP is typically asymptomatic. However, mild pruritus or erythema can be reported.[3]

Most patients will report a family history of KP as there is a genetic component to this condition.[4] Many patients will endorse a history of atopic dermatitis or dry skin.[5] It is also important to ask about associated comorbidities such as type 1 diabetes mellitus, and associated syndromes such as Down syndrome, Noonan syndrome, and cardio-facio-cutaneous syndrome.[9]

A thorough review of medications should be completed, as certain drugs, including cyclosporine, B-Raf inhibitors like dabrafenib, and tyrosine kinase inhibitors such as nilotinib, have been associated with KP-like lesions.[9] Finally, understanding previously attempted treatments for KP is crucial in developing an effective management plan.[3]

Physical examination

Physical findings of KP are most commonly bilateral and symmetric on the cheeks (Figure 1a and b) and the extensor surfaces of the upper arms (Figure 2), thighs (Figure 3), and buttocks. Less commonly, the condition may also involve the trunk, and distal extremities.[3] Affected areas are characterized by numerous small, keratotic papules surrounding hair follicles. These papules may

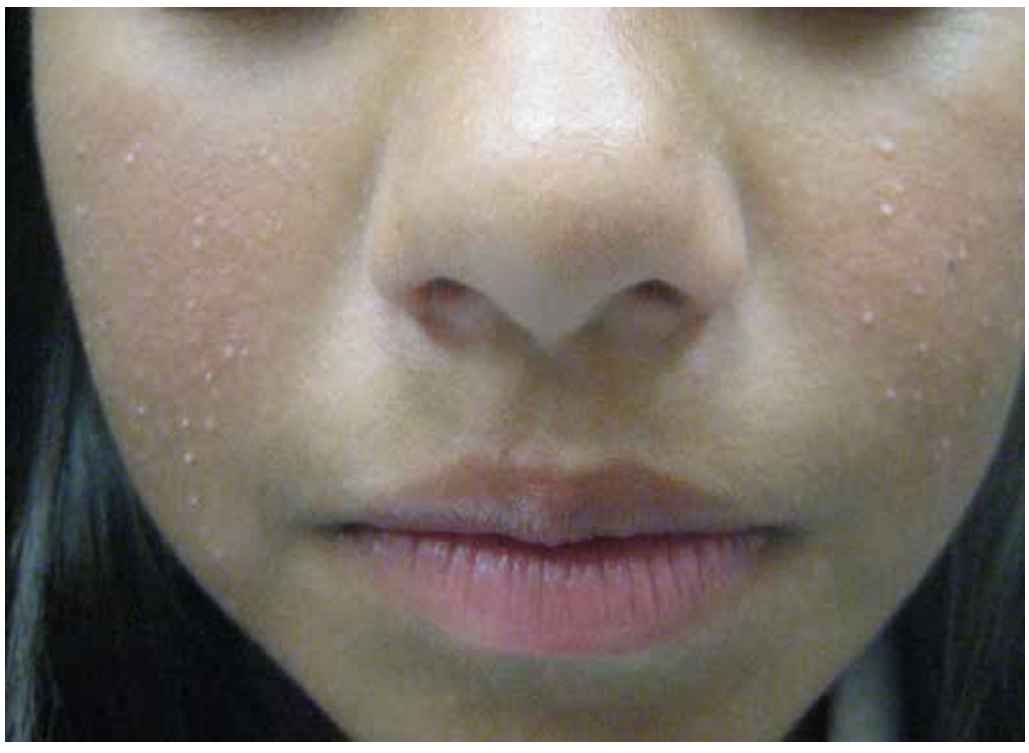

Figure 1a: Keratotic follicular papules with underlying erythema over the cheeks of a 13-year-old female

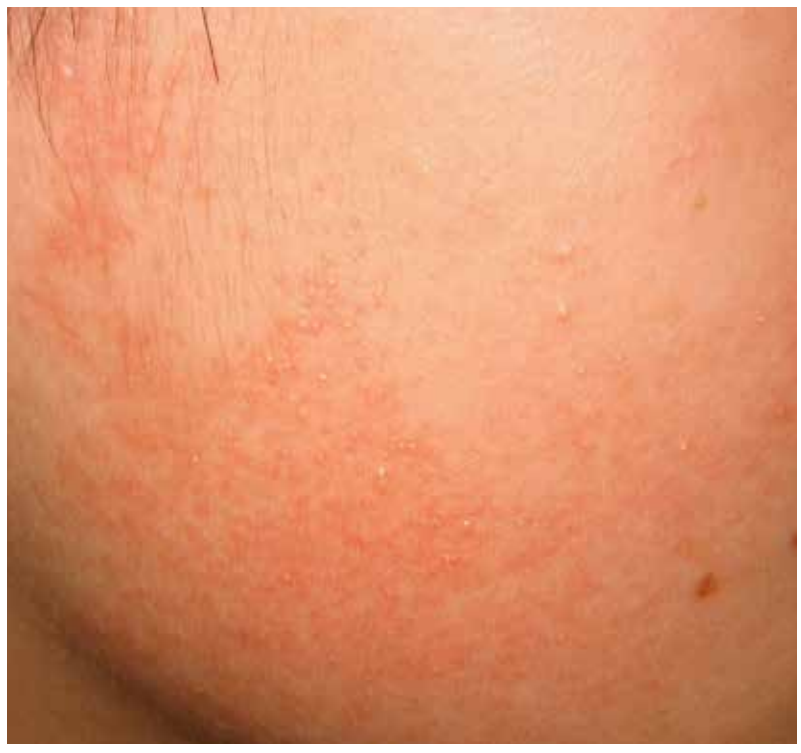

Figure 1b: Keratotic follicular papules with underlying erythema over the cheeks of a 14-year-old female

be skin-colored, erythematous, or surrounded by a perifollicular erythematous halo. Thin, coiled hairs are sometimes visible beneath the surface. The surrounding skin may be dry and scaly, as KP is often associated with xerosis. On palpation, the affected areas feel rough with sandpaper-like texture.[3]

It is important to assess for associated features related to the several variants of KP. For example, marked erythema may suggest a diagnosis of *keratosis pilaris rubra*, grayish-white lesions may suggest *keratosis pilaris alba*, and primary involvement of the lateral eyebrows and cheeks with scarring or alopecia may suggest *keratosis pilaris atrophicans faciei* (or *ulerythema ophryogenes*) (Figure 4).

Lastly, evaluate for dysmorphic features that might suggest an underlying syndrome associated with KP. For example, upslanting palpebral fissures, epicanthic folds, and brachycephaly are commonly seen in patients with Down syndrome, while a broad based nose, widely spaced eyes, and low-set ears are commonly seen in patients with Noonan syndrome.[9]

Investigations

The diagnosis of KP is most often made clinically, however dermoscopy and biopsy can be used to

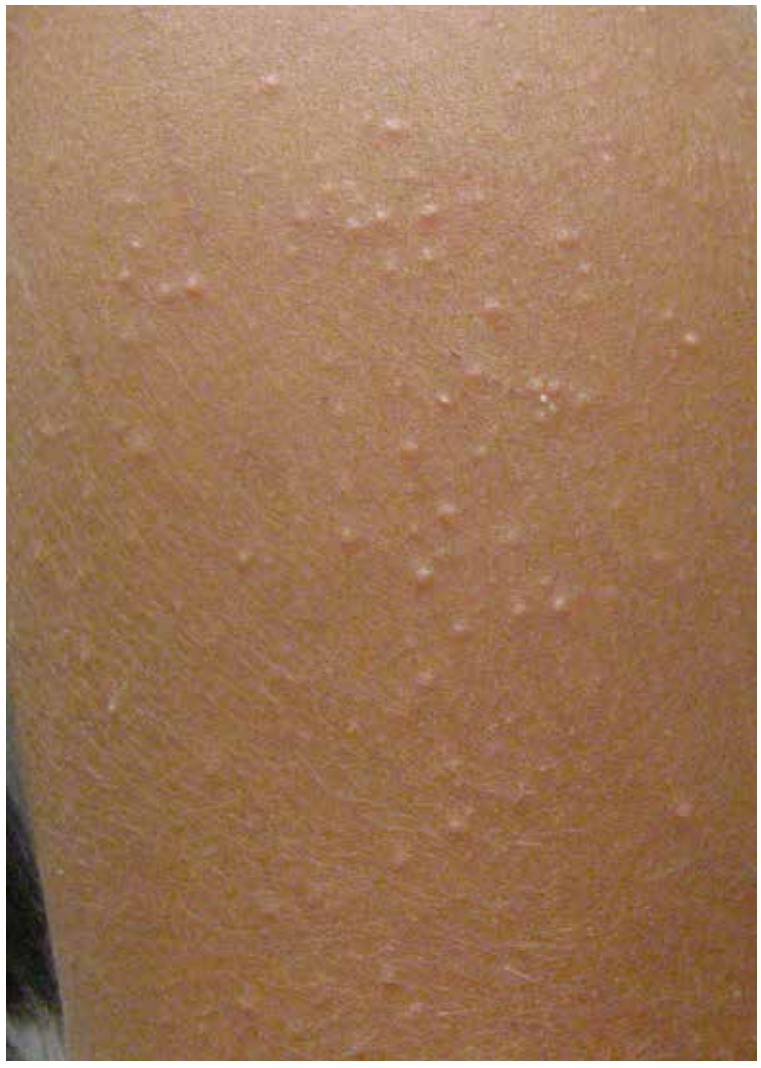

Figure 2: Keratotic follicular papules over the upper outer arms of a 13-year-old female

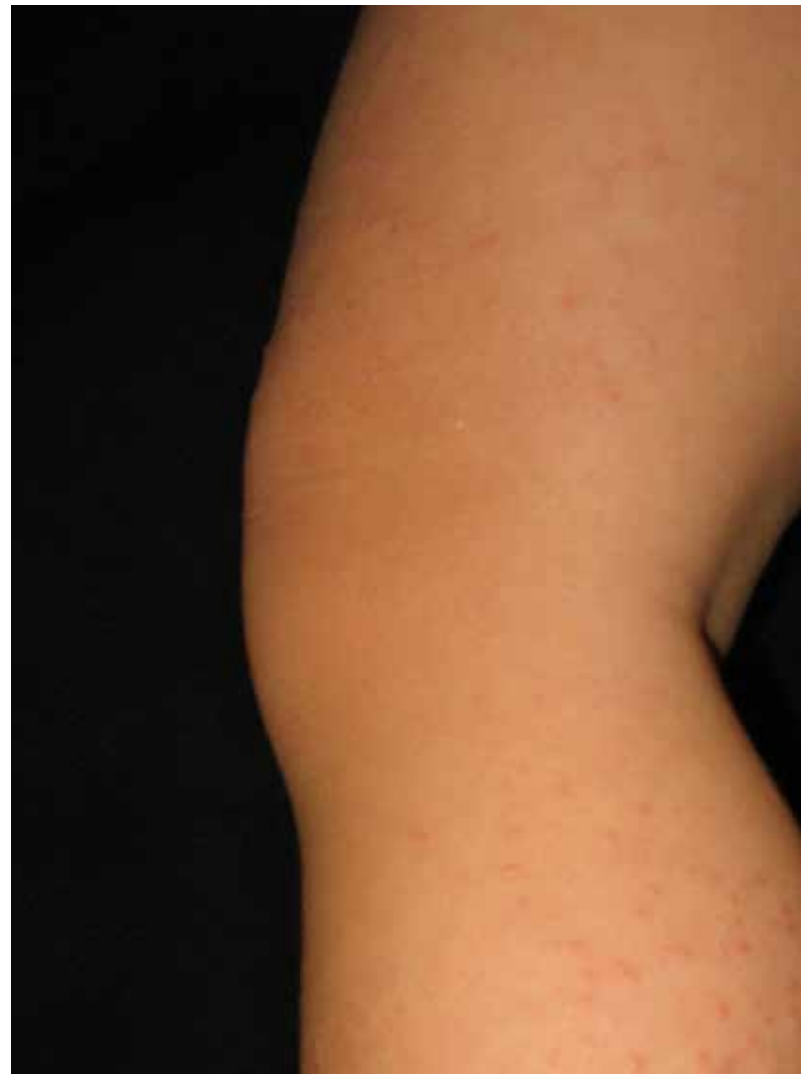

Figure 3: Keratotic follicular papules over the upper and lower legs of a 18-month-old toddler

evaluate lesions in challenging or atypical cases.[9]

Dermoscopy of KP reveals abnormally thin, short, and coiled hair shaft structure. Varying degrees of scaling, perifollicular erythema, and hyperpigmentation may also be observed.[12,14] Dermoscopy can be helpful in differentiating KP from other differential diagnoses. For example, follicular lichen planus would demonstrate keratotic plugs without the coiled or broken hairs observed in KP.[14]

Biopsies are rarely required for the diagnosis of KP. Histology of KP lesions demonstrate hyperkeratosis, hypergranulosis, marked plugging of hair follicles, absence of sebaceous glands, and mild lymphocytic infiltration.[10]

Management

KP is generally not treated as it is benign and often resolves with age.[4] There have been no controlled clinical trials that suggest a cure or means for prevention. Treatment aims to manage symptoms and improve cosmetic appearance.[3]

Skin care can be optimized to prevent irritation and excessive dryness by using mild hypoallergenic soaps, avoiding hot baths, enhancing home humidity, and refraining from manipulating and rupturing the lesions.[2,3] Emollients and moisturizers can be used to improve skin moisturization, however they will not likely resolve the bumpy appearance.[13]

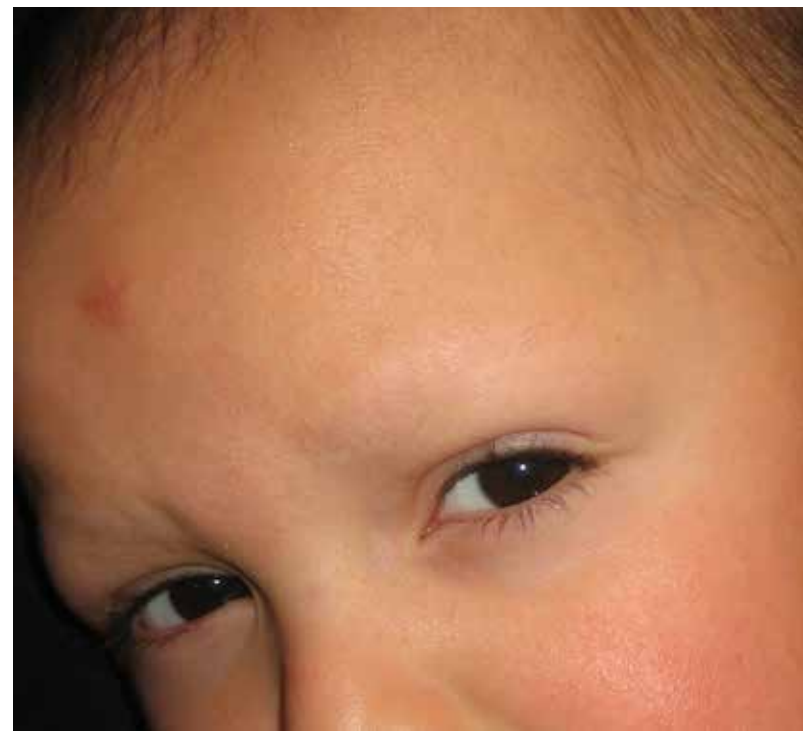

Figure 4: Erythema and follicular papules with alopecia of the eyebrows in a 2-year-old male

First-line treatment includes topical keratolytic agents that exfoliate the skin and reduce keratin build up. Examples include creams containing lactic acid, salicylic acid, glycolytic acid, or urea. Prescription retinoids such as tazarotene 0.01% can also be used.[2,9]

Associated redness and inflammation can be managed with intermittent mild-to-moderate topical steroids such as triamcinolone acetonide 0.1% or desonide 0.05% creams.[2] Non-steroidal ointments such as tacrolimus 0.03% or 0.1% ointment can also be effective.[13]

Laser therapy is another treatment option for KP. The Q-switched Nd:YAG laser has been the most commonly used as it can effectively penetrate to deeper dermal hair follicles. Studies also suggest efficacy in pulsed dye laser, alexandrite laser, and fractional carbon dioxide laser treatments.[9,15]

SUMMARY OF KEY POINTS

- Keratosis pilaris presents as numerous small follicular papules resembling "chicken skin" or goosebumps, most often on extensor surfaces of the proximal upper and lower extremities.
- Keratosis pilaris is a benign skin condition that often improves with age.
- Management of keratosis pilaris is aimed at reducing symptoms such as pruritus and improving cosmetic appearance. Keratolytic agents such as lactic acid or salicylic acid creams are most commonly used for treatment.

Conclusion

Keratosis pilaris is a common, benign skin condition that primarily affects children and adolescents. It presents as numerous small, keratotic papules surrounding hair follicles, most often on extensor surfaces of the proximal extremities. It is most often asymptomatic but can become mildly pruritic or inflamed. There is no definitive cure for KP, therefore treatment aims to manage these symptoms and improve cosmetic appearance.

References

1. Arnold, A. W., & Buechner, S. A. (2006). Keratosis pilaris und Keratosis pilaris atrophicans faciei [Keratosis pilaris and keratosis pilaris atrophicans faciei]. Journal der Deutschen Dermatologischen Gesellschaft
2. Journal of the German Society of Dermatology : JDDG, 4(4), 319–323. https://doi.org/10.1111/j.1610- 0387.2006.05933.x
3. Hwang, S., & Schwartz, R. A. (2008). Keratosis pilaris: a common follicular hyperkeratosis. Cutis, 82(3), 177–180.
4. Pennycook KB, McCready TA. Keratosis Pilaris. [Updated 2023 Jun 26]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: https://www.ncbi.nlm.nih.gov/sites/books/NBK546708/
5. Poskitt, L., & Wilkinson, J. D. (1994). Natural history of keratosis pilaris. The British journal of dermatology, 130(6), 711–713. https://doi.org/10.1111/j.1365-2133.1994.tb03406.x

CLINICAL PEARLS

Keratosis pilaris is the result of excess keratin production that deposits around the hair follicle, however the exact pathogenesis is not fully understood.

Diagnosis of keratosis pilaris is made on history and physical exam. Dermoscopy and biopsy are not often required, but can be used if other conditions are suspected.

Patients should understand that keratosis pilaris is benign and that there is no cure. It is important to provide reassurance that the condition often improves with age, however some topical treatments may be able to help improve cosmetic appearance.

6. Yosipovitch, G., Mevorah, B., Mashiach, J., Chan, Y. H., & David, M. (2000). High body mass index, dry scaly leg skin and atopic conditions are highly associated with keratosis pilaris. Dermatology (Basel, Switzerland), 201(1), 34–36. https://doi.org/10.1159/000018425
7. Mevorah, B., Marazzi, A., & Frenk, E. (1985). The preva- lence of accentuated palmoplantar markings and keratosis pilaris in atopic dermatitis, autosomal dominant ichthyosis and control dermatological patients. The British journal of dermatology, 112(6), 679–685. https://doi.org/10.1111/j.1365-2133.1985.tb02336.x
8. Boza, J. C., Trindade, E. N., Peruzzo, J., Sachett, L., Rech, L., & Cestari, T. F. (2012). Skin manifestations of obe- sity: a comparative study. Journal of the European Academy of Dermatology and Venereology : JEADV, 26(10), 1220–1223. https://doi.org/10.1111/j.1468- 3083.2011.04265.x
9. Firsowicz, M., Boyd, M., & Jacks, S. K. (2020). Follicular occlusion disorders in Down syndrome patients. Pedi- atric Dermatology, 37(1), 219-221.
10. Wang, J. F., & Orlow, S. J. (2018). Keratosis Pilaris and its Subtypes: Associations, New Molecular and Pharmacologic Etiologies, and Therapeutic Options. American journal of clinical dermatology, 19(5), 733–757. https://doi.org/10.1007/s40257-018-0368-3
11. Gruber, R., Sugarman, J. L., Crumrine, D., Hupe, M., Mauro, T. M., Mauldin, E. A., Thyssen, J. P., Brandner, J. M., Hen- nies, H. C., Schmuth, M., & Elias, P. M. (2015). Sebaceous gland, hair shaft, and epidermal barrier abnormalities in keratosis pilaris with and without filaggrin deficiency. The American journal of pathology, 185(4), 1012–1021. https://doi.org/10.1016/j.ajpath.2014.12.012
12. Kodali, N., Patel, V. M., & Schwartz, R. A. (2023). Keratosis pilaris: an update and approach to management. Italian journal of dermatology and venereology, 158(3), 217–223. https://doi.org/10.23736/S2784-8671.23.07594-1
13. Thomas, M., & Khopkar, U. S. (2012). Keratosis pilaris revisited: is it more than just a follicular keratosis?. International journal of trichology, 4(4), 255–258. https://doi.org/10.4103/0974-7753.111215
14. Lam, J. (2018). Keratosis pilaris. BCCH Pediatric Dermatology Clinic. http://www.bcchildrens.ca/dermatology-site/Documents/bc-childrens-hospital- dermatology-keratosis-pilaris.pdf
15. Panchaprateep, R., Tanus, A., & Tosti, A. (2015). Clinical, dermoscopic, and histopathologic features of body hair disorders. Journal of the American Academy of Dermatology, 72(5), 890–900. https://doi.org/10.1016/j.jaad.2015.01.024
16. Maghfour, J., Ly, S., Haidari, W., Taylor, S. L., & Feldman, S. R. (2020). Treatment of keratosis pilaris and its variants: a systematic review. Journal of Dermatological Treatment, 33(3), 1231–1242. https://doi.org/10.1080/09546634.2020.1818678

Keratosis Pilaris Key Points for the Practitioner

History (key points to cover)

- Onset, appearance, location, and associated symptoms
- Family history of keratosis pilaris
- Personal history of atopy
- Medication history (certain drugs, including cyclosporine, B-Raf inhibitors like dabrafenib, and tyrosine kinase inhibitors such as nilotinib, have been associated with keratosis pilaris-like lesions)
- Skin care (it is important to optimize skin moisturization by using mild hypoallergenic soaps, avoiding hot baths, enhancing home humidity, and refraining from manipulating and rupturing the lesions)
- Associated syndromes such as type 1 diabetes mellitus, Down Syndrome, Noonan syndrome, cardio-facio-cutaneous syndrome, and ectodermal dysplasias.

Physical Examination (key things to note)

- Most commonly affected areas are the extensor surfaces of the extremities and the buttocks, however there may also be involvement of the face, trunk, and distal extremities.
- Numerous small, keratotic papules surrounding hair follicles, with coiled hairs sometimes visible beneath the surface.
- Palpation reveals a rough, sandpaper-like texture.
- Examine for presence of erythema, dryness, or scaling.
- Examine for primary involvement of the eyebrows and cheeks, and for scarring or alopecia in these areas that might suggest keratosis pilaris *atrophicans faciei* (or *ulerythema ophryogenes*).
- Examine for dysmorphic features that might suggest an underlying syndrome associated with keratosis pilaris, such as Down syndrome or Noonan syndrome.

Information for the Patient and Family

What is keratosis pilaris?

Keratosis pilaris is a common and harmless skin condition that primarily affects children and adolescents. It typically appears as numerous small, rough bumps, often described as having a sandpaper-like texture or resembling “chicken skin” or goosebumps. These bumps most commonly develop on the outer upper arms, thighs, or buttocks.

What causes keratosis pilaris?

The exact cause of keratosis pilaris is unclear, however it is believed to be the result of an overproduction of keratin, which is a protein in the outer layer of our skin. The excess keratin plugs up the hair follicles in affected areas which causes the “chicken skin” or goosebump appearance and sandpaper-like texture.

Is keratosis pilaris contagious?

No, keratosis pilaris is not contagious. It is a harmless condition and cannot be spread from person to person.

How is keratosis pilaris treated?

Treatment aims to manage symptoms like itchiness and reduce the appearance of bumps. Skin care can be optimized by using mild soaps, avoiding hot baths, enhancing home humidity, and refraining from scratching the bumps. Emollients and moisturizers can be used to improve skin moisturization. First-line treatment includes topical keratolytic agents such as creams containing lactic acid, salicylic acid, glycolytic acid, or urea, which help to reduce the build up of keratin. Other treatment options include topical retinoids such as tazarotene 0.01%, intermittent mild topical steroids, topical tacrolimus, and various forms of laser therapy.

Can keratosis pilaris be prevented?

No, keratosis pilaris cannot be prevented. No studies have shown a potential cure or means for prevention. There appears to be a genetic component to developing keratosis pilaris as many affected people have a family history of keratosis pilaris.

Does keratosis pilaris cause any long-term effects?

Keratosis pilaris is a harmless condition that usually resolves spontaneously with age. There are no long-term effects of the condition itself, however scratching and irritating the bumps may worsen inflammation and cause scarring or hyperpigmentation.