

Table 1. Keratin tissue distribution and associated human disorders^a

Keratin pair Type II	Type I	Tissue expression	Disorders resulting from mutations
K1	K10	Suprabasal cells of cornified stratified squamous epithelia	Bullous congenital ichthyosiform erythroderma (BCIE) or epidermolytic hyperkeratosis (EHK) Epidermolytic palmoplantar keratoderma (EPPK) Non-epidermolytic palmoplantar keratoderma (NEPPK) Ichthyosis hystrix of Curth-Macklin (IHCM) Ichthyosis bullosa of Siemens (IBS) ?
K2e ^b		Upper suprabasal epidermis	?
K2p		Upper suprabasal cells in oral palate	?
K3	K12	Corneal epithelium	Meesman corneal dystrophy (MECD)
K4	K13	Suprabasal cells of orogenital stratified squamous epithelium	White sponge naevus (WSN)
K5	K14	Basal layer keratinocytes; basal cells in glands and stratified epithelia	Epidermolysis bullosa simplex (EBS): Dowling–Meara EBS; Weber–Cockayne EBS; Köbner EBS ?
K6a	K15 K16	Basal-layer keratinocytes Suprabasal cells in orogenital stratified squamous epithelium; palmoplantar epidermis; hair follicle outer root sheath; induced in interfollicular epidermis by trauma	Pachyonychia congenita type 1 (PC-1) Steatocystoma multiplex
K6b	K17	Suprabasal cells in orogenital stratified squamous epithelium Focal in palmoplantar epidermis; deep hair follicle; extensive in foetal stratified squamous epithelia; induced by trauma in interfollicular epidermis	EPPK PC-2 Steatocystoma multiplex
K6hf		Companion layer of the hair follicle	?
K7		Gland ducts; myoepithelia; many simple epithelia; hair follicle and nail bed	?
K8	K18	Simple epithelia, including early embryo; placenta	Cryptogenic liver disease ? Inflammatory bowel disorder (IBD)
	K9	Palmoplantar epidermis	EPPK
	K19	Simple epithelia; hair follicle bulge cells; some basal cells in orogenital epithelia	?
	K20	Gastrointestinal epithelia; Merkel cells; uroepithelium	?
K6irs1, K6irs2	IRSa1–IRSa3	Inner root sheath of the hair follicle	?
Hb1–Hb6	Ha1–Ha9	Hair keratins; hard keratin sites (hair shaft, nails, tongue filiform papillae)	Monilethrix

^aReferences in [50] (see also <http://www.interfil.org>).

^bSingle keratins listed denotes that this is an additional type I or type II keratin to the predominant keratin pair.

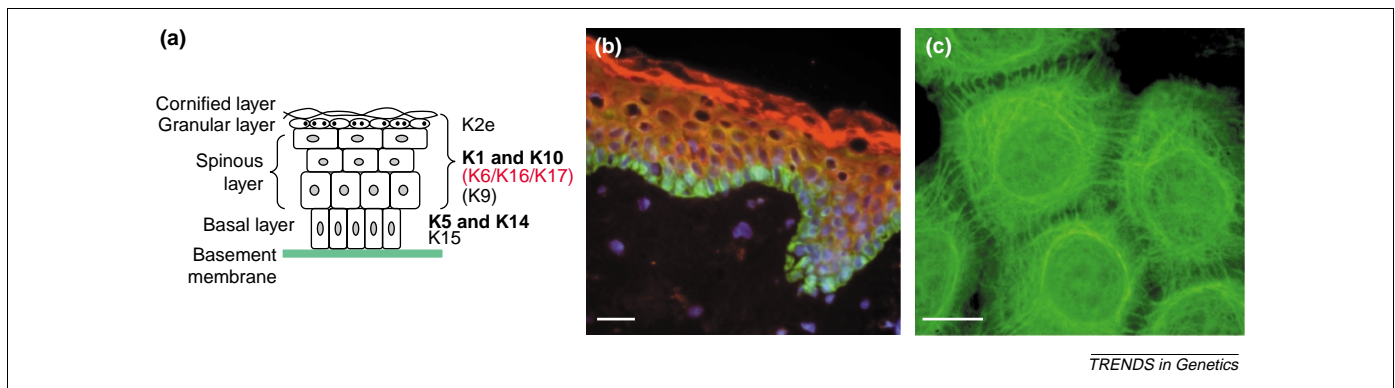


Fig. 1. Keratin expression in the epidermis. **(a)** The stratified epithelium of the epidermis. The basal layer of keratinocytes in contact with the basement membrane are actively dividing and express keratin K5 (type II) and K14 (type I) as the predominant keratin pair. Another type I keratin, K15, is also expressed in most basal cells. When the basal cells become committed to terminal differentiation they stop dividing and move into the suprabasal compartment. They then express K1 (type II) and K10 (type I) as the predominant keratin pair. These cells are also known as the spinous layer owing to the typically stellate appearance of the massive keratin bundles in these cells when visualized by light microscopy. As they differentiate further, the keratinocytes flatten out to form the granular layer, characterized by keratohyalin granules. The upper spinous layers and granular layer express an additional type II keratin, K2e. Finally, the keratinocyte becomes cornified – crosslinking of membrane proteins, such as loricrin, with the keratins to form an envelope – the nucleus breaks down and the cells die forming the cornified layer – a watertight, impermeable barrier. In wounded epidermis, the suprabasal layers switch-on the expression of K6, K16 and K17 in preference to K1 and K10. K6, K16 and K17 are normally confined to the appendages (hair follicle and nail bed) of the skin and to the thicker palmoplantar epidermis (soles and palms). K9 is unique to the palmoplantar epidermis. **(b)** Differentiation-specific expression of keratins as shown by immunofluorescence of the epidermis, with antibodies to K5 staining the basal cells (green), and antibodies to K10 staining the suprabasal cells (red). Scale bar: 20 μ m. **(c)** Keratin intermediate filaments of oral keratinocytes (TR146 cells) in culture, stained with a keratin-specific antibody. Scale bar 10 μ m. Another version of this micrograph originally appeared in Ref. [72].