

Epidermis

Molecule or structure of Importance	Location	Function	Associated Disease States
Keratin 5	Basal Layer	Attachment of basal keratinocyte to basement membrane by attachment to hemidesmosome.	Epidermolysis Bullosa Simplex
Keratin 14	Basal Layer	Attachment of basal keratinocyte to basement membrane by attachment to hemidesmosome.	Epidermolysis Bullosa Simplex
ornithine decarboxylase	Basal Layer	Marker of proliferative activity	Increased with UV exposure and other disease states
Tyrosinase	Basal melanocytes	Responsible for the first step of melanin synthesis (tyrosine to dopa)	Oculocutaneous albinism type I
Keratin 1	Spinous layer	Cytoskeletal component	bullous congenital ichthyosiform erythroderma affecting palms and soles, ichthyosis hystrix (Curth-Macklin)
Keratin 10	Spinous layer	Cytoskeletal component	bullous congenital ichthyosiform erythroderma not affecting palms and soles, ichthyosis en confetti
Desmosomes	Spinous layer	Moderate slow, strong intercellular adhesions. Composed of desmoglein, desmocollin, plakoglobin, plakophilin and desmoplakin	Naxos syndrome (plakoglobin), ectodermal dysplasia with skin fragility (plakophilin), pemphigus vulgaris (desmoglein 1,3) and various others
Adherens Junctions	Spinous layer	Moderate quick, weak cell-cell adhesions. Composed of E and P cadherins as well as alpha and beta catenin. Calcium dependent	
Lamellar Granules	Spinous and Granular layers	Secretory lysosomes which release lipid precursors (cholesterol, ceramide, free fatty acids) and hydrolytic enzymes such as steroid sulfatase.	Flegel's disease (decreased), Harlequin ichthyosis (absent), X linked ichthyosis (absent steroid sulfatase)
Keratin 2	Granular layer	Cytoskeletal component	Ichthyosis Bullosa of Siemens (keratin 2e)
Keratin 11	Granular layer	Cytoskeletal component	None
Filaggrin	Granular layer	Part of the cornified cell envelope. Catabolized to urocanic acid which provides UV protection	Ichthyosis vulgaris
Loricrin	Granular layer	Most abundant component of the cornified cell envelope	Vohwinkel syndrome without deafness
Involucrin	Granular layer	First protein to be crosslinked in the cornified cell envelope	Increased in inflammatory skin disease
Tissue Transglutaminase 1	Granular layer	Cross-links proteins in the cornified cell envelope	Lamellar ichthyosis
Tissue Transglutaminase 3	Granular layer	Cross-links proteins in the cornified cell envelope	Dermatitis Herpetiformis

Basement Membrane Zone

Molecule or structure of Importance	Location	Function	Associated Disease States
Plectin	Keratinocyte inner plasma membrane	Intracellular hemidesmosomal component	EBS with muscular dystrophy
BPAG1	Keratinocyte inner plasma membrane	Intracellular hemidesmosomal component	Bullous pemphigoid, paraneoplastic pemphigus
BPAG2	Keratinocyte plasma membrane to lamina densa	Transmembrane protein binding to BPAG1, plectin, integrins, laminin 5	BP, linear IgA, cicatricial pemphigoid, generalized atrophic benign EB, herpes gestationis, bullous LP
alpha 6 beta 4 integrin	Keratinocyte plasma membrane to lamina densa	Transmembrane protein binding to BPAG1, BPAG2, plectin, laminin 5	EB with pyloric atresia
Laminin 5 (laminin 332)	Lamina Densa	Binds integrins and BPAG2 from lamina lucia, and collagen VII from sub lamina densa	Junctional EB
Anchoring Fibrils	Sublamina densa	Composed mostly of collagen 7, binds laminin 5 from lamina densa and collagen I and III in sub lamina densa	Dystrophic EB, EBA, bullous lupus