

Disorders of Epidermal Maturation and Keratinization (Weedon Chapter 9)

Paul K. Shitabata, M.D.
Dermatopathology Institute

Localization of the Keratins

Epidermal Layer	Keratins
Basal	
Prickle Cell	
Nail bed	

Localization of the Keratins

Epidermal Layer	Keratins
Basal	K5 and K14
Prickle Cell	K1 and K10
Nail Bed	K6, K16, and K17

Match the Following

Keratinization	Characteristic
Keratohyaline granules	Involucrin
Inner root sheath	Trichohyalin
Cell envelope	Odland bodies

Match the Following

Keratinization	Characteristic
Keratohyaline granules	Odland bodies
Inner root sheath	Trichohyalin
Cell envelope	Involucrin

Match the following:

Disease	Defect
Darier's disease	Peroxisomes
Ichthyosis vulgaris	Desmosomes
Refsum disease	Calcium pump
Striate palmoplantar keratoderma	Filaggrin processing

Match the following:

Disease	Defect
Darier's disease	Calcium pump
Ichthyosis vulgaris	Filaggin processing
Refsum disease	Peroxisomes
Striate palmoplantar keratoderma	Desmosomes

Associations with cornoid lamella include all except:

- A. Decreased p53 expression
- B. Decreased p21 expression
- C. Decreased mdm2 expression
- D. Abnormal DNA ploidy
- E. Clonality

Answer A

It is associated with overexpression of p53

Grover's Disease vs. Darier's Disease

Characteristic	Grover's Disease	Darier's Disease
Abnormality of ATP ₂ A ₂		
Corp ronds/corp grains		
Direct immunofluorescence		
Projection of cytoplasmic processes from basal keratinocytes into dermis		

Grover's Disease vs. Darier's Disease

Characteristic	Grover's Disease	Darier's Disease
Abnormality of ATP ₂ A ₂		+
Corp ronds/corp grains	+	+
Direct immunofluorescence	-	-
Projection of cytoplasmic processes from basal keratinocytes into dermis		+

Hailey-Hailey Disease vs. Darier's Disease

Characteristic	Hailey-Hailey Disease	Darier's Disease
Defect in ATP ₂ C ₁		
Defect in ATP ₂ A ₂		
Autosomal Dominant		
Induced by UV radiation		
Keratinocyte abnormalities present in uninvolved skin		

Hailey-Hailey Disease vs. Darier's Disease

Characteristic	Hailey-Hailey Disease	Darier's Disease
Defect in ATP _{2C1}	+	
Defect in ATP _{2A2}		+
Autosomal Dominant	+	+
Induced by UV radiation	+	+
Keratinocyte abnormalities present in uninvolved skin	+	