

Pemphigus Vulgaris

Pemphigus is an **autoimmune disorder** against **desmoglein**. It's present in **desmosomes** that interconnect the epithelial cells of the epidermis. Because the destruction is between epithelial cells (**intra-epidermal**), the blister is **thin** and **tears easily** (+ **Nikolsky's Sign**). Diagnosis is made by **biopsy** showing a **tombstone** effect as basement membrane cells remain attached while epithelial cells split apart from each other. **Immunofluorescence** reveals **antibodies** on **epithelial cells throughout** the skin lesion. Because it's an autoimmune disease, treatment starts with **systemic steroids**. Once controlled, swap to **steroid sparing** immune modulators when possible (mycophenolate mofetil, rituximab). This disease is **life-threatening**, does **involve mucosa**, and occurs in people ages **30-50**.

Bullous Pemphigoid

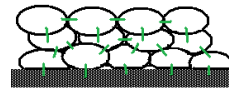
Pemphigoid is another **autoimmune disorder**, this time against the **hemidesmosomes** that attach basement membrane cells to the basement membrane (**sub-epidermal lesion**). The detachment causes a **blister** but the intact epithelium results in a **tense, rigid bullae** (⊖ Nikolsky's). Again, a biopsy is used for diagnosis showing **intact epithelium** that's **detached from the basement membrane**. **Immunofluorescence** shows antibodies at the **dermal-epidermal junction**. Treatment is with **steroids** during acute attacks – topical for limited disease, systemic for severe. This **ISN'T** life threatening and **DOESN'T** involve the mucosa. It's most commonly found in ages **70-80**.

Dermatitis Herpetiformis

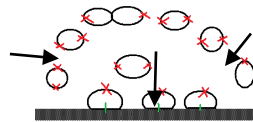
This isn't a true blistering disease but is commonly tested against them. It's another **autoimmune disease** caused by **IgA antibodies** against **transglutaminase**. It's the **cutaneous manifestation** of celiac sprue and has the same pathology. The antibody-antigen complex gets deposited at the **dermal papillae** and causes an extension of the epidermis. It manifests as multiple small vesicular eruptions that are **pruritic** and found on the buttocks / legs or **extensor surfaces**. A biopsy is not needed, though if performed it'll show "**neutrophilic abscess**." Make the diagnosis instead by diagnosing the Sprue with anti-endomysial or anti-transglutaminase antibodies and an endoscopy. Treat the skin manifestation by treating the underlying disease: **remove gluten from diet** entirely.

Porphyria Cutanea Tarda

The most common porphyria disease, it's the lowest yield of the four. **Bullae on sun-exposed areas** is highly suspicious for the disease. The diagnosis is made with **coral red urine under Wood's Lamp** caused by accumulation of urinary uroporphyrins. The underlying etiology is a deficiency of uroporphyrinogen decarboxylase but can be brought on by OCPs, alcohol, Hep C, or Hemochromatosis. **Avoid the sun**.

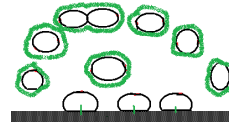


Basement Membrane

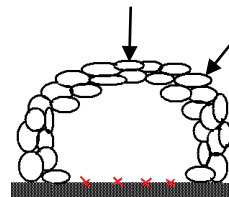


Weak Barriers with lots of gaps
(+ Nikolsky's)

X Desmosome Destroyed
Hemidesmosome Intact



IF lights up the epidermis
around the cells where the
antibodies are

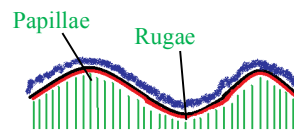


Strong barrier
(⊖ Nikolsky's)

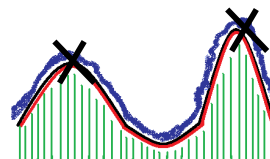
X Hemidesmosome Destroyed
Desmosome Intact



IF at the dermal epidermal
junction



Papillae
Rugae
Epidermis
Basement Membrane
Dermis



IgA Deposition
@ Papillae
Papillae enlarge

Disease	Age	Mucosa	Blisters	Target	Dx	IF	Tx
Pemphigus Vulgaris	40-50	Involved	+ Nikolsky Thin, Tears	Dermatomes (intracellular)	Bx	Within Epidermis	Steroids → MM
Bullous Pemphigoid	70-80	No	⊖ Nikolsky Tense, Tough	Hemidesmosomes (to BM)	Bx	On Basement Membrane Epidermal-Dermal Junction	Steroids
Dermatitis Herpetiformis	20-30	No	⊖ Nikolsky	IgA Deposition @ papillae	Antibodies	Deposition at the papillae (though this is not needed)	Remove Gluten from Diet
Porphyria Cutanea Tarda	Any	No	⊖ Nikolsky Tense, Tough	N/A	Wood's Lamp	Don't do it	Avoid the sun