



Abbreviations

DIF	Direct immunofluorescence
ELISA	Enzyme-linked immunosorbent assay
IgG	Immunoglobulin G
IIF	Indirect immunofluorescence
PAMS	Paraneoplastic autoimmune multiorgan syndrome
PNP	Paraneoplastic pemphigus

^aFor all suspected immunobullous diseases, biopsy specimens should be obtained from perilesional tissue because immunoreactants may not be present in lesional (blistered or eroded) tissue; perilesional tissue is defined as immediately adjacent to, but not involving, a blister or erosion and may include inflamed, intact skin or mucosa.

^bLesional biopsy specimens should be intact skin or mucosa from a newly developed lesion and/or inflamed skin.

^cIndividuals with PNP may demonstrate serum IgG antibodies to multiple epithelia (simple, columnar, transitional) and against envoplakin; desmoglein 1, 3; desmoplakin 1, 2; periplakin; and/or BP230 and BP180. Envoplakin IgG antibody is highly specific for PNP, and testing for this antibody should therefore be performed; desmoglein 1 and 3 IgG antibody testing and BP230 and BP180 IgG antibody testing should also be performed, along with IIF antibody testing on rodent substrates to increase sensitivity.

^dRare cases of IgA PNP have been reported; consider testing for IgA PNP antibodies by IIF using rodent substrates.

^eNotably, in some individuals with PNP/PAMS, serologic findings may be positive while DIF results are negative, and vice versa.

^fConsider antilaminin-332 pemphigoid and nonclassical pemphigus (ie, intercellular IgG/IgA dermatosis), both which have strong malignancy associations.

^gA general immunobullous disease workup may provide additional information useful to characterize presentation and, along with PNP/PAMS serologies, to monitor treatment response; refer to [Immunobullous Diseases Testing Algorithm - Epithelial Antibody Associated](#) for additional details on testing strategy.