

**Overview****Useful For**

Confirming a diagnosis of pemphigoid, pemphigus, epidermolysis bullosa acquisita, or bullous lupus erythematosus

**Method Name**

Indirect Immunofluorescence Assay (IFA)

**NY State Available**

Yes

**Specimen****Specimen Type**

Serum

**Specimen Required****Container/Tube:**

**Preferred:** Serum gel

**Acceptable:** Red top

**Specimen Volume:** 2 mL

**Specimen Minimum Volume**

0.5 mL

**Reject Due To**

Gross hemolysis	OK
Gross lipemia	Reject
Gross icterus	OK

**Specimen Stability Information**

Specimen Type	Temperature	Time	Special Container
Serum	Refrigerated (preferred)	14 days	
	Frozen	30 days	
	Ambient	14 days	

**Clinical and Interpretive****Clinical Information**

Immunoglobulin A anti-basement zone (BMZ) antibodies are produced by patients with pemphigoid. In most patients

with bullous pemphigoid, serum contains IgA anti-BMZ antibodies, while in cicatricial pemphigoid circulating IgA anti-BMZ antibodies are found in a minority of cases. Sensitivity of detection of anti-BMZ antibodies is increased when serum is tested using sodium chloride (NaCl)-split human skin as substrate.

Circulating IgA anti-BMZ antibodies are also detected in patients with epidermolysis bullosa acquisita (EBA) and bullous eruption of lupus erythematosus.

IgA anti-cell surface (CS) antibodies are produced by patients with pemphigus. The titer of anti-CS antibodies generally correlates with disease activity of pemphigus.

### Reference Values

Report includes presence and titer of circulating antibodies. If serum contains basement zone (BMZ) antibodies on split-skin substrate, patterns will be reported as: 1) epidermal pattern, consistent with pemphigoid or 2) dermal pattern, consistent with epidermolysis bullosa acquisita.

Negative in normal individuals

### Interpretation

Indirect immunofluorescence (IF) testing may be diagnostic when histologic or direct IF studies are only suggestive, nonspecific, or negative.

Anti-cell surface (CS) antibodies correlate with a diagnosis of pemphigus.

Anti-basement zone (BMZ) antibodies correlate with a diagnosis of bullous pemphigoid, cicatricial pemphigoid, epidermolysis bullosa acquisita (EBA), or bullous eruption of lupus erythematosus (LE).

If serum contains anti-BMZ antibodies, the pattern of fluorescence on sodium chloride (NaCl)-split skin substrate helps distinguish pemphigoid from EBA and bullous LE. Staining of the roof (epidermal side) or both epidermal and dermal sides of NaCl-split skin correlates with the diagnosis of pemphigoid, while fluorescence localized only to the dermal side of the split-skin substrate correlates with either EBA or bullous LE.

### Cautions

Results should be interpreted in conjunction with clinical information, histologic pattern, and results of direct immunofluorescence (IF) study. In particular, the finding of low titer (< or =1:80) anti-CS antibodies should not be used alone (ie, without histologic or direct IF support) to confirm a diagnosis of pemphigus.

### Clinical Reference

1. Caux F, Kirtschig G, Lemarchand-Venencie F, et al: IgA-epidermolysis bullosa acquisita in a child resulting in blindness. *Br J Dermatol.* 1997 Aug;137(2):270-275
2. Chorzelski TP, Jablonska S: IgA linear dermatosis of childhood (chronic Bullous disease of childhood). *Br J Dermatol.* 1979 Nov;101(5):535-542
3. Guide SV, Marinkovich MP: Linear IgA bullous dermatosis. *Clin Dermatol.* Nov-Dec 2001;19(6):719-727
4. Hashimoto T, Ebihara T, Nishikawa T: Studies of autoantigens recognized by IgA anti-keratinocyte cell surface antibodies. *J Dermatol Sci.* 1996 Apr;12(1):10-17
5. Lally A, Chamberlain A, Allen J, Dean D, Wojnarowska F: Dermal-binding linear IgA disease: an uncommon subset of a rare immunobullous disease. *Clin Exp Dermatol.* 2007 Sep;32(5):493-498
6. Tsuruta D, Ishii N, Hamada T, et al: IgA pemphigus. *Clin Dermatol.* 2011 Jul-Aug;29(4):437-442

7. Vodegel RM, de Jong MCJM, Pas HH, Jonkman MF: IgA-mediated epidermolysis bullosa acquisita: two cases and review of the literature. J Am Acad Dermatol. 2002 Dec;47(6):919-925

8. Willsteed E, Bhogal BS, Black MM, McKee P, Wojnarowska F: Use of 1M NaCl split skin in the indirect immunofluorescence of the linear IgA bullous dermatoses. J Cutan Pathol. 1990 Jun;17(3):144-148

9. Wilson BD, Beutner EH, Kumar V, Chorzelski TP, Jablonska S: Linear IgA bullous dermatosis. An immunologically defined disease. Int J Dermatol. 1985 Nov;24(9):569-574

10. Wojnarowska F, Collier PM, Allen J, Millard PR: The localization of the target antigens and antibodies in linear IgA disease is heterogeneous, and dependent on the methods used. Br J Dermatol. 1995 May;132(5):750-757

## Performance

### Method Description

Frozen sections of rhesus monkey esophagus and sodium chloride-split human skin are overlaid with dilutions of patient's serum, incubated, covered with fluorescein-conjugated IgA antiserum, and interpreted with a fluorescence microscope.(Unpublished Mayo method)

### PDF Report

No

### Day(s) Performed

Monday through Friday

### Report Available

2 to 7 days

### Specimen Retention Time

14 days

### Performing Laboratory Location

Rochester

## Fees and Codes

### Fees

- Authorized users can sign in to [Test Prices](#) for detailed fee information.
- Clients without access to Test Prices can contact [Customer Service](#) 24 hours a day, seven days a week.
- Prospective clients should contact their Regional Manager. For assistance, contact [Customer Service](#).

### Test Classification

This test was developed, and its performance characteristics determined by Mayo Clinic in a manner consistent with CLIA requirements. This test has not been cleared or approved by the US Food and Drug Administration.

### CPT Code Information

88346

88350

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**LOINC® Information**

Test ID	Test Order Name	Order LOINC Value
CIFA	Cutaneous Immfluo. Ab (IgA), S	In Process

Result ID	Test Result Name	Result LOINC Value
610628	Cell Surface Ab IgA	In Process
610629	Basement Membrane IgA	In Process
610630	Monkey Esophagus IgA	In Process
610631	Human Split Skin IgA	In Process
610632	Other	48767-8