# Test Definition: CIFA

Cutaneous Immunofluorescence Antibodies, IgA, Serum

## Overview

### Useful For

Confirming the presence of IgA antibodies to diagnose pemphigoid, pemphigus, epidermolysis bullosa acquisita, or bullous lupus erythematosus

### Testing Algorithm

For information see Autoimmune Blistering Disease Diagnostic Algorithm.

### Special Instructions

- Autoimmune Blistering Disease Diagnostic Algorithm

### Method Name

Indirect Immunofluorescence Assay (IFA)

### NY State Available

Yes

## Specimen

### Specimen Type

Serum

### Specimen Required

**Supplies:** Sarstedt Aliquot Tube, 5 mL (T914)
**Collection Container/Tube:**
**Preferred:** Serum gel
**Acceptable:** Red top
**Submission Container/Tube**: Plastic vial
**Specimen Volume:** 2 mL Serum
**Collection Instructions**: Centrifuge and aliquot serum into a plastic vial.

### Specimen Minimum Volume

Serum: 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 | |
| | Ambient | 14 days | |
| | Frozen | 30 days | |

## Clinical & Interpretive

### Clinical Information

Immunoglobulin A anti-basement membrane 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-split primate skin as substrate.

Circulating IgA anti-BMZ antibodies are also detected in patients with epidermolysis bullosa acquisita 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

Negative

### Interpretation

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

Results will be negative in individuals without any known autoimmune blistering disease.

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

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

Anti- basement membrane zone 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-cell surface 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;137(2):270-275
2. Chorzelski TP, Jablonska S. IgA linear dermatosis of childhood (chronic bullous disease of childhood). Br J Dermatol. 1979;101(5):535-542
3. Guide SV, Marinkovich MP. Linear IgA bullous dermatosis. Clin Dermatol. 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;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;32(5):493-498
6. Tsuruta D, Ishii N, Hamada T, et al. IgA pemphigus. Clin Dermatol. 2011;29(4):437-442
7. Vodegel RM, de Jong MC, Pas HH, Jonkman MF. IgA-mediated epidermolysis bullosa acquisita: two cases and review of the literature. J Am Acad Dermatol. 2002;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;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;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;132(5):750-757
11. Tirumalae R, Kalegowda IY. Role of BIOCHIP indirect immunofluorescence test in cutaneous vesiculobullous diseases. Am J Dermatopathol. 2020;42(5):322-328

## Performance

### Method Description

Frozen sections of primate esophagus and sodium chloride-split primate 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

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### Report Available
2 to 7 days

### Specimen Retention Time
30 days

### Performing Laboratory Location
Mayo Clinic Laboratories - Rochester Main Campus

## Fees & 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 account representative. 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. It has not been cleared or approved by the US Food and Drug Administration.

### CPT Code Information
88346

### LOINC® Information

| Test ID | Test Order Name | Order LOINC® Value |
|---|---|---|
| CIFA | Cutaneous Immfluor. Ab (IgA), S | 104828-9 |

| Result ID | Test Result Name | Result LOINC® Value |
|---|---|---|
| 610628 | Cell Surface Ab IgA | 104829-7 |
| 610629 | Basement Membrane IgA | 104830-5 |
| 610630 | Primate Esophagus IgA | 104833-9 |
| 610631 | Primate Split Skin IgA | 104834-7 |
| 610632 | Other | 48767-8 |