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**PATIENT REPORT**

Patient Age/Sex: 32 years Female

**Specimen Collected: 21-Jun-22 15:44**

**Pemphigus Ab, IgA** **|Received: 22-Jun-22 09:57** **Report/Verified: 22-Jun-22 11:48**

| Procedure | Result | Units | Reference Interval |
|---|---|---|---|
| Pemphigus Ab,IgA | See Note [f1] | | |

## Result Footnote

f1: Pemphigus Ab, IgA
CLINICAL INFORMATION
Blisters and pustules with pruritus. Presumptive diagnosis is dermatitis herpetiformis, folliculitis, IgA pemphigus.

Specimen Details
S22-IP0000503 - Serum; Collected: 6/21/2022; Received: 6/22/2022

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DIAGNOSTIC INTERPRETATION
Positive IgA cell surface antibodies by indirect immunofluorescence, consistent with IgA pemphigus

(See Results and Comments)

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RESULTS
Indirect Immunofluorescence (IIF)
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Cell Surface (CS)/Intercellular Substance (ICS) IgA Antibodies

IgA: Positive, titer 1:640 (H), monkey esophagus substrate
Positive, titer 1:160 (H), intact human skin substrate

Reference Range:
Negative - Titer less than 1:10
Borderline - Titer 1:10
Positive (H) - Titer greater than 1:10

(H) = high/positive

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COMMENTS
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Specific

These indirect immunofluorescence test results, demonstrating positive serum IgA cell surface (CS), also known as intercellular substance (ICS), antibodies reacting with both monkey esophagus substrate and intact human skin substrate, support the diagnosis of IgA pemphigus. Notably, IgA CS/ICS antibodies may be expressed in some pemphigus variants along with IgG CS/ICS antibodies.
Positive IgA CS/ICS antibody reactivity can be:
- Consistent with IgA pemphigus, a rare variant of pemphigus, including evolving or treated disease;
- Co-expressed with IgG CS/ICS antibodies in pemphigus foliaceus and pemphigus vulgaris;
- Observed in non-classical forms of pemphigus, including pemphigus herpetiformis, paraneoplastic pemphigus, and intercellular IgG/IgA dermatosis; and
- Found transiently and/or nonspecifically in normal

## Result Footnote

f1: Pemphigus Ab, IgA
individuals and in patients with various mucocutaneous
disorders including drug reactions and infections.
Approximately 40 percent of patients with nonclassical IgG/IgA pemphigus have an underlying systemic disease when diagnosed, malignancy being the most common.

Clinical correlation is needed, including with direct immunofluorescence findings on a biopsy specimen and treatment status, with further clinical evaluation as indicated. Additional serum testing for IgG CS/ICS antibodies by indirect immunofluorescence and ELISAs may be performed on this serum specimen by contacting ARUP Client Services at 1-800-242-2787, option 2, with add-on test request for: Pemphigus Antibody Panel, IgG (ARUP test number 0090650).

Detection, levels, and patterns of diagnostic antibodies may fluctuate with disease manifestations. Monitoring serum antibody profiles by indirect immunofluorescence and antibody levels by ELISAs may aid in assessing disease expression and activity, including response to therapy.

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General

IgA cell surface (CS)/intercellular substance (ICS) antibodies are positive in patients with IgA pemphigus, a rare type of pemphigus, also known as intercellular IgA dermatosis. IgA pemphigus presents as two major subtypes, the subcorneal pustular dermatosis (SPD) type and the intraepidermal neutrophilic (IEN) type; however, three other IgA pemphigus variants are recognized, IgA-pemphigus vegetans, IgA-pemphigus vulgaris, and unclassified IgA pemphigus. Positive IgA CS/ICS antibodies are found along with positive IgG CS/ICS antibodies in some pemphigus variants (References).

Monkey esophagus and intact normal human skin substrates may demonstrate differing sensitivities and specificities for disease-associated antibodies and, when tested together, increase the likelihood of detecting IgA cell surface antibodies. The presence of IgA epithelial antibody reactivity may give indication for treatment consideration with dapsone (if glucose-6-phosphate dehydrogenase, G6PD, is normal). IgA CS/ICS antibodies may be undetected in individual patients with IgA pemphigus whose disease is minimal and/or under control.

(References:

- Mentink LF, de Jong MC, Kloosterhuis GJ, et al. Coexistence of IgA antibodies to desmogleins 1 and 3 in pemphigus vulgaris, pemphigus foliaceus and paraneoplastic pemphigus. Br J Dermatol. 2007 Apr;156(4):635-41. doi: 10.1111/j.1365-2133.2006.07717.x. Epub 2007 Jan 30. PMID: 17263817.
- Porro AM, Caetano Lde V, Maehara Lde S, Enokihara MM. Non-classical forms of pemphigus: pemphigus herpetiformis, IgA pemphigus, paraneoplastic pemphigus and IgG/IgA pemphigus. An Bras Dermatol. 2014 Jan-Feb;89(1):96-106. doi: 10.1590/abd1806-4841.20142459. PMID: 24626654; PMCID: PMC3938360.
- Hashimoto T, Teye K, Hashimoto K, et al. Clinical and Immunological Study of 30 Cases With Both IgG and IgA Anti-Keratinocyte Cell Surface Autoantibodies Toward the Definition of Intercellular IgG/IgA Dermatosis. Front Immunol. 2018 May 7;9:994. doi: 10.3389/fimmu.2018.00994. PMID: 29867971; PMCID: PMC5950707.
- Criscito MC, Cohen JM, Toosi S, et al. A retrospective study on the clinicopathologic features of IgG/IgA pemphigus. J Am Acad Dermatol. 2021 Jul;85(1):237-240.

## Result Footnote

f1: Pemphigus Ab, IgA
doi: 10.1016/j.jaad.2020.07.126. Epub 2020 Aug 13.
PMID: 32798577.)

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TESTING METHODS
Indirect Immunofluorescence (IIF)
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IgA Epithelial Cell Surface (CS)/Intercellular Substance (ICS) Antibodies

Patient serum is progressively diluted in calcium-containing buffer beginning at 1:5 in three two-fold screening dilutions, layered on sections of intact normal human skin and monkey esophagus substrates, and reacted with fluorescein isothiocyanate (FITC)-conjugated antibody to IgA. When positive, the serum is further diluted in two-fold reductions to the limiting dilution of antibody detection or to a maximum dilution of 1:40,960. The limiting-dilution, end-point titer is reported for each substrate. This indirect immunofluorescence testing was developed and its performance characteristics determined by the Immunodermatology Laboratory at the University of Utah. It has not been cleared or approved by the FDA (US Food and Drug Administration). FDA clearance or approval currently is not required for this testing performed in a CLIA-certified laboratory (Clinical Laboratory Improvement Amendments) and intended for clinical use. [Indirect immunofluorescence, one antibody on two substrates (IIF X 2) with two limiting-dilution, end-point titers (antibody titer X 2)]

Electronically signed by Kristin M. Leiferman, MD, on 06/22/22 at 11:46 AM.
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