PATIENT REPORT

500 Chipeta Way, Salt Lake City, Utah 84108-1221 phone: 801-583-2787, toll free: 800-522-2787 Jonathan R. Genzen, MD, PhD, Chief Medical Officer

Patient Age/Sex:

Unknown

| Specimen Collected: 2/6/2025 09:35 | MST | | |
|--|------------------------------|--------------------|---|
| Autoimmune Pediatric CNS | Received: 2/6/2025 | 09:36 MST | Report/Verified: 2/6/2025 10:00 MST |
| Procedure | Result | Units | Reference Interval |
| Purkinje Cell/Neuronal Nuclear | ANNA Detected * | f1 i1 | [None Detected] |
| IgG Scrn | | | |
| MDA Receptor Ab IgG CBA-IFA, | 1:160 * f2 i2 | | [<1:10] |
| Gerum | | | |
| CASPR2 Ab IgG CBA-IFA Screen, | Detected * t1 i3 | | [<1:10] |
| Gerum | | | |
| GI1 Ab IgG CBA-IFA Screen,Serv | ım Detected * t2 i4 | | [<1:10] |
| MO/AQP4 Ab IgG CBA-IFA Screen, | Detected * t3 i5 | | [<1:10] |
| Gerum | | | |
| MPA Receptor Ab IgG CBA-IFA | Detected * t4 i6 | | [<1:10] |
| Scrn, Serum | | | |
| ABA-BR Ab IgG CBA-IFA Scrn,Ser | Detected * t5 i7 | | [<1:10] |
| MOG Ab IgG CBA-IFA Screen,Serum | Detected * t6 i8 | | [<1:10] |
| OPPX Ab IgG CBA-IFA Screen, Seru | ım Detected * t7 i9 | | [<1:10] |
| GABA-AR Ab IgG CBA-IFA Screen, | Detected * t8 i10 | | [<1:10] |
| Serum | | | |
| nGluR1 Ab IgG CBA-IFA Screen, | Detected * t9 i11 | | [<1:10] |
| Serum | | | |
| Slutamic Acid Decarboxylase | 10.0 H il2 | IU/mL | [0.0-5.0] |
| Antibody | | | |
| euronal Ab (TR/DNER) IgG, Ser | Received: 2/6/2025 | 09:36 MST | Report/Verified: 2/6/2025 10:0 MST |
| rocedure | Result | Units | Reference Interval |
| Meuronal Ab (TR/DNER) IgG,Ser | High Positive * | 113 | [Negative] |
| euronal Nuclear Ab (ANNA) IFA | Received: 2/6/2025 | 09:36 MST | Report/Verified: 2/6/2025 10:0 MST |
| rocedure | Result | Units | Reference Interval |
| euronal Nuclear Ab (ANNA) IFA | 1:640 * i14 | | [<1:10] |
| iter IgG | | | |
| Meuronal Nuclear Ab IgG, | Received: 2/6/2025 | 09:36 MST | Report/Verified: 2/6/2025 10:0 |
| mmunoblot, Hu | | | MST |
| rocedure | Result | Units | Reference Interval |
| Meuronal Nuclear Ab (Hu) IgG,IE | 3, Positive * ⁱ¹⁵ | | [Negative] |
| caronar Nacrear Ab (na, 190, 11 | | | |
| | | | |
| erum MPA Rptr Ab IgG Titer by | Received: 2/6/2025 | 09:36 MST | Report/Verified: 2/6/2025 10:0 MST |
| Gerum LMPA Rptr Ab IgG Titer by LBA-IFA, Ser | Received: 2/6/2025 | 09:36 MST Units | Report/Verified: 2/6/2025 10:0 MST Reference Interval |
| Serum | | | MST |

Unless otherwise indicated, testing performed at:

ARUP Laboratories

500 Chipeta Way, Salt Lake City, UT 84108

Laboratory Director: Jonathan R. Genzen, MD, PhD

ARUP Accession: Report Request ID: 20291687

25-037-900089

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^{*=}Abnormal, #=Corrected, C=Critical, f=Result Footnote, H-High, i-Test Information, L-Low, t-Interpretive Text, @=Performing lab

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| CASPR2 Ab IgG Titer by CBA-IFA, Ser | Received: 2/6/20 | 25 09:36 MST | Report/Verified: 2/6/2025 10:04 MST |
|---|---------------------------------------|--------------|--|
| Procedure CASPR2 Ab IgG CBA-IFA Titer, Serum | Result 1:320 * ¹¹⁷ | Units | Reference Interval [<1:10] |
| NMO/AQP4-Ab IgG Titer by CBA-IFA Ser | , Received: 2/6/20 | 25 09:36 MST | Report/Verified: 2/6/2025 10:04 MST |
| Procedure NMO/AQP4 Ab IgG CBA-IFA Titer Serum | Result , 1:20 * ¹¹⁸ | Units | Reference Interval [<1:10] |
| DPPX Ab IgG Titer by CBA-IFA, Set | r Received: 2/6/20 | 25 09:36 MST | Report/Verified: 2/6/2025 10:04 MST |
| Procedure DPPX Ab IgG CBA-IFA Titer, Ser | Result um 1:160 * ¹¹⁹ | Units | Reference Interval [<1:10] |
| GABA-A Receptor IgG CBA-IFA Titer, Serum | Received: 2/6/20 | 25 09:36 MST | Report/Verified: 2/6/2025 10:04 MST |
| Procedure GABA-AR Ab IgG CBA-IFA Titer, Serum | Result 1:80 * ⁱ²⁰ | Units | Reference Interval [<1:10] |
| GABA-B Rptr Ab IgG Titer by CBA-IFA, Ser | Received: 2/6/20 | 25 09:36 MST | Report/Verified: 2/6/2025 10:04 MST |
| Procedure GABA-BR Ab IgG CBA-IFA Titer, | Result Ser 1:2560 * ¹²¹ | Units | Reference Interval [<1:10] |
| LGI1 Ab IgG Titer by CBA-IFA, Set | r Received: 2/6/20 | 25 09:36 MST | Report/Verified: 2/6/2025 10:04 MST |
| Procedure LGI1 Ab IgG CBA-IFA Titer, Ser | Result um 1:640 * ¹²² | Units | Reference Interval [<1:10] |
| mGluR1 Ab IgG CBA-IFA Titer, Serum | Received: 2/6/20 | 25 09:36 MST | Report/Verified: 2/6/2025 10:04 MST |
| Procedure mGluR1 Ab IgG CBA-IFA Titer, Serum | Result >1:2560 * i23 | Units | Reference Interval [<1:10] |
| MOG Ab IgG Titer by CBA-IFA, Ser | Received: 2/6/20 | 25 09:36 MST | Report/Verified: 2/6/2025 10:04 MST |
| Procedure MOG Ab IgG CBA-IFA Titer, Seru | Result m 1:80 * 124 | Units | Reference Interval [<1:10] |
| <u>Interpretive Text</u> t1: 2/6/2025 09:35 MST (CASPR2 Ab |) IgG CBA-IFA Screen, | Serum) | |

CASPR2 Antibody, IgG is detected. Titer results to follow.

t2: 2/6/2025 09:35 MST (LGI1 Ab IgG CBA-IFA Screen, Serum)

LGI1 Antibody, IgG is detected. Titer results to follow.

2/6/2025 09:35 MST (NMO/AQP4 Ab IgG CBA-IFA Screen, Serum) t3:

Aquaporin-4 Receptor Antibody, IgG is detected. Titer results to follow.

2/6/2025 09:35 MST (AMPA Receptor Ab IgG CBA-IFA Scrn, Serum) t4:

AMPAR Antibody, IgG is detected. Titer results to follow.

2/6/2025 09:35 MST (GABA-BR Ab IgG CBA-IFA Scrn, Ser) t5:

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Interpretive Text

t5: 2/6/2025 09:35 MST (GABA-BR Ab IgG CBA-IFA Scrn, Ser)

GABA-BR Antibody, IgG is detected. Titer results to follow.

2/6/2025 09:35 MST (MOG Ab IgG CBA-IFA Screen, Serum) t6:

MOG Antibody, IgG is detected. Titer results to follow.

2/6/2025 09:35 MST (DPPX Ab IgG CBA-IFA Screen, Serum) t7:

DPPX Antibody, IgG is detected. Titer results to follow.

2/6/2025 09:35 MST (GABA-AR Ab IgG CBA-IFA Screen, Serum) t8:

GABA-AR Antibody, IqG is detected. Titer results to follow.

t9: 2/6/2025 09:35 MST (mGluR1 Ab IgG CBA-IFA Screen, Serum)

mGluR1 Antibody, IgG is detected. Titer results to follow.

Result Footnote

Purkinje Cell/Neuronal Nuclear IgG Scrn

Antibodies detected, therefore IFA titer and Immunoblot testing to be performed.

f2: NMDA Receptor Ab IgG CBA-IFA, Serum

Antibodies to NMDA were detected; titer was performed at an additional charge.

The ExTINGUISH Trial (safety and efficacy of Inebilizumab in anti-NMDA receptor encephalitis, NCT04372615) is actively recruiting patients. To learn more, or to refer your patient, call 1-844-427-2465, email ExTINGUISH@hsc.utah.edu, or visit https://neuronext.org/projects/nnll1-extinguish.

<u>Test Information</u>

i1: Purkinje Cell/Neuronal Nuclear IgG Scrn INTERPRETIVE INFORMATION: Purkinje Cell/Neuronal Nuclear IgG Scrn

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

NMDA Receptor Ab IgG CBA-IFA, Serum i2:

INTERPRETIVE INFORMATION: NMDA Receptor Ab IqG CBA-IFA, Serum

NMDA receptor antibody is found in a subset of patients with autoimmune limbic encephalitis and may occur with or without associated tumor. Decreasing antibody levels may be associated with therapeutic response. In addition, positive results have been reported in patients with non-autoimmune phenotypes. A negative test result does not rule out a diagnosis of autoimmune limbic encephalitis. Results should be interpreted in correlation with the patient's clinical history and other laboratory findings. Serum testing should be paired with CSF testing for improved diagnostic sensitivity.

This indirect fluorescent antibody assay utilizes full-length GluN1 transfected cell lines for the detection and semiquantification of NMDA receptor IgG antibody.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug

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Jonathan R. Genzen, MD, PhD, Chief Medical Officer

Patient Age/Sex:

Unknown

Test Information

i2: NMDA Receptor Ab IgG CBA-IFA, Serum

Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i3: CASPR2 Ab IgG CBA-IFA Screen, Serum

INTERPRETIVE INFORMATION: CASPR2 Ab IgG CBA-IFA Screen,

Serum

Contactin-associated protein-2 (CASPR2) IgG antibody may occur as part of the voltage-gated potassium channel (VGKC) complex antibodies.

The presence of CASPR2 IgG antibody is associated with a wide spectrum of clinical manifestations, including acquired neuromyotonia, limbic encephalitis, painful neuropathy, and Morvan syndrome. Tumors such as thymoma, small cell lung cancer, and other rarer tumors may occur. The full-spectrum of clinical disorders and tumors associated with the CASPR2 IgG antibody continues to be defined. Results should be interpreted in correlation with the patient's clinical history and other laboratory findings.

This indirect fluorescent antibody assay utilizes CASPR2 transfected cell lines for the detection and semiquantification of the CASPR2 IgG antibody.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i4: LGI1 Ab IgG CBA-IFA Screen, Serum

INTERPRETIVE INFORMATION: LGI1 Ab IgG CBA-IFA Screen, Serum

Leucine-rich, glioma-inactivated 1 protein (LGI1) IgG antibody may occur as part of the voltage-gated potassium channel (VGKC) complex antibodies.

The presence of LGI1 IgG antibody is mainly associated with limbic encephalitis, hyponatremia, and myoclonic movements. LGI1 IgG antibody is rarely associated with tumors but may occur infrequently in Morvan syndrome, neuromyotonia, and idiopathic epilepsy. The full-spectrum of clinical disorders associated with the LGI1 IgG antibody continues to be defined. Results should be interpreted in correlation with the patient's clinical history and other laboratory findings.

This indirect fluorescent antibody assay utilizes LGI1 transfected cell lines for the detection and semiquantification of the LGI1 IgG antibody.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

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Jonathan R. Genzen, MD, PhD, Chief Medical Officer

Patient Age/Sex:

Unknown

Test Information

i5: NMO/AQP4 Ab IgG CBA-IFA Screen, Serum

INTERPRETIVE INFORMATION: NMO/AQP4 Ab IgG CBA-IFA Screen,

Serum

Neuromyelitis optic (NMO) commonly presents with optic neuritis or longitudinally extensive transverse myelitis. Approximately 75 percent of patients with NMO have antibodies to the aquaporin-4 (AQP4) receptor. While the absence of AQP4 receptor antibodies does not rule out a diagnosis of NMO, presence of this antibody is diagnostic for NMO.

This indirect fluorescent antibody assay utilizes AQP4 receptor transfected cell lines for the detection and semiquantification of AQP4 IgG antibody.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i6: AMPA Receptor Ab IgG CBA-IFA Scrn, Serum

INTERPRETIVE INFORMATION: AMPA Receptor Ab IgG CBA-IFA Scrn,

Serum

Alpha-amino-3-hydroxy-5-methyl-4-isoxazoleproprionic acid receptor (AMPAR) antibody is found in a subset of patients with autoimmune limbic encephalitis and may occur with or without associated tumor. Decreasing antibody levels may be associated with therapeutic response. A negative test result does not rule out a diagnosis of autoimmune encephalitis. Results should be interpreted in correlation with the patient's clinical history and other laboratory findings.

This indirect fluorescent antibody assay utilizes AMPAR transfected cell lines for the detection and semiguantification of AMPAR IgG antibody.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i7: GABA-BR Ab IgG CBA-IFA Scrn, Ser

INTERPRETIVE INFORMATION: GABA-BR Ab IgG CBA-IFA Scrn, Ser

Gamma-amino butyric acid receptor, type B (GABA-BR) antibody is found in a subset of patients with autoimmune epilepsy and other autoimmune neurologic phenotypes; it may occur with or without associated tumor. Decreasing antibody levels may be associated with therapeutic response. A negative test result does not rule out a diagnosis of autoimmune neurologic disease. Results should be interpreted in correlation with the patient's clinical history and other laboratory findings.

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Patient Age/Sex:

Unknown

Test Information

i7: GABA-BR Ab IgG CBA-IFA Scrn, Ser

This indirect fluorescent antibody assay utilizes GABA-BR transfected cell lines for the detection and semiquantification of GABA-BR IgG antibody.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i8: MOG Ab IgG CBA-IFA Screen, Serum

INTERPRETIVE INFORMATION: MOG Ab IgG CBA-IFA Screen, Serum

Myelin oligodendrocyte glycoprotein (MOG) antibody is found in a subset of patients with neuromyelitis optica spectrum disorders including optic neuritis and transverse myelitis, brainstem encephalitis, and acute disseminated encephalomyelitis. Persistence of antibody positivity may be associated with a relapsing course. A negative test result does not rule out a diagnosis of CNS demyelinating disease. Results should be interpreted in correlation with the patient's clinical history and other laboratory findings.

This indirect fluorescent antibody assay utilizes full-length MOG transfected cell lines for the detection and semiquantification of MOG IgG antibody

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i9: DPPX Ab IgG CBA-IFA Screen, Serum INTERPRETIVE INFORMATION: DPPX Ab IgG CBA-IFA Screen, Serum

DPPX antibody is found in a subset of patients with autoimmune encephalitis, and is often associated with prodromal gastrointestinal symptoms and unintentional weight loss. It may occur with or without associated tumor. Decreasing antibody levels may be associated with therapeutic response. A negative test result does not rule out a diagnosis of autoimmune neurologic disease. Results should be interpreted in correlation with the patient's clinical history and other laboratory findings.

This indirect fluorescent antibody assay utilizes DPPX transfected cells for the detection and semiquantification of the DPPX IgG antibody.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the U.S. Food and Drug Administration. This test was performed in a CLIA-certified laboratory and is intended for clinical purposes.

i10: GABA-AR Ab IgG CBA-IFA Screen, Serum INTERPRETIVE INFORMATION: GABA-AR Ab IgG CBA-IFA Screen,

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Unknown

Test Information

i10: GABA-AR Ab IgG CBA-IFA Screen, Serum

Serum

Gamma-aminobutyric acid receptor, type A (GABA-AR) antibody is found in a subset of patients with autoimmune encephalitis or autoimmune epilepsy and may occur with or without associated tumor. A negative test result does not rule out a diagnosis of autoimmune limbic encephalitis or autoimmune epilepsy. Interpretation of any antineural antibody test requires clinical correlation.

This indirect fluorescent antibody assay utilizes GABA-AR transfected cell lines for detection and semi-quantification of GABA-AR IgG antibody.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the U.S. Food and Drug Administration. This test was performed in a CLIA-certified laboratory and is intended for clinical purposes.

ill: mGluR1 Ab IgG CBA-IFA Screen, Serum

INTERPRETIVE INFORMATION: mGluR1 Ab IgG CBA-IFA Screen,
Serum

Metabotropic glutamate receptor 1 (mGluR1) antibody is found in a subset of patients with autoimmune cerebellar ataxia or autoimmune encephalitis and may occur with or without associated tumor. A negative test result does not rule out a diagnosis of autoimmune cerebellar ataxia or limbic encephalitis. Interpretation of any antineural antibody test requires clinical correlation.

This indirect fluorescent antibody assay utilizes mGluR1 transfected cell lines for detection and semi-quantification of mGluR1 IgG antibody.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the U.S. Food and Drug Administration. This test was performed in a CLIA-certified laboratory and is intended for clinical purposes.

i12: Glutamic Acid Decarboxylase Antibody

INTERPRETIVE INFORMATION: Glutamic Acid Decarboxylase Antibody

A value greater than 5.0 IU/mL is considered positive for Glutamic Acid Decarboxylase Antibody (GAD Ab). This assay is intended for the semi-quantitative determination of the GAD Ab in human serum. Results should be interpreted within the context of clinical symptoms.

i13: Neuronal Ab (TR/DNER) IgG, Ser

INTERPRETIVE INFORMATION: Neuronal Ab (TR/DNER) IgG, Ser

This test detects IgG antineuronal antibodies to Tr (DNER) antigens.

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Unknown

Test Information

i13: Neuronal Ab (TR/DNER) IgG, Ser

Antineuronal antibodies serve as markers that aid in discriminating between a true paraneoplastic neurological disorder (PND) and other inflammatory disorders of the nervous system. Anti-Tr(DNER) is associated with Hodgkin's lymphoma.

The presence of these antineuronal antibodies supports a clinical diagnosis of PND and should lead to a focused search for the underlying neoplasm.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i14: Neuronal Nuclear Ab (ANNA) IFA Titer IgG

INTERPRETIVE INFORMATION: Neuronal Nuclear Ab (ANNA) IFA Titer IgG

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i15: Neuronal Nuclear Ab (Hu) IgG, IB, Serum

INTERPRETIVE INFORMATION: Neuronal Nuclear Ab IgG,

Immunoblot, Hu

This test detects IgG antineuronal antibodies to Hu antigens.

Antineuronal antibodies serve as markers that aid in discriminating between a true paraneoplastic neurological disorder (PND) and other inflammatory disorders of the nervous system. Anti-Hu (antineuronal nuclear antibody, type I) is associated with small-cell lung cancer.

The presence of these antineuronal antibodies supports a clinical diagnosis of PND and should lead to a focused search for the underlying neoplasm.

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the U.S. Food and Drug Administration. This test was performed in a CLIA-certified laboratory and is intended for clinical purposes.

i16: AMPA Receptor Ab IgG CBA-IFA Titer, Ser

INTERPRETIVE INFORMATION: AMPA Receptor Ab IgG CBA-IFA Titer, Ser

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i17: CASPR2 Ab IgG CBA-IFA Titer, Serum

INTERPRETIVE INFORMATION: CASPR2 Ab IgG CBA-IFA Titer, Serum

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i17: CASPR2 Ab IgG CBA-IFA Titer, Serum

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i18: NMO/AQP4 Ab IgG CBA-IFA Titer, Serum

INTERPRETIVE INFORMATION: NMO/AQP4 Ab IgG CBA-IFA Titer,

Serum

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i19: DPPX Ab IgG CBA-IFA Titer, Serum

INTERPRETIVE INFORMATION: DPPX Ab IgG CBA-IFA Titer, Serum

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i20: GABA-AR Ab IgG CBA-IFA Titer, Serum

INTERPRETIVE INFORMATION: GABA-AR Ab IgG CBA-IFA Titer,

Serum

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i21: GABA-BR Ab IgG CBA-IFA Titer, Ser

INTERPRETIVE INFORMATION: GABA-BR Ab IgG CBA-IFA Titer, Ser

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i22: LGI1 Ab IgG CBA-IFA Titer, Serum

INTERPRETIVE INFORMATION: LGI1 Ab IgG CBA-IFA Titer, Serum

This test was developed and its performance characteristics determined by ARUP Laboratories. It has not been cleared or approved by the US Food and Drug Administration. This test was performed in a CLIA certified laboratory and is intended for clinical purposes.

i23: mGluR1 Ab IgG CBA-IFA Titer, Serum

INTERPRETIVE INFORMATION: mGluR1 Ab IgG CBA-IFA Titer, Serum

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Unknown

Test Information

i23: mGluR1 Ab IgG CBA-IFA Titer, Serum

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i24: MOG Ab IgG CBA-IFA Titer, Serum

INTERPRETIVE INFORMATION: MOG Ab IgG CBA-IFA Titer, Serum

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ARUP Laboratories

500 Chipeta Way, Salt Lake City, UT 84108 Laboratory Director: Jonathan R. Genzen, MD, PhD **ARUP Accession:** 25-037-900089 **Report Request ID:** 20291687

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