

well. AIND and HC participant serum samples have been collected to obtain NfL, GFAP, UCH-L1, and Tau protein levels using Quanterix SR-XTM SIMOA. We plan to analyze and compare age and sex matched HC samples to AIND patients for each biomarker, ages ranging from 20-80 years old. Concentrations will be log transformed and analyzed with mixed model regression.

## Results

Healthy Controls (130), AIND (255), and newly diagnosed/treatment naïve MS samples (681) have already been collected. Nineteen HC and AIND (includes NMDA, LGI1, TRIM46, CRMP5, MOG, DPPX, GABA-A, GAD-65) participants were analyzed. Preliminary results for HC show mean NfL (10.3 pg/mL), GFAP (78.9 pg/mL), UCH-L1 (5.65 pg/mL), and Tau (0.53 pg/mL) levels, and for AIND patients mean NfL (186.48 pg/mL), GFAP (434.92 pg/mL), UCH-L1 (71.38 pg/mL), and Tau (51.85 pg/mL) levels. While ongoing biomarker analysis will be completed with HC that are age and sex matched, the significantly higher levels in AIND patients highlights the importance of creating baseline values in HC to understand these same biomarkers in AIND patients.

## Conclusions

Preliminary results show NfL and GFAP levels are significantly higher in AIND patients versus HC. Baseline biomarker values are essential for understanding further research in biomarkers related to AIND.

**Disclosure:** Mr. Borko has nothing to disclose. Ms. Barrera has nothing to disclose. Mr. Mizenko has nothing to disclose. The institution of Aurelie Ledreux has received research support from Alzheimer's Association. The institution of Dr. Kammeyer has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Genentech. Mrs. Ritchie has nothing to disclose. Sean Selva has nothing to disclose. The institution of Stefan Sillau has received research support from Alzheimer's Association. The institution of Stefan Sillau has received research support from Hewitt Family Foundation; State of Colorado. The institution of Stefan Sillau has received research support from PCORL. The institution of Stefan Sillau has received research support from NINR. The institution of Stefan Sillau has received research support from Michael J. Fox Foundation. The institution of Stefan Sillau has received research support from Department of Defense. The institution of Stefan Sillau has received research support from Colorado Department of Public Health and Environment. The institution of Stefan Sillau has received research support from Benign Essential Blepharospasm Research Foundation. Stefan Sillau has a non-compensated relationship as a Statistician with Novartis that is relevant to AAN interests or activities. Stefan Sillau has a non-compensated relationship as a Statistician with Biogen that is relevant to AAN interests or activities. Mr. Engebretson has nothing to disclose. Ms. Seale has stock in Johnson & Johnson. Ms. Valdez has a non-compensated relationship as a Valdez with The Rocky Mountain MS Center non-profit partner that is relevant to AAN interests or activities. Dr. Bennett has received personal compensation in the range of \$10,000-\$49,999 for serving as a Consultant for Horizon Therapeutics. Dr. Bennett has received personal compensation in the range of \$10,000-\$49,999 for serving as a Consultant for Alexion. Dr. Bennett has received personal compensation in the range of \$10,000-\$49,999 for serving as a Consultant for Genentech-Roche. Dr. Bennett has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for TG Therapeutics. Dr. Bennett has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Reistone Bio. Dr. Bennett has received personal compensation in the range of \$5,000-\$9,999 for serving as a Consultant for Abbvie. Dr. Bennett has received personal compensation in the range of \$0-\$499 for serving as a Consultant for Novartis. Dr. Bennett has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Chugai. Dr. Bennett has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Mitsubishi Tanabe. Dr. Bennett has received personal compensation in the range of \$500-\$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Clene Nanomedicine. Dr. Bennett has received personal compensation in the range of \$500-\$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Roche. Dr. Bennett has received personal compensation in the range of \$500-\$4,999 for serving as an Expert Witness for Parker, Whiteford Taylor. Dr. Bennett has received personal compensation in the range of \$5,000-\$9,999 for serving as an Expert Witness for Podoll. Dr. Bennett has received personal compensation in the range of \$5,000-\$9,999 for serving as an Expert Witness for Hogan Lovells.

The institution of Dr. Bennett has received research support from Novartis. The institution of Dr. Bennett has received research support from Alexion. Dr. Bennett has received intellectual property interests from a discovery or technology relating to health care. Dr. Bennett has received publishing royalties from a publication relating to health care. The institution of Dr. Vollmer has received personal compensation in the range of \$5,000-\$9,999 for serving as a Consultant for Biogen IDEC. The institution of Dr. Vollmer has received personal compensation in the range of \$10,000-\$49,999 for serving as a Consultant for Genentech/Roche. The institution of Dr. Vollmer has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Siranax. The institution of Dr. Vollmer has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Celgene. The institution of Dr. Vollmer has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for EMD Serono. The institution of Dr. Vollmer has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Bristol Meyers Squibb. The institution of Dr. Vollmer has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Viela Bios. The institution of Dr. Vollmer has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Novartis. The institution of Dr. Vollmer has received research support from Rocky Mountain MS Center. The institution of Dr. Vollmer has received research support from Biogen. The institution of Dr. Vollmer has received research support from Actelion. The institution of Dr. Vollmer has received research support from Genentech/Roche. The institution of Dr. Vollmer has received research support from Anokion. The institution of Dr. Vollmer has received research support from TG Therapeutics. Dr. Nair has received personal compensation in the range of \$5,000-\$9,999 for serving as a Consultant for Bristol Meyers Squibb. Dr. Nair has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Novartis. Dr. Nair has received personal compensation in the range of \$500-\$4,999 for serving on a Speakers Bureau for Sanofi-Genzyme. The institution of Dr. Nair has received research support from Genentech. The institution of Dr. Nair has received research support from Novartis. The institution of Dr. Nair has received research support from Genentech. The institution of Dr. Nair has received research support from Phrma Foundation. The institution of Dr. Nair has received research support from Bristol Meyers Squibb. The institution of Dr. Piquet has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Genentech. Dr. Piquet has received personal compensation in the range of \$500-\$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Genentech. Dr. Piquet has received personal compensation in the range of \$500-\$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Alexion. The institution of Dr. Piquet has received research support from Rocky Mountain MS Center. The institution of Dr. Piquet has received research support from Novartis. The institution of Dr. Piquet has received research support from Abbvie. The institution of Dr. Piquet has received research support from Roche/Genentech. The institution of Dr. Piquet has received research support from NYU. Dr. Piquet has received publishing royalties from a publication relating to health care. Dr. Piquet has received publishing royalties from a publication relating to health care. Dr. Piquet has received personal compensation in the range of \$5,000-\$9,999 for serving as a Litigative Consultant with US-Dept HHS/DICP.

## A Severe Case of Lupus Cerebritis

Christopher Szweczyk, Hemil Gonzalez

### Objective

Lupus cerebritis is a rare but potentially fatal complication of SLE. Prompt diagnosis and rapid initiation of therapy can prove lifesaving.

### Background

We present the case of a 31-year-old woman who presented to the emergency department with one week of fevers, headaches and confusion. Review of systems was notable for diffuse muscle and joint pains, weakness and a rash. SLE and Sjogren's were both diagnosed six months prior to admission and treatment included prednisone, methotrexate, and hydroxychloroquine. She self-discontinued therapy six weeks prior to seek natural remedies in her native Ecuador. Vitals on admission were 102.2 F, 111 beats/min, 96/67 mmHg, 16 breaths/min and 95% SO<sub>2</sub> on 2 L/min via nasal cannula. Physical exam was notable for conjunctival injection, hypopyon (OD), and oral ulcers with visible bleeding. Pelvic exam showed shallow ulcerations of the vaginal mucosa. Neurological exam was significant for diffuse weakness. There were punched-out ulcers on the digits of both hands. Laboratory markers of lupus disease

activity were markedly abnormal. An MRI of the brain showed numerous punctate foci of restricted diffusion in the supra and infratentorial brain parenchyma. Few of the lesions showed subtle rim-enhancement and microhemorrhagic foci. CSF analysis showed 220 WBC/mL (70% PMNs), glucose 13 mg/dL and protein 63.7 mg/dL. No CSF oligoclonal bands were detected. CSF/serum albumin index showed mild impairment of the blood brain barrier. Cultures of CSF, blood, urine and sputum showed no growth.

### Design/Methods

NA.

### Results

The patient improved significantly upon initiation of pulsed corticosteroids, plasma exchange, and cyclophosphamide. She was transitioned to steroid-sparing agents and is doing well.

### Conclusions

Lupus cerebritis can be the dominant syndrome in a patient presenting with uncontrolled SLE. Imaging and CSF findings can be dramatic and evoke infectious syndromes. Once alternative diagnoses have been ruled out lupus cerebritis should be managed aggressively to ensure good outcomes.

**Disclosure:** Mr. Szewczyk has nothing to disclose. The institution of Hemil Gonzalez has received research support from NIH.

## A Rapidly Fatal Case of Anti-GFAP Receptor Encephalitis Due to Acute Brain Edema and Herniation

Roua Kahila, Zafar Kaleem

### Objective

Glial fibrillary acidic protein antibody (GFAP) is a newly recognized biomarker for an immunotherapy responsive autoimmune meningo-encephalomyelitis with a wide variety of clinical presentations. We report the second GFAP antibody positive case in a young man who died despite appropriate and aggressive immunomodulatory treatment.

### Background

29 year old previously healthy male with childhood immune disorder presented with 3 week history of acute progressive worsening headaches, bloody emesis, nausea, blurry vision and generalized weakness. Exam was significant for downbeating nystagmus, limb ataxia and tremor and later progressing into inattention, confusion, urinary retention, asymmetric pupils, hyperreflexia and lack of motor or sensory response. Lumbar puncture revealed lymphocytic pleocytosis with elevated protein and opening pressure of 36 cm H<sub>2</sub>O. MRI demonstrated areas of restricted diffusions symmetrically involving white matter of the corpus callosum, middle cerebellar peduncle, cerebellar white matter bilaterally as well as within the pons centrally. Patient was started on intravenous immunoglobulin (IVIG) and pulse corticosteroids along with broad spectrum antimicrobial therapy. After an initial apparent response to treatment, repeat head CT showed Diffuse Sulci effacement. Shortly after, He rapidly decompensated with clinical findings indicating brainstem herniation, cardiac arrest and brain death was diagnosed. CSF studies subsequently were reported as positive for GFAP antibodies. An autopsy reported the cause of death as cerebellar tonsillar herniation secondary to diffuse cerebral edema. all sections showed perivascular inflammation and gliosis.

### Design/Methods

NA.

### Results

NA.

### Conclusions

This reported case of anti-GFAP meningo-encephalomyelitis is unusual for the rapid onset of cerebral edema and rapid progression to herniation and brain death occurring only 4 weeks after symptom onset. While this may be a rare complication of Anti GFAP encephalitis, clinicians should be vigilant for acutely increased intracranial pressure in patients with clinical findings of encephalitis in general.

**Disclosure:** Dr. Kahila has nothing to disclose. Zafar Kaleem has nothing to disclose.

## Characterization of Retinal Nerve Fiber Layer Thickness in a Cohort with Glutamic Acid Decarboxylase 65 and Glycine Receptor Autoimmunity

Yoji Hoshina, Ka-Ho Wong, Jonathan Galli, John Greenlee, Julia Klein, M. Paz Soldan, Stacey Clardy, Anette Fjeldstad, John Rose, Robert Kadish

### Objective

To describe the retinal nerve fiber layer (RNFL) with the demographic and clinical profile in patients with glutamic acid decarboxylase 65 (GAD65) and glycine receptor (GlyR) neurological autoimmunity.

### Background

GAD65 and GlyR autoimmunity can cause a wide range of clinical phenomena, including stiff-person spectrum disorder (SPSD) and epilepsy. Both GAD65, through  $\gamma$ -aminobutyric acid-ergic neurons, and GlyR interact in the retina. Optical coherence tomography (OCT) has previously been used in a variety of neurological disorders to establish baseline characteristics and monitor disease course. This presents a noninvasive opportunity to evaluate for a biomarker that may assist with the treatment of these rare but debilitating disorders.

### Design/Methods

OCT measures of RNFL by sectors were studied in patients with GAD65 and GlyR neurological autoimmunity and compared to that of 148 healthy control eyes. Patients' baseline characteristics were also reviewed retrospectively from medical records.

### Results

Of the 14 patients included in this study, 12 patients were female, and the mean age was  $52.6 \pm 16.8$  (22-79) years when OCT was performed. Ten had GAD65 autoimmunity and 4 had GlyR autoimmunity. Patients with GAD or GlyR autoimmunity showed lower RNFL thickness in multiple sectors compared to the healthy control group. This result was most apparent in the anti-GAD65 antibody subgroup. Eleven patients had PSD, one patient had epilepsy, and two had non-specific symptoms.

### Conclusions

This study provides insight into baseline RNFL thickness in a group with GAD65 and GlyR autoimmunity, two conditions that may produce varied symptoms. While limited by sample size, RNFL thinning was seen in the GAD65 and GlyR autoimmunity groups, and it was most evident in the anti-GAD65 subgroup. This provides a baseline characterization and suggests that future studies should be conducted to determine the utility of OCT as a biomarker for these conditions.

**Disclosure:** Dr. Hoshina has nothing to disclose. The institution of Mr. Wong has received research support from Biogen Idec. Dr. Galli has nothing to disclose. Dr. Greenlee has received personal compensation in the range of \$500-\$4,999 for serving as an Editor, Associate Editor, or Editorial Advisory Board Member for Medlink. Dr. Greenlee has received publishing royalties from a publication relating to health care. Dr. Greenlee has received publishing royalties from a publication relating to health care. An immediate family member of Ms. Klein has received personal compensation for serving as an employee of Amgen. An immediate family member of Ms. Klein has received personal compensation in the range of \$500-\$4,999 for serving on a Speakers Bureau for Amgen. Dr. Paz Soldan

# Neurology®

**A Severe Case of Lupus Cerebritis**  
Christopher Szewczyk and Hemil Gonzalez  
*Neurology* 2022;99;S9-S10  
DOI 10.1212/01.wnl.0000903100.95793.8a

**This information is current as of December 5, 2022**

<b>Updated Information &amp; Services</b>	including high resolution figures, can be found at: <a href="http://n.neurology.org/content/99/23_Supplement_2/S9.full">http://n.neurology.org/content/99/23_Supplement_2/S9.full</a>
<b>Subspecialty Collections</b>	This article, along with others on similar topics, appears in the following collection(s): <b>Cerebrospinal Fluid</b> <a href="http://n.neurology.org/cgi/collection/cerebrospinal_fluid">http://n.neurology.org/cgi/collection/cerebrospinal_fluid</a> <b>CT</b> <a href="http://n.neurology.org/cgi/collection/ct">http://n.neurology.org/cgi/collection/ct</a> <b>Low pressure syndrome</b> <a href="http://n.neurology.org/cgi/collection/low_pressure_syndrome">http://n.neurology.org/cgi/collection/low_pressure_syndrome</a>
<b>Permissions &amp; Licensing</b>	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: <a href="http://www.neurology.org/about/about_the_journal#permissions">http://www.neurology.org/about/about_the_journal#permissions</a>
<b>Reprints</b>	Information about ordering reprints can be found online: <a href="http://n.neurology.org/subscribers/advertise">http://n.neurology.org/subscribers/advertise</a>

*Neurology*® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright © 2022 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

