

ANA 2018 Program

Autoimmune Neurology

S101. A Combined Immunogenetic and Cerebrospinal Fluid Cytokine Study in Anti-Neurofascin155 Antibody-Positive Neuropathy

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S102. Quantifying Biomarkers of Neuronal Injury, Neuroinflammation and Neurotransmission in Antibody-Mediated Encephalitis

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S103. Clinical Features of Glycine Receptor Antibody Syndrome: A Series of Eleven Cases

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S104. Reversal of Immune Exhaustion against HIV in Two Patients Following Anti-PD1 Therapy

Lauren Bowen Reoma, MD, Bryan Smith, MD, Matthew Schindler, MD PhD, Irene Cortese, MD PhD, Peter Burbelo, PhD, Yoshimi Akahata, PhD, Steven Jacobson, PhD, Daniel

Reich, MD PhD, Avindra Nath, MD. NINDS, Bethesda, MD, USA.

S105. A Euthyroid Case Presenting with Unilateral Thyroid Eye Disease

Golnaz Yadollahikhales, M.D., Amy Myre Fallen, M.D., Fred Zar, M.D. University of Illinois Hospital, Chicago, IL, USA.

S106. A Tale of Two Children: Anti-MOG-Associated Encephalitis

Payal Patel, M.D., Ian Ferguson, M.D., Elijah Paintsil, MBChB, Naila Makhani, M.D., MPH, Serena Spudich, M.D. Yale University, New Haven, CT, USA.

S107. Newly Diagnosed Anti-Myelin Oligodendrocyte Glycoprotein Syndromes in the Inpatient Setting: Six-Month Experience at a Tertiary Pediatric Center

Cynthia X. Wang, MD, Benjamin Greenberg, MD MHS. UT Southwestern, Dallas, TX, USA.

S108. Seronegative Paraneoplastic Limbic Encephalitis Masquerading as Dementia

Hatem Hassanein, MD, **Sukriye D. Kara, MD**, James Ghattas, DO, Rossitza Chichkova, MD, Erika Abel, MD. University of South Florida, Tampa, FL, USA.

S109. Treatment-Responsive Cerebellar Ataxia in a Patient with Monoclonal Gammopathy of Uncertain Significance

Stephen L. Jaffe, M.D.^{1,2}, Noel G. Carlson, PhD^{3,4}, **John E. Greenlee, M.D.**^{3,4}. ¹Hunter Holmes McGuire VA Medical Center, Richmond, VA, USA, ²Virginia Commonwealth University School of Medicine, Richmond, VA, USA, ³George E Wahlen Veterans Affairs Health Care System, Salt Lake City, UT, USA, ⁴University of Utah School of Medicine, Salt Lake City, UT, USA.

S110. Neurosarcoidosis Manifesting as an Acute Stroke - An Unusual Presentation

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S111. Atypical Presentation of Posterior Reversible Encephalopathy Syndrome with Spinal Cord Lesion

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S112. A Rare Case of MAID Responding to Plasmapheresis

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contribute towards long-term reorganization and neurological dysfunction. This model may help explain acute findings of injury as they relate to long-term changes.

Cerebrovascular Disease

CD108. Depression Following Intracerebral Hemorrhage

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Background: Survivors of Intracerebral Hemorrhage (ICH) are at high risk for recurrent stroke, mortality, and cognitive impairment. Prevention and/or treatment of post-ICH depression may represent a novel strategy to ameliorate long-term outcomes, but little is known about its incidence, risk factors and prognostic significance.

Objective: To conduct a systematic investigation of the incidence, risk factors and prognostic significance of depression after ICH.

Methods: We followed ICH survivors with no prior history of depression enrolled in a single center prospective longitudinal study. Exposures of interest included hematoma characteristics, patient clinical information, APOE genotype, and MRI markers of cerebral small vessel disease. We captured outcomes of interest (depression, mortality, recurrent stroke and dementia) via medical records and telephone interviews. We utilized univariable and multivariable (Cox model) survival analyses to evaluate incidence of post-ICH depression, and to determine its prognostic impact on subsequent clinical endpoints.

Results: We enrolled and followed 695 ICH survivors for a median time of 48.5 months. Among these, 419/695 (60%) developed post-ICH depression. Post-ICH depression was associated with increased risk for mortality (Hazard Ratio [HR] 2.20, 95% CI 1.26-3.83), recurrent stroke (HR 1.88, 95% CI 1.16-3.04), and dementia (HR 1.50, 95% CI 1.05-2.14). IVH was the only hematoma characteristic associated with increased post-ICH depression risk (HR 1.99, 95% CI 1.12-3.54). Higher educational level, better functional performance, APOE ϵ 4, and MRI/CT measures of chronic white matter damage were all associated with increased post-ICH depression risk (all $p < 0.05$).

Conclusion: This systematic evaluation of ICH survivors revealed a high incidence of post-ICH depression, which in turn was associated with increased risk for recurrent stroke and post-ICH dementia. We identified intraventricular involvement as a novel risk factor for post-ICH depression. However, patient characteristics appear to play a predominant role in determining risk of post-ICH depression.

Dementia and Aging

CD110. Subclinical Epileptiform Abnormalities in Sporadic Alzheimer's Disease

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The role of epileptiform activity and hyperexcitability in Alzheimer's disease (AD) has gained significant interest recently. The first study utilizing prolonged (24 hour) EEG recordings as well as MEG recordings in subjects with AD reported subclinical epileptiform activity in 42% of people with AD without a prior history of epilepsy (Vossel et al, 2016). Subclinical epileptiform activity in these subjects was associated with faster rates of cognitive decline, suggesting that epileptiform activity may accelerate the course of AD. However, subjects in that study were young (mean age \sim 62 years old, with mean age of onset of cognitive symptoms of \sim 57 years old) and were more typical of patients with early-onset AD. Whether subclinical epileptiform activity is common or rare in late-onset (sporadic) AD remains unknown. Moreover, little is known about the brain electrical abnormalities that arise in sporadic Alzheimer's disease, particularly during sleep. We performed 1 to 3 day ambulatory scalp EEGs in elderly participants, including cognitively normal controls as well as those with amnesic mild cognitive impairment (aMCI) or mild dementia due to probable Alzheimer's disease (AD). Some AD participants had no prior history of seizures, while others had developed epilepsy secondary to their AD. Subjects were recruited from the Massachusetts Alzheimer's Disease Research Center, as well as from memory disorders clinics at the Massachusetts General Hospital and Brigham and Women's Hospital / South Shore Hospital (Boston, MA). Scalp EEG electrodes were placed according to the international 10-20 system with additional T1 and T2 anterior temporal electrodes. Two board-certified epileptologists (ADL, RAS) visually reviewed the ambulatory EEGs independently and blinded to each participant's cognitive and seizure status. They identified all seizures, epileptiform discharges, and any other abnormalities present in the recordings, and a consensus read for each EEG was generated after discussion. Among participants with AD and no prior history of epilepsy, a significant proportion of participants were found to have subclinical epileptiform abnormalities. Most epileptiform discharges had a temporal or fronto-temporal localization and occurred during sleep. Epileptiform abnormalities were almost never seen in the first 30 minutes of recording, and thus would not have been captured on a routine EEG. More work is needed to better understand the natural history of epileptiform abnormalities in Alzheimer's disease and their effect on disease trajectory.

Epilepsy

CD112. Hippocampal Gamma Predicts Associative Memory Performance as Measured by Acute and Chronic Intracranial EEG

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Direct recordings from the human brain have historically involved epilepsy patients undergoing invasive electroencephalography (iEEG) for surgery. However, these measurements are temporally limited to days and weeks and affected by clinical variables, including distraction, pain, and seizures. The RNS® System (NeuroPace, Inc.) is a chronic, closed-loop electrographic seizure detection and stimulation system. When adapted by investigators for research, the device permits cognitive testing in a controlled ambulatory setting, with measurements collected over months to years. We utilized an associative learning paradigm in 5 patients with traditional iEEG and 3 patients with chronic iEEG, and found increased hippocampal gamma (60-100 Hz) sustained at 1.3-1.5 seconds during encoding in successful versus failed trials in surgical patients, with similar results in our RNS System patients (1.4-1.6 seconds). Our findings replicate other studies demonstrating that sustained hippocampal gamma supports encoding. Importantly, we have validated the RNS System to make sensitive measurements of hippocampal dynamics during cognitive tasks in a chronic ambulatory research setting.

Global Neurology

CD115. Performance of Novel Diagnostics for Tuberculous Meningitis in Zambia

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Objective: To evaluate the performance of GeneXpert for mycobacterium tuberculosis and rifampicin resistance (Xpert MTB/RIF) on cerebrospinal fluid (CSF) and lipoarabinomannan (LAM) lateral flow assay on CSF and urine samples for the diagnosis of tuberculous meningitis. **Background:** TBM is a devastating infection of the nervous system in Zambia, where ~ 12% of the population is HIV-infected. Traditional diagnostic tests have low sensitivity or require several weeks for results. Xpert MTB/RIF is an automated, real-time PCR-based test that provides a diagnosis of pulmonary tuberculosis in 90 minutes. LAM lateral flow assay has been used to evaluate disseminated TB in urine at the bedside. Both tests have the potential to expedite TBM diagnosis in resource-limited setting, but have not been fully evaluated in sub-Saharan Africa.

Methods: From 4/4/14 to 8/31/17, adults with suspected CNS infection requiring lumbar puncture were enrolled at University Teaching Hospital in Lusaka, Zambia. Exclusion criteria included positive gram stain and cryptococcal antigen in the CSF. CSF samples underwent LAM and Xpert MTB/RIF testing. Urine samples underwent LAM testing only. Sensitivities and specificities of urine LAM, CSF LAM, and Xpert MTB/RIF were calculated using CSF MGIT TB liquid culture as the gold standard. Xpert MTB/RIF results were provided to treating providers within 24 hours to see if they impacted inpatient mortality.

Results: Five-hundred and fifty patients were recruited into the study. Seventy-four (21.6%) CSF samples were MTB culture-positive, including 68 (91.9%) from HIV-infected patients. The sensitivities of urine LAM, CSF LAM, and CSF Xpert MTB/RIF were 24.1% [14.9, 33.3], 21.9% [14.0, 29.8], and 52.9% [43.3, 62.5], respectively. The specificities were 76.1% [71.4, 80.1], 94.2% [92.0, 96.4], and 98.8% [97.8, 99.9], respectively. Xpert MTB/RIF detected rifampicin resistance on 7/55 (13%) positive samples. The cumulative inpatient and one-year mortality for patients that were diagnosed as CSF Xpert MTB/RIF positive was 47% and 65%

Conclusions: Compared to culture, CSF Xpert MTB/RIF had a sensitivity of 52.9% for TBM; advantages include rapid and inexpensive testing, as well as immediate information about rifampicin resistance. While urine and CSF LAM exhibited low sensitivities, they may still be useful as cheap, rule-in, point-of-care tests in remote settings. CSF LAM and Xpert MTB/RIF were highly specific for TBM. Real time use of CSF Xpert did not reduce inpatient mortality.

Movement Disorders

CD118. Preferential Role of the Subthalamic Nucleus and Dorsal Anterior Cingulate Cortex in Avoidant Decision Making

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Background: Neuropsychiatric symptoms including anxiety, impulsivity, apathy and depression are common in Parkinson's disease (PD). Motor symptoms can be alleviated by deep brain stimulation (DBS) of the subthalamic nucleus (STN) or globus pallidus internus (GPI). The STN and GPI are also important nodes in associative and limbic cortical-subcortical networks implicated in impulsivity, anxiety and depression. Here we aimed to elucidate the role of the STN and GPI in approach-avoidance behavior, a core neuropsychiatric dimension in depression and anxiety.

Methods: We recorded from single neurons in the human STN (n = 17 subjects, 94 neurons), GPI (n = 7 subjects, 22 neurons) and dACC (n = 5 subjects, 14 neurons) during intraoperative microelectrode mapping for placement of deep brain stimulation electrodes or cingulotomy. During