Washington University
School of Medicine
Department of Neurology

Residents' Day Symposium
Friday, May 25, 2018
7:30am – 9:15am
Farrell Learning and Teaching Center ~ Atrium

Poster presentations will begin at 7:30am

Awards will be presented at 9:00am

We encourage everyone to attend this important event.

Agner, Shannon

A 10-year-old girl with autoantibody positive cerebrospinal fluid in the setting of transient headache and neurologic deficits with CSF lymphocytosis (HANDL)

Shannon C. Agner, Craig Zaidman, Soe Mar, Rejean Guerriero

Background: Transient headache and neurologic deficits with cerebrospinal fluid (CSF) lymphocytosis (HaNDL) is an episodic, acute onset neurologic syndrome that presents with a variety of neurologic deficits including unilateral weakness and sensory changes, aphasia, nausea, vomiting, visual acuity and field changes, photophobia, papilledema and increased intracranial hypertension. It is most commonly observed in older children and young adults. The etiology is not well understood. A 10 year old previously healthy girl presented with a 3 day history of headache, vomiting and 1 day of acute onset sixth never palsy. These symptoms improved after lumbar puncture. Brain MRI was normal at that time. One week later, she developed right sided weakness, confusion and emesis. CSF showed a persistent lymphocytosis. EEG showed left sided slowing, and repeat brain MRI showed increased deoxyhemoglobin content as evidenced by increased signal of susceptibility weighted imaging (SWI) of the left cerebral hemisphere pial vasculature. Extensive infectious and oncologic workups were unremarkable. Her autoimmune encephalopathy panel was, however, positive for calcium channel binding antibody, N-type (0.24 nmol/L). Over the next week, the patient's mental status and other symptoms improved without intervention, and she was discharged home with a mild right sided weakness and papilledema as her only residual symptoms and signs. Clinical exam 8 months after presentation demonstrated a very mild residual right sided weakness, and brain MRI was normal. The patient met clinical criteria for HaNDL. However, her brain imaging was strikingly similar to that seen frequently in patients with hemiplegic migraine. We propose that HaNDL may represent an auto-antibody mediated process. Specifically, it may be a calcium channel mediated process with a similar mechanism to hemiplegic migraine.

Blattner, Maggie

Sleep disturbances are prevalent in autoimmune encephalitis

M Blattner, G de Bruin, RC Bucelli, GS Day

Introduction: Autoimmune encephalitis (AE) is increasingly recognized as an important cause of subacute cognitive decline, seizures, and encephalopathy in hospitalized patients, with an ever broadening clinical phenotype. Sleep disturbances have been reported in patients with AE, including REM sleep behavior disorder, hypersomnia and fragmented sleep, and sleep-disordered breathing. There is a need to determine the prevalence of sleep pathology in patients with AE, and to clarify the relationship between specific autoantibodies and sleep symptomatology, with the potential to improve patient outcomes through diagnosis and management of comorbid sleep dysfunction.

Methods: Clinical history and diagnostic data, including results of serum and CSF testing, EEG, and brain MRI, were retrospectively obtained from 25 consecutively-encountered patients with definite AE diagnosed at our tertiary care hospital. Polysomnography (PSG) was performed in a subset of patients, and raw data reviewed. Written informed consent was obtained from all patients or delegates, and study protocols were approved by our institution's human research protections office.

Results: Median age of this AE cohort was 52 years (range, 18-83) with intracellular autoantibodies (including ma and Hu autoantibodies) identified in 6/25 (24%) patients and cell-surface autoantibodies (including NMDAR and LGI1) identified in the remainder (76%). New sleep complaints were reported in 18/25 (72%) of patients AE in this cohort, including dream enactment (24%, 6/25), insomnia (20%, 5/25), hypersomnia (16%, 4/25), other parasomnias (16%, 4/25), and sleep/ wake confusional states (8%, 2/25). PSG data showed poor sleep efficiency (55%) relative to healthy populations. Sleep disordered breathing was diagnosed in 32% (8/25).

Conclusion: Sleep disturbances were prevalent in this retrospective cohort of AE patients, including dream enactment behavior, and poor sleep efficiency. This finding emphasizes the need to screen for sleep disorders in this population. Improved identification and treatment of sleep disorders may reduce morbidity associated with AE and affect long-term outcomes of these patients.

Support: Patient recruitment and data collection was supported through and American Brain Foundation Clinical Research Training Fellowship (GSD).

Bonnette, Michael

Risk Factors for Acute Care Transfers in the Inpatient Rehabilitation Setting: A Review of the Literature

Abstract:

Objective: Acute care transfers (ACTs) are common in inpatient rehabilitation settings. Our aim was to analyze the current literature to identify common characteristics and risk factors that lead to ACTs and appraise the Early Warning Systems (EWS) literature with the eventual goal of constructing a risk assessment tool for Acute Care Transfer clinicians.

Design: We reviewed published literature searching through databases Ovid, Medline, Embase, Scopus, and Web of Science "acute care transfers", "unplanned transfers", "inpatient rehabilitation"", "risk assessment tools" and "early warning systems." We came across a total 338 articles of which 11 articles fit our inclusion criteria.

Setting: Inpatient rehabilitation facilities throughout the United States

Participants: None

Interventions: None

Main Outcome Measures: ACT rates, risk factors for acute transfers, use of risk assessment tools and early warning systems.

Results: Rehab patients with cancer diagnoses, SCI, complicated amputations, and low admission FIM scores are at a higher risk of being ACT'ed. Main reasons for ACT included infection, DVTs, and cardiac arrhythmias

Conclusions: Important to be vigilant in patients with those certain diagnoses (SCI, TKA, neoplasms) and be aware of potential complications

Level of Evidence: Systemic Review of Descriptive and Qualitative Studies

Creighton, Andrew

Sacral Pedicle Stress Fracture in an African-American Male, Adolescent, Competitive Basketball and Track and Field Athlete with a Prior Femoral Physeal Injury: A Case Report

Andrew Creighton, DO, Gregory Holtzman, PT, DPT, Craig Ziegler, MD, Heidi Prather, DO

Abstract:

Setting: Tertiary university orthopedic department

Patient: 15-year-old African American male basketball and track & field athlete with left-sided low back pain (LBP)

Case Description: The patient's pain started five months prior, but worsened four months prior when he participated in a relay race. While reaching out with his right arm to grab a baton pass, the patient's left knee hit the ground with his knee firmly extended and he felt a sudden, jarring and painful sensation in his left lower back. Pain was 8/10 at its worst, and located in the left PSIS region. Pain was better with ibuprofen, heat and ice applications, and worse with flexion, extension, running and jumping. After multiple rounds of physical therapy (PT), the patient experienced minimal relief. The patient had been on relative rest for three months prior to the visit. Past medical history was significant for a left distal femur physeal injury a year and a half prior, requiring multiple months of immobilization and crutches. Since this injury, the patient had grown about three inches. Additionally, at the time of onset of LBP, the patient was diagnosed with an estimated 15 degree scoliotic curve. Physical exam was significant for a lowered left iliac crest and left PSIS when compared to the right, positive standing flexion test on the left, positive FABER test bilaterally reproducing posterior pelvic pain, and left leg length 1.5cm shorter than the right. MRI revealed a small non-displaced extra-articular stress fracture at the left S1 pedicle. Vitamin D level was found to be low normal at 33 (normal range 30-100). The patient was restricted from sports participation for eight weeks, sent to PT for stabilization exercises, and prescribed an orthotic for leg length discrepancy.

Assessment/Results: At the eight-week mark, the patient had no pain with lumbar extension and was able to hop and skip flat-footed without pain. Vitamin D had been supplemented. The patient was referred to a physical therapist who specializes in running biomechanics.

Discussion: Sacral stress fractures are being reported with increasing frequency in literature; however, the incidence of these fractures in patients with leg length discrepancies is unknown.

Conclusion: A sacral pedicle stress fracture can be an unusual source of unilateral low back pain in a patient with a leg length discrepancy.

Decker, Gregory

Hip Radiograph Findings in Patients with Posterior Pelvic Pain

Gregory Decker MD, Michael Bonnette MD, Heidi Prather DO

Background: Posterior pelvic pain often presents as a diagnostic treatment dilemma for healthcare providers as it includes disorders of the lumbar spine, hip, and both intra-articular and peri-articular structures of the sacroiliac joint (SIJ).Little is understood about the links between pelvic girdle pain and hip motion and hip deformity prior to the onset of osteoarthritis.

Purpose: The purpose of this study was to describe the prevalence of radiographic hip deformity in patients who met the clinical diagnostic criteria for treatment of posterior pelvic pain with an image guided intra-articular sacroiliac joint injection.

Methods: A retrospective chart review found 148 adults 18-40 years of age who underwent an SIJ injection for refractory posterior pelvic pain in a tertiary university orthopedic setting. Nearly exclusively, 144/148 (97.3%) patients showed no radiographic findings of hip osteoarthritis. Radiographic measurements were completed to assess hip deformity including femoral acetabular impingement (FAI), developmental hip dysplasia (DDH), and acetabular retroversion. A measurement consistent with (FAI) was found in 26.5% (α angle $\geq 55°$) to 3.0% (LCE angle $>39°$) of patients. Findings indicative of DDH ranged from 20.4% (LCE angle $<20°$) to 12.3% (ACE angle $<20°$) of patients. Findings representative of acetabular retroversion were found in 42.6% (crossover sign) and 39.2% (prominent ischial spine) of patients.

Conclusion: Approximately 40% of adult patients under the age of 40 with posterior pelvic pain and signs and symptoms of SIJ pain were found to have acetabular retroversion. This is a significantly higher percentage than the 5-6% reported in asymptomatic people. Acetabular retroversion affects load transmission across the hip and pelvis which may provide a link to posterior pelvic pain and the hip not previously realized. This is the first study to investigate hip radiograph measurements to assess hip deformity without moderate or greater degenerative changes of the hip in patients seeking treatment for posterior pelvic pain.

Drace, Amelia

aEEG Background Indicates Seizure Burden, MRI Abnormality, and 2-Year Developmental Outcomes in Cooled Term Encephalopathic Neonates

To identify predictive value of aEEG background for seizure burden, MRI Injury Global Score (GS), and later development, we studied 44 term infants with moderate/severe HIE undergoing therapeutic hypothermia with cEEG for 96 hours and scored their aEEG backgrounds hourly.

Any Flat-Inactivity during the full study predicted increased mean seizure burden (515.2 vs. 32.86 minutes, p=0.011) and MRI injury (GS 131.8 vs 54.03, p=0.036). In the first 24 hours, appearance of Discontinuity was associated with decreased seizure burden (28.76 vs. 204.35 min, p=0.024) and favorable MRI abnormality score (51.21 vs. 77, p=0.031). Over the full study, Continuity was associated with decreased seizure burden (39.12 vs. 150.77, p=0.048), and favorable MRI (52.23 vs. 71.21, p=0.048). A stepwise linear regression of seizure burden showed independent contributions by percentage of total time spent in Flat-Inactivity (B=420.959, p<0.001) and Low-Burst-Suppression (B=866.049, p<0.001). Flat-Inactive percentage over the whole study was the sole independent predictor of MRI global score (B=143.9, p<0.001).

On 2-year follow-up, subjects who had a predominantly continuous aEEG had higher mean Bailey scores in cognitive (p=0.025), language (p=0.001), and motor (p=0.002) domains versus other background patterns. No subject with a continuous aEEG had a Bailey score showing moderate or severe delay (Chi2 = 26.5, p=0.001)

Severely abnormal aEEG background patterns are more closely associated with poor short-term outcomes while normal backgrounds are better predictors of favorable long-term outcome.

Eby, Brendan

Does Changing Technique During Mechanical Thrombectomy Improve Recanalization Rates?

Abstract:

Introduction: In mechanical thrombectomy for acute ischemic stroke, recanalization of the occluded vessel correlates with improved outcomes. When initial recanalization attempts fail, it is unclear whether introducing another technique (rescue therapy) might confer an advantage over continuing the same technique. This study aims to investigate the effect of a mid-procedure technique change on recanalization rates during mechanical thrombectomy.

Methods: Retrospective chart review was performed on all mechanical thrombectomies performed at a single tertiary care academic hospital between January 2013 and October 2016. Cases were grouped by initial technique (stent retriever, aspiration catheter). All cases with more than one pass attempt were included, evaluated for whether a mid-procedure technique change was employed, and subsequently analyzed.

Results: 53 cases were identified for analysis. Technique change was utilized in 6 of 29 (21%) stent retriever cases and 15 of 24 (63%) aspiration catheter cases (p=0.004). In the stent retriever group, recanalization was achieved in 4 of 6 (67%) cases utilizing a technique change, compared with 14 of 23 (61%) without technique change (p=1.00). In the aspiration catheter group, recanalization was achieved in 10 of 15 (67%) cases utilizing a technique change, compared with 6 of 9 (67%) without change (p=1.00).

Conclusion: Among thrombectomy cases who fail first pass recanalization, final recanalization rates are comparable between cases with or without a mid-procedure technique change, regardless of initial technique choice.

Giles, James

Caring for Neuromyelitis Optica Patients in Yangon, Myanmar

Giles JA, Silbermann E, Lin HH, Mar S, Sein MMA.

Background: Neuromyelitis optica (NMO) makes up a larger proportion of demyelinating disease in Asian populations, yet data is lacking on NMO in Southeast Asia. Myanmar is a lower-middle income country with a population of 55 million. Most of the population has no private medical insurance and must pay out-of-pocket for medical diagnosis and treatment, where this is not funded by the government. The Yangon NMO Registry was established based on the clinical observation of high incidence of NMO in Myanmar. Here we present the data from the first six months of the registry, and to discuss the current clinical practice caring for these patients.

Method: All consecutive cases presenting to the Neurology Unit at Yangon General Hospital (YGH) with longitudinally extensive transverse myelitis (LETM) and/or bilateral optic neuritis (ON), or those with a known diagnosis of NMO were added prospectively to the registry. Data included demographics, disease presentation, laboratory and imaging findings, and details of immunosuppressive therapy. Disability was measured on admission via the Extended Disability Status Scale (EDSS).

Results: 19 cases were identified in the first six months. Median age of onset was 35 (range 16-55) and 84.2% of patients were women. 89.4% of patients presented with LETM, and 47.3% had concomitant ON. Average hospital stay was 30 days. The mean EDSS at the time of initial diagnosis was 8.3. While 84% of NMO patients received spine imaging, only 52.6% received MRI brain. 81.2% of patients had their diagnosis confirmed with NMO antibody testing (AQP4-IgG). 68% of patients were treated with azathioprine (AZ) while 31.6% received mycophenolate mofetil (MMF).

Conclusion: This is the first study describing the care of NMO patients in Myanmar. Most patients presented highly symptomatic from spinal cord pathology. Due to the high financial cost and long waiting time, most patients receive partial diagnostic assessment; hence the diagnosis of NMO is made via differing combination of clinical criteria, serological testing and imaging. Patients are preferentially treated with AZ over MMF due to cost. This registry provides insight into providing care for NMO patients in a resource-limited setting.

Kim, Albert

Defining dorsal and ventral subthalamic nucleus stimulation networks via resting state functional connectivity

Albert Kim, Aimee Morris, Mikhail Milchenko, Aaron Tannenbaum, Joel Perlmutter, Scott Norris

Objective: To determine the networks corresponding to dorsal and ventral subthalamic nucleus (STN) deep brain stimulation (DBS) sites in Parkinson's disease (PD)

Background: Deep brain stimulation to the bilateral subthalamic nucleus (STN DBS) improves symptoms in advanced PD, but the mechanism of action remains unclear. Furthermore, there is substantial inter-individual STN DBS-mediated motor variability. Anatomical heterogeneity may be to blame, as in vivo animal tracers and recent advanced imaging have suggested distinct regions within the STN that may subserve different functions. Here, we seek to define the networks affected by dorsal and ventral STN stimulation, and correlate with DBS stimulation effects of motor tasks.

Design / Methods: 133 patients with PD received resting-state functional connectivity MRI scans (rsfcMRI) prior to STN DBS placement. The rsfcMRI scans underwent registration to Talariach atlas space and validated processing steps to negate the effects of various confounders (i.e. head motion, CSF, white matter) on BOLD signal. Dorsal and ventral contact localization was determined using structural MRI at time of surgery and post-operative head CT. A seed-based analysis was performed to define networks correlating with dorsal and ventral STN stimulation. Dorsal-STN minus ventral-STN difference correlation maps were computed for each subject. Dorsal-STN versus ventral-STN differences were examined using a one-sample t-test on subject difference maps. Statistically significant voxels, accounting for multiple comparisons, were computed on a cluster-wise basis using a voxel-wise $p < 0.001$, followed by $p < 0.05$ family-wise error correction. Z-score thresholds were computed by Monte Carlo permutation (n=10,000 iterations) analysis.

Results: On both the left and right electrode, the dorsal STN had significantly increased functional connectivity bilaterally within the globus pallidus and thalamus as compared to the ventral STN. On the left electrode, there was increased connectivity to the right frontal lobe and left insular cortex.

Discussion: This finding is in agreement with known tractography from the dorsal STN to basal ganglia, but it is unclear why connectivity differences exist with respect to laterality. Additional analyses will be performed. We plan to correlate strength of these dorsal and ventral STN networks to UPDRS III data. Finally, we will run a two-sample t test comparing alterations in these networks with a cohort of age-matched normal controls. By identifying changes in connectivity for the disease state and correlating strength in network to motor outcomes, we hope to provide insight into the neural control of gait and balance and perhaps an optimal stimulation site for improving gait and balance in patients with PD.

Laurido-Soto, Osvaldo

Systematic Review of AMPA-R Encephalitis: An attempt at Defining an Illness

Abstract:

Objective: To review the literature available to report on the clinical features, comorbidities, and outcomes of patients with antibodies to the alpha-amino-3-hydroxy-5-methyl-4-isoxazolepropionic acid receptor (AMPAR). In addition to adding two patients from our institution to this repertoire.

Methods: A retrospective review of patients diagnosed with AMPAR encephalitis proven by commercially available assays was performed following the preferred reporting items for systematic reviews and meta-analyses guidelines (PRISMA).

Results: 260 papers met search criteria which was further narrowed to 22 papers that met inclusion criteria. It was found that only 69 patients have been reported in the literature as of this moment and we added two internal patients, for a total of 71 patients. M=21, F=47 with an average age of 51.2 for which the average MRS at diagnosis was 3.93 and at last day of f/u 1.76. The first symptoms were: Confusion 57%, Limbic encephalitis 34%, and convulsions 31%, usually with most patients having an average of 4 neurological symptoms. From a diagnostic standpoint – 87% of patients with bMRI abnormalities had temporal lobe involvement and 67% had abnormalities in their CSF characterized primarily by pleocytosis. Of the patients for which the information was available 70% had some sort of tumor and 98% of patients received some sort of immunotherapy.

Conclusion: AMPAR encephalitis is a heterogeneous disease with multiple potential phenotypes. It appears that the majority will have a limbic encephalitis phenomenology with a reactive CSF, but this is not ubiquitous. Further analyses to elucidate associations with outcomes are currently pending.

Long, Justin

Sensitivity and Specificity of CSF VZV Antibody and PCR Testing in Suspected VZV Vasculopathy

Justin Long, James Giles, Peter Kang, Laura Baldassari, Kristen Gehrking, Lynn Zhang, Carey-Ann Burnham, Craig Wilen, Beau Ances, Robert Bucelli

Background: Vasculopathy due to varicella zoster virus (VZV) infection is a secondary cause of stroke in patients with zoster reactivation, with or without rash. A limited series of previous studies in patients with VZV vasculopathy have demonstrated a significantly increased sensitivity when testing for the presence of CSF anti-VZV IgG as compared to CSF VZV DNA via PCR. However, there is limited independent verification of this finding and lack of evidence as to the specificity of anti-VZV IgG antibodies in CSF of patients without VZV, most crucially in those with strokes not related to VZV infection.

Goal: To assess the sensitivity and specificity of CSF anti-VZV IgG and CSF VZV DNA testing by PCR, both in isolation and in combination, in patients with suspected VZV vasculopathy.

Methods: We prospectively identified and collected blood and CSF specimens from 10 control subjects without stroke, 20 disease control subjects with stroke unrelated to VZV infection and 9 subjects with stroke due to suspected VZV vasculopathy (i.e. stroke in the setting of active or recent history of zoster rash or VZV meningoencephalitis). Prior to CSF analysis, patients were assigned to these groups on clinical grounds used pre-defined criteria. Specimens were analyzed for VZV DNA copies by PCR and anti-VZV IgG and IgM levels.

Results: Among 9 patients with suspected VZV vasculopathy, 4 (44%) had CSF VZV DNA and 6 (67%) had CSF anti-VZV IgG. Among 20 patients with stroke due to other causes, 0 patients had CSF VZV DNA and 1 had CSF anti-VZV IgG. Among 10 patients without stroke, no patient had CSF VZV DNA or anti-VZV IgG. For presence of CSF VZV DNA alone, sensitivity/specificity for detection of VZV vasculopathy was 44%/100%. For presence of CSF VZV anti-IgG alone, sensitivity/specificity was 67%/97%. For presence of either CSF VZV DNA or anti-IgG, sensitivity/specificity was 78%/97%. The difference in sensitivity for presence of CSF VZV DNA alone as compared to presence of either CSF VZV DNA or anti-IgG was not statistically significant by McNemar test (p=0.2482), although sample size for this comparison was quite small (n=9).

Conclusions: The presence of either CSF VZV DNA or CSF VZV anti-IgG is highly specific for VZV vasculopathy. Sensitivity is improved by testing for both VZV PCR and anti-IgG, although sample size was too small in this study to demonstrate statistical significance.

Study supported by: This work was supported, in part, by the Paula C and Rodger O Riney Fund

Majmudar, Bittu

NMNAT2 Effects on Neuronal Survival and Learning in Immature Neurons and Brain

Majmudar, B. and Galindo, R.
Department of Neurology, Division of Pediatric & Developmental Neurology
Washington University in St. Louis

Background: Nicotinamide mononucleotide adenylyl transferase 2 (NMNAT2) is an NAD-synthetizing enzyme widely abundant in the brain and thought to be important in the survival of peripheral neurons and axons by delaying Wallerian degeneration. NMNAT2 knockout mice do not survive and have decreased amount of peripheral nervous system neurons and axons. Alternatively, NMNAT overexpression provides neuroprotection against various stress-related and toxic insults in peripheral nerves. However, little is known regarding the role of NMNAT2 in the developing brain specifically in response to cerebral injury. Here, we hypothesize that NMNAT2 downregulation promotes the degeneration of immature developing neurons following NMDA-mediated excitotoxicity in vitro and after neonatal cerebral hypoxia-ischemia (HI) in vivo.

Methods: Cultured immature cortical and hippocampal neurons were transiently exposed to the NMDA-agonist ibotenic acid in the presence or absence of an NMNAT2-reducing or NMNAT2-synthetizing AAV8 virus. Neuronal survival was then evaluated by MAP2+ neurite staining and by the lactate dehydrogenase cell death assay. To evaluate the potential neurodegenerative effects of NMNAT2 depletion in the neonatal injured brain, we utilized two different NMNAT2 depletion approaches: (1) NMNAT2-depleted heterozygote (HET) and wild-type (WT) mice (N2-Blad mice) were subjected to the Rice-Vannuci model of neonatal HI at near term-equivalent age followed by volumetric histochemical analysis of cerebral injury 7 days after HI, (2) Newborn postnatal day 0 N2-blad WT and HET mice were injected with the AAV8-NMNAT2-depleting virus and then subjected to HI injury 7 days later followed by histochemical analysis of injury 7 and 14 days after HI. Lastly, we examined the potential effect of NMNAT2 depletion on hippocampal-mediated learning utilizing the Barnes Maze in SHAM-operated and neonatal HI-exposed NMNAT2-depleted HET and WT mice.

Results: Depletion of NMNAT2 resulted in the progressive degeneration of immature cortical and hippocampal neurons even in the absence of excitotoxic injury. NMDA-mediated excitotoxicity further promoted neuronal degeneration of NMNAT2-depleted neurons. In contrast, upregulation of NMNAT2 appeared to favor neuronal survival and was able to decrease the pro-degenerative effects of AAV8-mediated NMNAT2 knockdown. Heterozygote mice containing half of the native amount of NMNAT2 demonstrated a trend towards an increase in cerebral degeneration following neonatal HI (HET, N=58; WT, N=39; % degeneration for hippocampus = 32±4.6% HET vs. 26±5% WT, p=0.19; striatum = 38±10% HET vs. 21±4% WT, p=0.15; cortex = 19±6% HET vs. 12±7% WT, p=0.22). Interestingly, despite the absence of a statistically significant increase in cerebral neurodegeneration in the NMNAT2-heterozygote Blad mice, SHAM and HI-induced N2-Blad HET mice demonstrated a decrease in learning when exposed to the Barnes Maze task when compared to WT mice. Quantitation of the neurodegenerative effects in the N2-Blad injured and SHAM brains exposed to the AAV8-NMNAT2 downregulating virus is currently ongoing.

Conclusions: Depletion of NMNAT2 promotes the degeneration of naïve and injured cortical and hippocampal developing neurons in vitro. In contrast, exogenous upregulation of NMNAT2 favors in vitro neuronal survival and appears to rescue the degeneration caused by NMNAT2 downregulation. HET mice with 50% NMNAT2 levels when compared to WT mice did not demonstrate a statistically-significant increased susceptibility to cerebral tissue loss after hypoxia-ischemia. This could potentially indicate that cerebral NMNAT2 levels need to be decreased further to produce a neurodegenerative effect. Thus, a

Majmudar, Bittu - continued

group of HET mice were injected with a viral vector to see if further decreasing NMNAT2 levels by 80-90% produces significant neuronal loss after hypoxia-ischemia. This data is currently in the process of analysis. Even if there is no difference in volume noted between WT and HET mice exposed to hypoxia-ischemia, it is possible that there are still neurobehavioral differences in these animals with respect to learning. Thus, behavioral testing of WT and HET animals exposed to hypoxia-ischemia was conducted using the Barnes Maze. Preliminary results show that HET mice in general do not show the expected learning trend compared to WT mice, regardless of exposure to HI. This suggests that decreased NMNAT2 levels potentially negatively impact learning. However, further detailed work with a larger cohort of mice will need to be done to confirm the above findings.

Obretenova, Souzana

Visual Hallucinations and Field Deficit Associated with Non-ketotic Hyperglycemia

Abstract:

Introduction: Focal seizures are often seen in non-ketotic hyperglycemia, but may present atypically and delay diagnosis and appropriate management. Occipital seizures have been reported to present with visual hallucinations and post-ictal hemianopia. These cases are associated with MR imaging findings of FLAIR white matter hypointensity rather than hyperintesinty in occipital regions.

Case: A 51 year old man presented with 5 days of recurrent visual hallucinations followed by left homonymous hemianopia in the setting of non-ketotic hyperglycemia. Symptoms correlated with electrographic seizures originating in the right posterior region and did not resolve within 24 hours after improved glycemic control. The patient demonstrated white matter FLAIR hypointensity in the right occipital region on MRI, which correlated with the electrographic origin of his seizures. Symptoms resolved after initiation of carbamazepine.

Conclusion: It is important to recognize atypical focal seizure symptoms in patients with non-ketotic hyperglycemia. Visual hallucinations and subsequent hemianopia have been described in rare case reports, and appear to correlate with FLAIR hypointensity in occipital regions that may be missed on initial radiologic evaluation. Treatment with glycemic control and an antiepileptic agent leads to full neurologic recovery.

Patel, Meera

Characterization of the Sleep Disorders of Children with Cerebral Palsy

Meera S. Patel, M.D. and Amy Licis, M.D.

Background: Children with developmental disorders such as cerebral palsy often have sleep disorders that have not been well-characterized. Prior studies have shown that children with cerebral palsy have a higher prevalence of sleep disorders compared with the general population. The most frequently identified problems were disorders of initiation and maintenance of sleep, of sleep-wake transition, of sleep breathing, of excessive daytime somnolence, and of arousal. While prior studies have characterized sleep disorders in children with cerebral palsy, few have compared sleep disorders in children with cerebral palsy to a control group. Using the Sleep Disturbances Scale for Children (SDSC) questionnaire, this study aims to characterize the sleep disorders in children with cerebral palsy and the sleep disorders in the siblings of children with cerebral palsy. Additionally, this study seeks to compare the SDSC questionnaire scores between these two groups.

Methods: This was a prospective study with one-time questionnaire administration. Participants ages 2 to 17 years old were recruited from the Washington University Department of Neurology Cerebral Palsy Center at Saint Louis Children's Hospital and assessed using the SDSC. One hundred and eighty nine children with completed SDSC questionnaires were included in analysis (109 children with cerebral palsy and 80 siblings of children with cerebral palsy). Mann Whitney nonparametric T-tests were performed to assess whether there was a statistically significant difference in the means of the scores on the SDSC between the children with cerebral palsy and their siblings.

Results: A pathological total sleep score on the SDSC was found in 26% of children with cerebral palsy compared with 11% of siblings of children with cerebral palsy. Children with cerebral palsy had significantly higher total sleep scores than their siblings without cerebral palsy, as well as significantly higher sleep scores in disorders of initiation and maintenance of sleep, disorders of sleep-wake transition, disorders of sleep breathing, and disorders of excessive daytime somnolence.

Conclusions: Sleep disorders are more common in children with cerebral palsy than in their siblings without cerebral palsy. Better characterization of the sleep disorders in children with cerebral palsy may be an initial step toward implementing therapies and improving quality of life.

Robison, Leah

Acute motor and sensory axonal neuropathy: a unique complication of DRESS syndrome

Drug rash with eosinophilia and system symptoms (DRESS) syndrome and Guillain-Barre Syndrome (GBS) are both relatively uncommon diseases with an immunologically mediated pathogenesis. We present a case of both diseases overlapping in the same patient. Neurological involvement of DRESS overall is considered to be rare, although central nervous system involvement has been well-described in the literature. However, acute peripheral nervous system involvement of DRESS is not well-described. To the best of our knowledge, there have been only two prior descriptions of GBS in the setting of a drug hypersensitivity reaction [1, 2], and one case of a peripheral neuropathy not otherwise defined [3]. Herein, we describe a case of definite DRESS with GBS representing acute peripheral nervous system involvement of this multi-system disease. The purpose of this poster is to raise awareness of the possible neurologic complications in the acute phase of DRESS syndrome.

Rudock, Robert

Comparison of Home Sleep Apnea Testing and In-laboratory Polysomnography at a Single Academic Outpatient Sleep Center

Introduction: Home sleep apnea testing (HSAT) is being utilized more often to screen for and diagnose obstructive sleep apnea (OSA). While this approach has many advantages (patient convenience, access, potential cost-effectiveness), in-laboratory attended polysomnography (PSG) remains the gold standard for diagnosing sleep disordered breathing. Recognizing the limitations of HSAT, such as false positive rates and grading of OSA severity, is paramount to a sleep provider's ability to correctly interpret results and recommend appropriate treatment options.

Objectives: To determine the accuracy of ambulatory (type III) home sleep studies compared to in-laboratory polysomnography at a single academic outpatient sleep center.

Methods: At risk patients for sleep disordered breathing, who had an ambulatory (type III) home sleep study with a respiratory event index (REI) between 5.0 and 10.0, followed by an in-laboratory diagnostic polysomnogram were identified. The REI from the home sleep study was then compared to the RDI (respiratory disturbance index) from the polysomnogram. Patient demographics were also analyzed to ascertain patient populations who may be more likely to have inaccurate HSAT results.

Results: Of 31 patients who were identified as having a REI between 5 and 10 respiratory events per hour, followed by an in-laboratory polysomnogram, 21 (68%) were confirmed to have OSA (6 mild; 10 moderate; 5 severe). 10 of 31 patients did not have OSA, representing a potential false positive rate of 32%. Additionally, male sex and BMI ≥30 kg/m², were associated with higher positive predictive values (0.71 and 0.75) compared to female sex and BMI <30k kg/m² (0.6 and 0.64).

Conclusion: HSAT offers patients who are at risk for OSA several potential benefits compared to traditional PSG. However, HSAT resulting in a REI between 5.0 and 10.0, can be falsely positive in 32% of patients leading to incorrect diagnosis and costly, unneeded treatments. Therefore, a dedicated in-laboratory attended PSG in patients who have a REI of 5.0 to 10.0 respiratory events per hour is strongly recommended.

Smith, Sean

Case Diagnosis: Autonomic dysreflexia due to stercoral colitis in a tetraplegic patient

Case Description: A 34-year-old man with a history of incomplete C4 ASIA Impairment Scale B tetraplegia presented with transient episodes of autonomic dysreflexia (AD) over a 12-hour period. Initial episodes resolved with repositioning, bladder and bowel management per protocol. However, a noxious stimulus source was unable to be identified in the last episode, and the patient required pharmacologic management for dangerously high blood pressure. Further evaluation with CT chest/abdomen/pelvis was performed. Abdominal CT scan was consistent with developing stercoral colitis. Stercoral colitis is an uncommon but important complication of fecal impaction. It is an inflammatory process caused by increased intraluminal pressures that is associated with high morbidity and mortality.

Discussions: This case highlights the evaluation and management of AD. Spinal cord injury patients are at increased risk for fecal impaction, so stercoral colitis may develop. This has the potential to trigger AD. If stercoral colitis is not diagnosed and treated promptly, bowel ulceration and perforation may also occur, leading to peritonitis, sepsis, and death. To our knowledge, this is the first reported case of AD due to stercoral colitis in an individual with spinal cord injury.

Conclusions: Autonomic dysreflexia (AD) is a potentially life-threatening condition that may occur in spinal cord injury patients with lesions above T6. When the underlying cause for AD is elusive and the episodes are persistent or recurrent, further investigation, including imaging, is warranted to identify other possible etiologies. This case identified stercoral colitis as a cause of AD in an incomplete C4 tetraplegic patient.

Smith, Sean

Assessment of Compassion Fatigue in Physical Medicine and Rehabilitation Residents

Abstract: The problems of burnout and depression in the medical profession have been well established in recent years with a focus on resident wellness programs to address these issues. Compassion is a way to relate to suffering and an attempt to relieve suffering. Self-compassion has been shown to be significantly correlated with positive mental health. We aimed to identify the current state of compassion satisfaction and compassion fatigue among resident physicians in a Physical Medicine and Rehabilitation (PM&R) Program. Method: We distributed the self-administered Professional Quality of Life Scale (PQLS) questionnaire and analyzed each category on compassion fatigue (burnout, secondary traumatic stress) and compassion satisfaction. Analyses: Data was scored according to the PQLS scales for compassion satisfaction, burnout, and secondary traumatic stress and analyzed. Means, standard deviations and ranges were calculated for each category. Results: Descriptive statistics revealed the residents were moderate to low for all categories. The mean compassion satisfaction score was 33.9 ± 4.56 corresponding to an average level of compassion satisfaction. No individual resident scored a high level of compassion satisfaction. Conclusion: Compassion satisfaction was moderate to low for PM&R residents. Residents may benefit from a compassion training module as compassion has been shown to improve patient outcomes and increase physician well-being.

Stunkel, Leanne

Evaluating the utility of a post-processing algorithm for MRI evaluation of optic neuritis

Leanne Stunkel1, Aseem Sharma2, Matthew S. Parsons2, Amber Salter3, Gregory P. Van Stavern4
1. Department of Neurology, Washington University in St. Louis School of Medicine, St. Louis, MO.
Mallinckrodt Institute of Radiology, Washington University in St. Louis School of Medicine, St. Louis, MO.
Division of Biostatistics, Washington University in St. Louis School of Medicine, St. Louis, MO.
Department of Ophthalmology and Visual Sciences, Washington University in St. Louis School of Medicine, St. Louis, MO

DISCLOSURES: Dr. Sharma is co-inventor of algorithms used to process images in this study and has a financial [ownership] interest in Correlative Enhancement LLC. He did not participate in patient selection, image analysis, or statistical analysis.

INTRODUCTION: MRI evaluation of the optic nerve requires comparison of the nerve's signal intensity with that of normal white matter, but the different visual environments of the optic nerve and the white matter make it difficult for the human eye to compare their signal intensities. Correlative Image Enhancement (CIE) is a post-processing algorithm aims to improve conspicuity of a structural detail of interest on existing MRI scans by increasing its contrast-to-noise ratio.

METHODS: Retrospective study of 44 patients (88 eyes) who underwent MRI for clinical suspicion of optic neuritis. 31 eyes diagnosed with optic neuritis and 28 eyes without clinical evidence for optic neuritis were analyzed. 29 asymptomatic fellow eyes were not analyzed. Coronal FLAIR and contrast-enhanced MRI images were processed using CIE. Contrast-to-noise ratio (CNR) between optic nerves and ipsilateral white matter on both FLAIR and contrast-enhanced images was calculated for baseline and processed images using identical regions of interest. Baseline and processed images were reviewed by 6 masked readers to assess FLAIR signal intensity of the optic nerves and presence of contrast enhancement.

RESULTS: For FLAIR images of eyes with optic neuritis, processing resulted in an increase in median CNR from 17.8 to 85.0 ($p<0.0001$), with increased CNR seen in 27/28 eyes. CNR for control eyes was not significantly affected ($p=0.0821$), with CNR increase noted only in 1/28 control eyes. For contrast-enhanced images of eyes with optic neuritis, processing resulted in an increase in median CNR from 19.35 to 93.7 ($p<0.0001$), with increased CNR seen in 24/28 eyes. CNR for control eyes was not affected ($p=0.125$), with CNR increase noted only in 1/18 control eyes.

CIE processing improved sensitivity for most readers for both FLAIR and contrast-enhanced images. CIE processing did not worsen specificity for most readers. CIE improved sensitivity for detection of optic neuritis for most readers.

CONCLUSIONS: CIE increased CNR in eyes with optic neuritis, and improved sensitivity for detection of optic neuritis, and did not affect CNR for eyes without optic neuritis or worsen specificity.

Younce, John

A literature review and case series of ziprasidone for psychosis in Parkinson disease

John R Younce, Albert A Davis, Kevin J Black

The atypical antipsychotic ziprasidone has been considered inappropriate for use in patients with Parkinson disease, as most atypical antipsychotics worsen parkinsonian severity. However, this opinion has been based on very limited clinical experience. Here we review published experience with ziprasidone for treating psychosis in Parkinson disease, and add 7 cases from our center of ziprasidone exposure in patients with idiopathic parkinsonism. We conclude that, although ziprasidone occasionally can produce substantial worsening of motor signs, it usually is well tolerated, and may provide in some cases a useful alternative to better proven treatment particularly in the acute care setting. Further prospective controlled studies are needed.