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Case report: listeria rhombencephalitis in a healthy 64 year old woman

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Background: Listeria rhombencephalitis is a rare and serious complication of Listeria monocytogenes infection. We present a case of presumed Listeria rhombencephalitis with dramatic recovery from a highly morbid state. Methods: A previously healthy 64 year old woman with a remote and stable history of a major depressive episode and no history to suggest immune compromise presented with nausea and vomiting followed by the acute onset of diplopia and gait disturbance 28 days after exposure to an identified infectious source of spring rolls and 21 days after a severe diarrheal illness from that exposure. Our patient was evaluated by emergency physicians and general internists over a period of 1 week after the onset of diplopia and gait disturbance and given a diagnosis of serotonin syndrome before receiving a consultation from Neurology. Her presentation featured a deep encephalopathy and an unusual hyperkinetic movement disorder with startle myoclonus, palatal myoclonus and diffuse tremor. Results: Her MRI scan showed FLAIR hyperintensities in the bilateral cerebellum and pons with adjacent pial enhancement, characteristic of Listeria rhombencephalitis. Her CSF showed a lymphocytic pleocytosis with normal chemistry. Conclusions: She recovered dramatically to treatment with IV ampicillin. This case report illustrates the importance of considering Listeria rhombencephalitis in immunocompetent patients with brainstem symptoms following a diarrheal illness.

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Causes of albuminocytologic dissociation and the impact of an age-adjusted reference limit on review of 2,627 CSF samples

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Background: We set out to test the discriminative power of an age-adjusted upper reference limit (URL) for CSF total protein (CSF-TP) in identifying pathological causes of albuminocytologic dissociation (ACD). Methods: We reviewed the charts of 2,627 adult patients who underwent a lumbar puncture at a tertiary care center over a 20-year period. Samples with CSF-TP above 45 mg/dL (0.45 g/L) were included. Samples with white blood cell count > 5×10^9 /L, red blood cell count > 50×109/L, and glucose < 2.5 mmol/L (45 mg/ dL) were excluded. Patients with CSF-TP elevated above 45 mg/dL were considered to have 'pseudo' albuminocytologic dissociation (ACD) or 'true' ACD if their CSF-TP was in excess of age-adjusted norms. Results: Among all patients with ACD, a pathological source of CSF-TP elevation was identified in 57% (1490/2627) of cases, 51% of those with 'pseudo' ACD, and 75% with 'true' ACD (p< 0.001). Use of an age-adjusted upper reference limit favored the detection of polyneuropathy patients (13.5% proportionate increase) and excluded a larger number of patients with isolated headache

(10.7% proportionate decrease; p < 0.0001). **Conclusions:** Elevated CSF-TP is a relatively common finding. Use of age-adjusted upper reference limits for CSF-TP values improve diagnostic specificity and help to avoid over-diagnosis of ACD.

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Effects of hypoglycemia on sensitive brain structures in a patient with an insulinoma

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Background: A previously healthy 26 year-old male presented with confusion and recurrent hypoglycemia (blood glucose lows of 2.5 mmol/L) while on vacation in Las Vegas. He denied substance or heavy alcohol use and the toxicology screen was negative. He was transferred home to Winnipeg for further care and was found to have only patchy memories of his trip and the days leading up to the trip, consistent with mixed anterograde and retrograde amnesia. MoCA score at presentation was 16/30 with points lost on orientation, delayed recall and visuospatial-executive tasks. MRI revealed T2 hyperintensities and diffusion abnormalities in bilateral hippocampi and globus pallidi. Electroencephalography showed triphasic waves. The patient was found to have a pancreatic insulinoma, which was surgically resected. In follow-up nine weeks later he was near his cognitive baseline, though he had ongoing difficulties with delayed recall. Repeat MRI showed improvement but not resolution of hippocampal and pallidal signal change, with mild hippocampal atrophy.

Neuropathological and animal studies have shown that structures most sensitive to hypoglycemic neural injury include the hippocampus, basal ganglia, and neocortex. The clinical and radiographic findings in this case illustrate an unusual presentation of insulinoma and the effects of hypoglycemia on the brain. **Methods:** N/A **Results:** N/A **Conclusions:** N/A

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Initial validation of symptom scores derived from the Orthostatic Discriminant and Severity Scale

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Background: To develop a scale to quantify and discriminate orthostatic from non-orthostatic symptoms. We present initial validation and reliability of orthostatic and non-orthostatic symptom scores taken from the Orthostatic Discriminate and Severity Scale (ODSS). Methods: Validity and reliability were assessed in participants with and without orthostatic intolerance. Convergent validity was assessed by correlating symptoms scores with previously validated tools (Autonomic Symptom Profile (ASP) and the Orthostatic Hypotension Questionnaire (OHQ)). Clinical validity was assessed by correlating scores against standardized autonomic testing. Test-retest reliability was calculated using an intra-class correlation coefficient. Results: Convergent Validity: Orthostatic (OS) and Non-Orthostatic (NS) Symptom Scores from 77 controls and 67 patients with orthostatic intolerance were highly correlated with both the Orthostatic Intolerance index of the ASP (OS:r=0.903;NS:r=0.651; p<0.001) and the OHQ: (OS:r=0.800;NS:r=0.574; p<0.001). Clinical Validity: Symptom Scores were significantly correlated with the blood pressure change during head-up tilt (OS:r=-0.445;NS:r=-0.354; p<0.001). Patients with orthostatic intolerance had significantly higher symptom scores compared to controls (OS:66.5±18.1 vs. 17.4±12.9; NS:19.9±11.3 vs. 10.2±6.8; p<0.001, respectively). Test-retest reliability: Both symptom scores were highly reliable (OS:r=0.956;NS:r=0.574, respectively; p<0.001) with an internal consistency of 0.978 and 0.729, respectively. Conclusions: Our initial results demonstrate that the ODSS is capable of producing valid and reliable Orthostatic and Non-Orthostatic Symptom Scores.

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Hospital readmission following neurology discharge: A systematic review

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Background: Unplanned hospital readmission is inconvenient for patients, puts them at risk of harm, and is a resource strain. We reviewed available literature on risk factors for readmission following discharge specifically from neurology inpatient services with a focus on factors unique to non-stroke neurology admissions. Methods: We conducted a systematic search using PRISMA methodology of MEDLINE, EMBASE, and CENTRAL databases up to January 1, 2018. Two independent reviewers screened articles for inclusion. English-language articles were included that identified factors related to hospital readmission after discharge from a neurology service. Admissions with stroke as the primary focus were excluded. Results: Of 9508 unique abstracts, 25 met inclusion criteria and were included for review. Multiple factors impacting probability of readmission were identified including age, living alone, history of nonepileptic seizure, length of stay, services consulted during hospital stay, hospital volume, and severity of illness. Conclusions: There are identifiable risk factors that influence likelihood of readmission to hospital following discharge from neurology inpatient services, although the non-stroke literature is sparse. There is a need for future prospective work to investigate modifiable risk factors and opportunities to reduce readmission rates and improve patient safety.

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Down Syndrome: robust neurophysiological perspectives

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Background: Down Syndrome (DS) has a mosaicism of presentations, but a number of common features. Cerebral evoked potentials (somatosensory, visual and auditory) can be higher in amplitude in DS. The aim of this study is to explore the value of the neurophysiological amplitude of three different modalities in DS individuals undergoing spinal surgery, or epilepsy evaluation. **Methods:** Standard procedure of EEG evaluation was conducted. We routinely monitor somatosensory (SSEP) and motor evoked potentials (MEP), using peripheral nerves stimulation and transcranial electrical stimulation during surgery. We report findings from 14 DS individuals age-matched to 14 individuals with idiopathic scoliosis **Results:** The amplitude of the SSEP is significantly higher in DS individuals than in age-matched controls using the same parameters.

SSEP;10.2±2.5µV vs 2.4±2.3µV (p<0.05, paired t-test). The threshold for eliciting MEPs was also significantly lower in DS in comparison to controls, 175±20V vs 629±100V, (p<0.05, paired t-test). Interictal EEG showed high amplitude spike and waves, and greater intracortical coherence in DS with epilepsy than non-DS patients Conclusions: Robust neurophysiological findings showed high amplitude sensory evoked potentials, low threshold motor evoked potentials, and high amplitude spikes and wave, all reflect a common process of increased neuronal synchronicity and oscillatory behaviour in Down Syndrome.

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Twice negative PCR in a patient with HSV-1 Encephalitis

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Case Description: A 64 year-old male presented with left-sided weakness and altered level of consciousness after a suspected seizure. MR Brain demonstrated right mesial temporal lobe diffusion restriction. Empiric antiviral and antibiotic treatments were initiated despite CSF negative for HSV/VZV and enteroviruses. Lumbar puncture on admission day five was unchanged and empiric treatments were discontinued. On day 13 he deteriorated into status epilepticus necessitating ICU transfer. A third lumbar puncture demonstrated elevated protein and HSV-1 positive PCR. Acyclovir was restarted with guarded prognosis. **Discussion:** Detection of HSV-1 in CSF is considered the diagnostic gold standard for HSV-1 encephalitis. The validated multiplex assay used in Alberta, Canada has a 95% level of detection significantly better than the recommended threshold for HSV laboratory diagnosis. Previous reports have indicated that CSF PCR may be negative early in the disease course. Others have suggested that initially negative/follow up positive HSV PCR cases may represent secondary reactivation or release from underlying tissue damage. Consideration of the full clinical picture is crucial in patients with HSV negative PCR. Continuation of antiviral therapy may be appropriate in select HSV PCR negative patients.

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Association of phantogeusia with Parkinson Disease

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Background: Phantogeusia associated with Parkinson Disease has not heretofore been reported. Methods: A 59 year old right handed female presented with a four year history of a bitter, sour and sweet taste on her entire tongue and roof of her mouth, 8/10 intensity, constant, persistent, without any external stimuli. Drinking water tasted bitter and sour. The phantogeusia was unresponsive to dietary changes, gabapentine, and allergy medications. Results: Abnormalities in Neurological examination: Decreased blink frequency. Hypokinetic. Hypomimetic face. Mood appears sad. Cranial Nerve (CN) examination: CN lll, IV, VI: Saccadization of horizontal eye movements. Motor Examination: Pill rolling tremor in right hand. 1+ cogwheel rigidity in left upper extremity. Gait: 2+ retropulsion. Chemosensory testing: Olfactory: Alcohol Sniff Test: 6 (anosmia). SNAP Phenylethyl Alcohol Threshold Testing left -2.5 (hyposmia)