Clinical Neurophysiology (CSCN) Neurocritical Care

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Desynchronization in high-definition transcranial direct current stimulation of refractory status epilepticus

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Background: Recently, we showed cathodal high-definition transcranial direct current stimulation (HD-tDCS) inhibits spikes in refractory status epilepticus (RSE) patients. We sought to characterize relative power changes within regional frequency bands to explore connectivity throughout stimulation. Methods: 28 ICU-EEGs from 10 patients were filtered with 1Hz-lower/127Hz-upper/60Hz-notch. Power spectral density was computed for each individual bipolar-longitudinal (excluding midline) electrode chain in delta/theta/alpha/beta/gamma frequency bands. Relative power was extracted throughout recordings by Welch's method with 2.5-second Hanning window size with 50% overlap every 10 seconds. The average of relative powers from ≤20-minute pre/during/post stimulation windows were calculated per session/chain/band. Absolute maximal relative power change between pre-during and during-post were computed. ANOVA statistically tested for differences. Results: Change in pre-during for delta/theta/alpha/beta/gamma were respectively -0.7%/-3.1%/-2.8%/-2.1%/8.2%, and during-post were 1.3%/2.8%/2.3%/1.7%/-7%. Positive average changes are increases in average relative power where negative changes show decreases. Changes in absolute maximal average relative power between pre-during and during-post stimulation between frequency bands were significant respectively (p=0.0011, p=0.0139). Conclusions: Relative power in all bands drop pre-during and increase during-post except gamma which correlates with desynchronization being a mechanism for HD-tDCS efficacy in RSE. These frequency band power changes may imply brain connectivity changes that need to be further explored.

Neuromuscular Disease and EMG

P.079

A novel case of pickle ball neuropathy

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Background: We report a case of distal right radial cutaneous sensory neuropathy secondary to participation in Pickle Ball. Methods: Case Report & Review of the LiteratureResults: Our 67yoF patient developed acute onset persistent numbness and dysesthesia in the distal right radial sensory distribution while participating in Pickle Ball, a sport undergoing rapid growth of popularity over the past decade. Nerve conduction studies (NCS) were performed in our lab following symptom onset, and the patient had previous NCS that allowed for comparison. NCS in our lab demonstrated asymmetrically reduced amplitude of right distal radial sensory responses (14.5 uV), in comparison to normal (42 uV) radial sensory responses from her previous NCS using the same protocol. All remaining NCS were normal in both studies. Conclusions: This study represents the first reported case within the literature of a Pickle Ball induced distal radial cutaneous sensory neuropathy demonstrated objectively using comparative nerve conduction studies. It is important that clinicians be aware of this entity and the possibility that it can manifest in the context of activities involving repetitive extension of the wrist. We present this case along with a review of the current literature of focal neuropathies involving the upper & lower extremities.

Multi-society MS/Neuroinflammatory Disease

P.082

White matter abnormalities suggestive of multiple sclerosis in Wolfram syndrome: report of two unrelated cases

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Background: Wolfram syndrome (WFS) is a genetic disorder clinically characterized by optic atrophy (OA), diabetes mellitus, sensorineural deafness, and diabetes insipidus. It is caused by mutations in *WFS1* (mono- or biallelic) or *CISD2* (biallelic) genes. Neuroradiological features include cerebellar and/or brainstem atrophy with visual pathway and white matter involvement. We report two subjects with WFS in which multifocal, progressive, and contrast-enhancing white matter abnormalities (WMA) led to the consideration of multiple sclerosis (MS). Methods: We retrospectively analyzed the clinical, genetic, and radiological data from two unrelated subjects with genetically confirmed WFS and multifocal WMA. Results: Subject I: a 43-year-old woman, heterozygous for a known *WFS1* variant, had a history of congenital deafness and OA. The brain MRI documented progressive multifocal WMA including pericallosal lesions. Subject II: a 28-year-old woman, compound heterozygous for two *WFS1* variants, was known for OA and diabetes mellitus. The brain MRI revealed multifocal periventricular, callosal, subcortical, and juxtacortical WMA, with some enhancing after gadolinium injection. Conclusions: Our report expands the WFS spectrum of white matter involvement to include progressive, seemingly inflammatory lesions. Although we cannot exclude a dual diagnosis, the roles of *WFS1* and *CISD2* in myelination suggest a selective white matter vulnerability in WFS.