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HIGHLIGHTS

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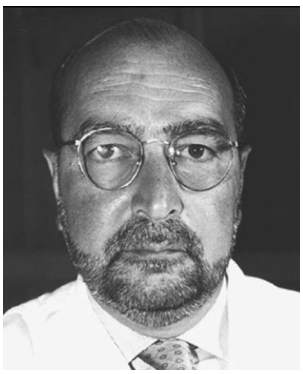

Antonio Federico
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EDITORIAL

The history of the first 40 years of Neurological Sciences

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REVIEW ARTICLES

Basal ganglia calcifications (Fahr's syndrome): related conditions and clinical features

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Basal ganglia calcifications could be incidental findings up to 20% of asymptomatic patients undergoing CT or MRI scan. The presence of neuropsychiatric symptoms associated with bilateral basal ganglia calcifications (which could occur in other peculiar brain structures, such as dentate nuclei) identifies a clinical picture defined as Fahr's Disease. This denomination mainly refers to idiopathic forms in which no metabolic or other underlying causes are identified. Recently, mutations in four different genes (SLC20A2, PDGFRB, PDGFB, and XPR1) were identified, together with novel mutations in the Myogenic Regulating Glycosylase gene, causing the occurrence of movement disorders, cognitive decline, and psychiatric symptoms. On the other hand, secondary forms, also identified as Fahr's syndrome, have been associated with different conditions: endocrine abnormalities of PTH, such as hypoparathyroidism, other genetically determined conditions, brain infections, or toxic exposure. The underlying pathophysiology seems to be related to an abnormal calcium/phosphorus homeostasis and transportation and alteration of the blood-brain barrier.

Statin, cholesterol, and sICH after acute ischemic stroke: systematic review and meta-analysis

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Background and purpose Conflicts exist regarding relationship between prior/new statin use, cholesterol, and early poststroke intracranial hemorrhage (ICH) in acute ischemic stroke (AIS) patients. This meta-analysis is aimed at evaluating the safety of prior/new statin use, cholesterol level and risk of ICH in AIS patients. Methods The AA searched PubMed and Embase for studies examining relation between statin use, cholesterol level, and early poststroke ICH in AIS. Included studies should report risk of early poststroke symptomatic ICH (sICH) or overall ICH. A random-effects model was used to pool the data. Results Twenty-five articles involving 26,327 participants were included, among whom 925 had sICH. Prior statin use was not associated with overall ICH (adjusted odds ratio (OR), 1.478; 95%confidence interval (CI), 0.924–2.362; p = 0.103) and sICH in patients who received thrombolysis (adjusted OR, 1.567; 95% CI, 0.994–2.471; p = 0.053) or overall ICH in patients, most of whom had not received recanalization therapy (crude OR, 1.342; 95% CI, 0.872–2.065;

p = 0.181). New statin use was associated with decreased sICH after recanalization therapy (crude OR, 0.292; 95% CI, 0.168–0.507; p < 0.001). Cholesterol level was not associated with overall ICH. Conclusion Prior/new statin use and lower cholesterol level are not risk factors for sICH and overall ICH in AIS patients, whether or not the patient has received recanalization therapy. New statin use is likely associated with decreased sICH.

Exploring the role of music therapy in multiple sclerosis: brief updates from research to clinical practice

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Background Physical, cognitive and mood-behavioral disturbances are very common in people with multiple sclerosis (MS) representing a relevant disease burden. Recently, in this field, several studies investigated the role of music therapy (MT) as a complementary therapeutic approach especially in terms of rehabilitation strategy. Objectives The aim of this review is to report and discuss the effectiveness of various music-based interventions (MBIs) for clinical outcomes in MS patients. Data sources All English medical papers registered in theWeb of Knowledge, PubMed, Google Scholar, and ScienceDirect from March 1999 to March 2019. Inclusion and exclusion criteria We selected all the articles concerning MBIs in MS including papers that dealt with human samples and excluding non-human samples, reviews and case reports. Results Out of 46 articles, we selected 24 papers of which 13 completely following the inclusion criteria were evaluated for the present analysis. Discussion The AA explored the efficacy of several MT programs, taking into account the different aspects of application feasibility in the clinical management of MS patients and the future challenges.

ORIGINAL ARTICLES

UNC13A variant rs12608932 is associated with increased risk of amyotrophic lateral sclerosis and reduced patient survival: a meta-analysis

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Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease associated with both genetic and environmental risk factors. Previous studies trying to find an association between ALS and unc-13 homolog A (UNC13A) gene variants have shown inconsistent results. This study aimed to conduct a meta-analysis of the association between the C allele of rs12608932, a single-nucleotide polymorphism located in an intron of UNC13A, and risk of ALS and patient survival.

PubMed, Web of Science, Embase, Chinese National Knowledge Infrastructure, Wanfang, and SinoMed databases were systematically searched for genome-wide association studies or case-control studies published up to January 2019 on the association between this variant in UNC13A and risk and/or prognosis of ALS. Data from eligible studies were extracted and analyzed.

The pooled data (28,072 patients with sporadic ALS and 56,545 controls) showed that rs12608932(C) was associated with an increased risk of ALS (OR = 1.13, 95%CI 1.07–1.20). Subgroup analysis revealed that rs12608932(C) increased the risk of sporadic ALS in non-Asian individuals, including those from the USA and Europe (OR 1.17, 95%CI 1.10–1.25, P < 0.000), but not in Japanese or Chinese subjects (OR 1.01, 95%CI 0.92–1.10, P = 0.85). The available data demonstrated that the CC genotype decreased the survival time of patients with ALS (OR 1.33, 95%CI 1.19–1.49, P < 0.001). The present meta-analysis suggests that rs12608932(C) is associated with increased ALS susceptibility, especially in Caucasian and European subjects, and that the CC genotype of rs12608932 is associated with reduced ALS patient survival.

Biallelic pathogenic variants in TBCD-related neurodevelopment disease with mild clinical features

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Microtubule dynamics is crucial for neuronal function and survival. The disrupted function of microtubule dynamics would lead to neurodegenerative and neurodevelopmental disorders. Tubulin-specific chaperone D (TBCD) is one of five tubulin co-chaperones acted in assembly and disassembly dynamics of microtubule. The biallelic pathogenic variants of TBCD gene were reported to be associated with severe degenerative encephalopathy accompanied with seizures previously. Compound heterozygous variants were identified in three patients from three families. The in silico prediction software and ACMGstandards and guidelines proved the pathogenicity of the TBCD pathogenic variants. The clinical features of the three patients presented with mild neurodevelopmental manifestations including autism spectrum disorder (ASD) and occasional generalized tonic-clonic seizures (GTCSs) responding well to antiepileptic drugs. This research expanded the clinical spectrum of TBCD-related neurodevelopmental disease which contributed to understanding the genotype-phenotype correlations of the disease.

Visuospatial learning is fostered in migraine: evidence by a neuropsychological study

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Cognitive profile in migraine patients still remains undefined. Contradictory evidence has been provided, with impairments in different cognitive domains, normal cognition, or even better performance compared to healthy controls (HC). The latter is of particular interest considering the evidence of glutamatergic upregulation in migraine, particularly in the visual cortex, and the role of the glutamatergic system in synaptic plasticity and learning. The aim of our study is to compare cognitive performance for visuospatial memory and learning (supraspan modality) between migraineurs without aura (MwoA) and HC. Twenty-one subjects suffering from MwoA and 21 HC were enrolled. Migraineurs during the interictal phase and HC underwent visuospatial memory test (Corsi test) and verbal memory test (Buschke Selective Reminding Test) in supraspan modality, Trial Making Test A (TMTA) and B (TMTB) as test exploring attention, and TMTB-TMTA as test of executive functioning. Depression was assessed with the Beck Depression Inventory Short Form (BDI-SF). Migraine characteristics (i.e., disease duration and frequency expressed as attacks per month) were collected. Subjects withMwoA showed better performance than HC in test exploring both short ($p = 0.002$) and long-term ($p= 0.001$) visuospatial memory. No significant difference between groups was found in verbal memory, attention, executive functioning, and depression (BDI-SF). No significant association emerged between cognitive performance and migraine characteristics. Subjects with MwoA had significant better performance in visuospatial memory and learning than HC. Occipitoparietal hyperexcitability (in particular in the visual cortex), which is a hallmark of the migraine brain, would probably explain these results. These data need to be confirmed in larger samples of migraineurs.

Complex therapies for advanced Parkinson's disease: what is the role of doctor-patient communication?

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Communication processes play a key role in the patient-doctor relationship. Few studies have considered communicative processes in advanced Parkinson's disease (PD), and in particular in the phase of proposing complex therapies (CT). Therefore, we explored the role of communication and patient-doctor relationship in the transition phase to CT for advanced PD, analysing satisfaction, factors influencing the relationship and patients' unmet needs. Twenty-four PD patients (mean age 61.7 ± 8.8 years; mean disease duration 12 ± 4.8 years) eligible for deep brain stimulation or infusion therapies were submitted to a semi-structured interview aimed to investigate communication related cognitions, feelings and behaviours concerning PD and the possible transition towards CT. The Patient-Doctor Relationship Questionnaire (PDRQ-9) was administered along with neuropsychological and behavioural screening tests. All patients discussed the possible transition to CT with a neurologist. A high degree of satisfaction about the relationship with the neurologist was revealed (mean PDRQ-9 score 37.3 ± 7.3). The communication not only aroused feelings of fear (11/24 patients) and concern (15/24 patients), but also fostered the hope for motor improvement (15/24 patients). Half the patients (12/ 24) wanted to receive more information about CT after communication. This pilot study highlights the importance of doctor-patient communication in PD when facing the transition to CT. Trust in the physician emerged as a key point in favour of the therapeutic alliance. Neurologists should carefully consider patients' reactions and preferences for eliciting collaboration and treatment adherence, favouring a patient-centred standard of care.

BRIEF COMMUNICATION

"Accent issue": foreign accent syndrome following ischemic stroke

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Foreign accent syndrome (FAS) is a rare syndrome associated with altered speech rhythm and prosody, which listeners perceive as foreign; cerebrovascular accidents, tumors and multiple sclerosis are reported as possible causes of FAS. The pathophysiology of FAS is not yet understood. A 68-year-old Italian man was admitted to the Emergency Department for non-fluent aphasia and dysarthria. Computed tomography (CT) scan did not show abnormalities; the patient was treated with systemic thrombolysis. A repeated brain CT and magnetic resonance imaging (MRI) confirmed an infarct in the left primary motor cortex and mild extension to cortico-subcortical frontal regions. In the following days he gradually improved, speaking Italian fluently with a typical German accent. In conclusion, FAS is a rare motor speech disorder, often related to cerebrovascular accidents involving critical regions in the dominant hemisphere. In addition, the present case adds further evidence to the role of the left primary motor cortex in modulation of prosody. In rare cases FAS can be the only sign of stroke or can appear after recovery from post-stroke aphasia.