

Highlights of the issue 10, 2019

Published online: 7 September 2019
© Fondazione Società Italiana di Neurologia 2019



Antonio Federico
Editor-in-Chief

REVIEW ARTICLES

Posterior circulation ischaemic stroke—a review. Part I: anatomy, etiology and clinical presentations

Marco Sparaco, Ludovico Ciolli, Andrea Zini
(Italy)

<https://doi.org/10.1007/s10072-019-03977-2>

Posterior circulation ischaemia is a clinicopathological condition with complex symptomatology associated with an infarction within the vertebrobasilar arterial system. Posterior circulation strokes account for about 20–25% of all ischemic strokes and remain a significant cause of patient disability and mortality. Diagnosis can be challenging because presenting symptoms are often non-focal and because there is a substantial overlapping symptoms and signs of ischaemia in the anterior circulation. Despite better imaging techniques, diagnosis and treatment of life-threatening conditions, such as basilar artery occlusions, are often delayed. Therefore, early detection of symptoms and causes of posterior circulation ischaemia is essential for choosing the most appropriate therapy. In this review, the AA summarize the anatomy, etiology, typical presentations and characteristic findings of common strokes resulting from disease in the vertebrobasilar arterial system.

Management of antibody-mediated autoimmune encephalitis in adults and children: literature review and consensus-based practical recommendations

Luigi Zuliani, Margherita Nosadini, Matteo Gastaldi, Marianna Spatola, Raffaele Iorio, Marco Zoccarato, Sara Mariotto, Piera De Gaspari, Francesco Perini, Sergio Ferrari, Amelia Evoli, Stefano Sartori, Diego Franciotta, Bruno Giometto
(Italy)

<https://doi.org/10.1007/s10072-019-03930-3>

Autoimmune encephalitis associated with antibodies against neuronal surface targets (NSAE) are rare but still under recognized conditions that affect adult and pediatric patients. Clinical guidelines have recently been published with the aim of providing diagnostic clues regardless of antibody status. These syndromes are potentially treatable but the choice of treatment and its timing, as well as differential diagnoses, long-term management, and clinical and paraclinical follow-up, remain major challenges. In the absence of evidence-based guidelines, management of these conditions is commonly based on single-center expertise. Taking into account different published expert recommendations in addition to the multi-center experience of the Italian Working Group on Autoimmune Encephalitis, both widely accepted and critical aspects of diagnosis, management and particularly of immunotherapy for NSAE have been reviewed and are discussed. Finally, the AA provide consensus-based practical advice for managing hospitalization and follow-up of patients with NSA.

Dietary pattern in relation to the risk of Alzheimer's disease: a systematic review

Mehnoosh Samadi, Shima Moradi, Mehdi Moradinazar, Roghayeh Mostafai, Yahya Pasdar
(Iran)

<https://doi.org/10.1007/s10072-019-03976-3>

Alzheimer's disease (AD) is a progressive neurodegenerative disease leading to a gradual and irreversible loss of memory, linguistic skills, and perception of time and space, thinking, and behavior. Dietary pattern has been presented as a contributor to the incidence of Alzheimer's. This study aimed at

reviewing the evidence on the relation between dietary pattern and AD. This systematic search was performed on the articles available in PubMed, Scopus, and Web of Sciences databases until May 2019 using keywords, including (diet, food, dietary pattern, food pattern) and (Alzheimer's disease) among observational studies. After excluding duplicated, and irrelevant studies, 26 studies were eligible for this review study. The AA categorized the studied dietary patterns into two groups: healthy and unhealthy diet. This study reviewed two case-control, five cross-sectional, and 19 prospective studies. Eight studies assessed unhealthy diet (high-fat diet, high-glycemic diet, sweetened sugary beverage, etc.) and the risk of AD. In addition, the other studies considered the effect of healthy diet such as Mediterranean diet, dietary approaches to stop hypertension (DASH), Mediterranean-DASH intervention for neurodegenerative delay, and seafood-rich diet on AD. This literature review indicated that adherence to a healthy dietary pattern has neuroprotective effects on AD prevention, while unhealthy diet can cause neurodegenerative effects in AD etiology. In conclusion, our findings showed that adherence to healthy diet can decrease oxidative stress and inflammation and accumulation of amyloid- β and consequently can decrease the risk of AD.

ORIGINAL ARTICLES

Diaphragm ultrasonography in amyotrophic lateral sclerosis: a diagnostic tool to assess ventilatory dysfunction and disease severity

Ferdinando Sartucci, Alessio Pelagatti, Michela Santin, Tommaso Bocci, Cristina Dolciotti, Paolo Bongioanni.
(Italy)

<https://doi.org/10.1007/s10072-019-03938-9>

Respiratory failure represents an unavoidable step in patients with amyotrophic lateral sclerosis (ALS) and other motor neuron diseases (MND). The development of diaphragm ultrasonography (DUS) provides an alternative useful and risk free tool to supply clinical, functional, and neurophysiological assessment of respiratory muscle weakness. Our aim was to evaluate if sonographic changes (thickness and echogenicity in the costal portion of the diaphragm, at rest and during respiratory movements) may be used in ALS patients to assess disease severity overtime, to rule out any risk or discomfort due to traditional neurophysiological investigations. Twenty ALS patients (mean age, 64.6 ± 10.5 years) were enrolled and data were compared with age-matched healthy volunteers; DUS data were correlated with respiratory function and disease severity scale. Examinations were performed using Teled Echo-wave II or Esaote MyLab Gamma devices in conventional B-Mode. Mean resting thickness was reduced in all cases; changes in thickness during inspiration and expiration were also reduced ($p < 0.0001$) and lost in severe cases ($n = 3$). In bulbar-onset disease, respiratory scores were strictly

correlated with the difference in diaphragm thickness between full inspiration—and expiration—as well as on the diaphragm thickness in expiration ($p < 0.001$). Conclusions DUS represents a simple, painless, and risk-free tool; moreover, it provides useful functional and structural insights to the understanding of diaphragm function and the degree of respiratory failure in ALS.

Brain volume is related to neurological impairment and to copper overload in Wilson's disease

Lukasz Smolinski, Tomasz Litwin, Barbara Redzia-Ogrodnik, Karolina Dziezyc, Iwona Kurkowska-Jastrzebska, Anna Czlonkowska
(Poland)

<https://doi.org/10.1007/s10072-019-03942-z>

To determine whether brain volume was associated with functional and neurological impairments and with copper overload markers in patients with Wilson's disease. In 48 treatment-naïve patients, the AA assessed functional and neurological impairments with the Unified Wilson's Disease Rating Scale, measured normalized brain volumes based on magnetic resonance images, and assessed concentration of non ceruloplasmin-bound copper. The AA correlated brain volume measures with functional and neurological impairment scores and copper overload indices. Results Functional and neurological impairments correlated with all brain volume measures, including the total brain volume and the volumes of white matter and gray matter (both peripheral gray matter and deep brain nuclei). Higher non-ceruloplasmin bound copper concentrations were associated with greater functional and neurological impairments and lower brain volumes. Conclusions All findings provided the first in vivo evidence that the severity of brain atrophy is a correlate of functional and neurological impairments in patients with Wilson's disease and that brain volume could serve as a marker of neurodegeneration induced by copper.

Longitudinal study of a cohort of MSA-C patients in South Italy: survival and clinical features

Maria Lieto, Alessandro Roca, Dario Bruzzese, Antonella Antenora, Girolamo Alfieri, Francesco Saccà, Marta Bellofatto, Leonilda Bilo, Stefano Barbato, Giuseppe De Michele, Alessandro Filla
(Italy)

<https://doi.org/10.1007/s10072-019-03948-7>

Sixty-six patients with possible or probable MSA (multiple system atrophy) cerebellar type, personally observed between 2006 and 2018 were retrospectively reviewed. The time point of data collection was January 1, 2019. Forty-nine patients lost independent walking after a median time of 5 years (95% C. I. 4–6). Thirty-two patients were confined to wheelchair after a

median time of 7 years (95% C.I. 7–8). Twenty-seven patients were deceased after a median time of 9 years (95% C.I. 8–10). A later onset predicted an earlier loss of independent walking (HR 1.07; 95% C.I. 1.03–1.11; $p = 0.001$). Higher UMSARS score predicted shorter time to loss of independent walking (HR 1.04; 95% C.I. 1.02–1.06; $p = 0.001$) and to wheelchair (HR 1.03; 95% C.I. 1.01–1.06; $p = 0.021$). No predictor of time to death was found.

A novel S379A TARDBP mutation associated to late-onset sporadic ALS

Teresa Sprovieri, Carmine Ungaro, Benedetta Perrone, Giuseppina Daniela Naimo, Rossella Spataro, Sebastiano Cavallaro, Vincenzo La Bella, Francesca Luisa Conforti (Italy)

<https://doi.org/10.1007/s10072-019-03943-y>

Since 2008, several groups have reported a lot of dominant mutations in TARDBP gene as a primary cause of Amyotrophic lateral sclerosis (ALS). Mutations in TARDBP gene are responsible for 4–5% of familial ALS (fALS) and nearly 1% of sporadic ALS (sALS). To date, over 50 dominant mutations were found in TDP-43 in both familial and sporadic ALS patients, most of which were missense mutations in the C-terminal glycine-rich region. Herein, the AA describe the clinical and genetic analysis of an Italian non-familial ALS patient with a late onset and a rapid disease progression, which led to the discovery of a novel TARDBP mutation. After neurological evaluation, molecular investigation highlighted the heterozygous substitution in exon 6 of TARDBP gene (S379A), which has previously neither been described nor reported in the ALS database. Several evidences supported the S379A mutation as causative in this patient: (a) it was neither found in ExAC nor 1000G and it was absent in the AA database of control subjects; (b) the position of the mutation involves an evolutionarily highly conserved residue; (c) two different amino acid substitutions in the same 379 codon were already reported in Swedish and Italian fALS cases, supporting the critical role of this codon for the protein function. The identification of this novel mutation enlarges the number of TARDBP mutations in ALS patients.

Early extubation after thymectomy is good for the patients with myasthenia gravis

Li Chen, Wenfeng Xie, Donghua Zheng, Siqi Wang, Ganping Wang, Jiaqi Sun, Qiang Tai, Zhenguang Chen (China)

<https://doi.org/10.1007/s10072-019-03941-0>

Patients with myasthenia gravis (MG) often benefit from thymectomy, but the optimal timing of extubation following thymectomy in these patients remains unknown. This study of MG patients compared the effect of early and late extubation following thymectomy on clinical outcome. **Methods** We performed a study of data from 96 patients with MG who received thymectomy procedures, followed by early (< 6 h) or late (> 6 h) extubation, at our institution between October 2011 and November 2017. Patient clinical and demographic characteristics, preoperative data, and postoperative clinical outcomes were analyzed. Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study. **Results** The patients in the early extubation group ($n = 53$) and late extubation group ($n = 43$) had similar preoperative clinical and demographic characteristics. However, the early extubation group had a significantly longer duration of MG (24 months vs. 12 months, $P < 0.013$) and a lower incidence of reintubation (11.3% vs. 37.2%, $P = 0.003$). Postoperative pulmonary infection was significantly more common in the late extubation group (39.5% vs. 11.3%, $P = 0.001$; adjusted odds ratio = 6.94, 95% CI 1.24–38.97). Also, patients in the late extubation group had a longer duration of ICU stay (6.4 ± 4.0 h vs. 4.3 ± 1.8 h; $P = 0.003$) and had a longer adjusted duration of ICU stay by 0.93 days (95% CI 0.02–1.85). **Conclusions** Our analysis of patients with MG who received thymectomy procedures indicated that early extubation was associated with improved clinical outcomes, in particular with reduced risk of postoperative pulmonary infection and reduced ICU stay.

Publisher's note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations