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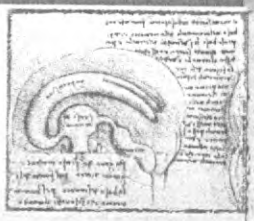

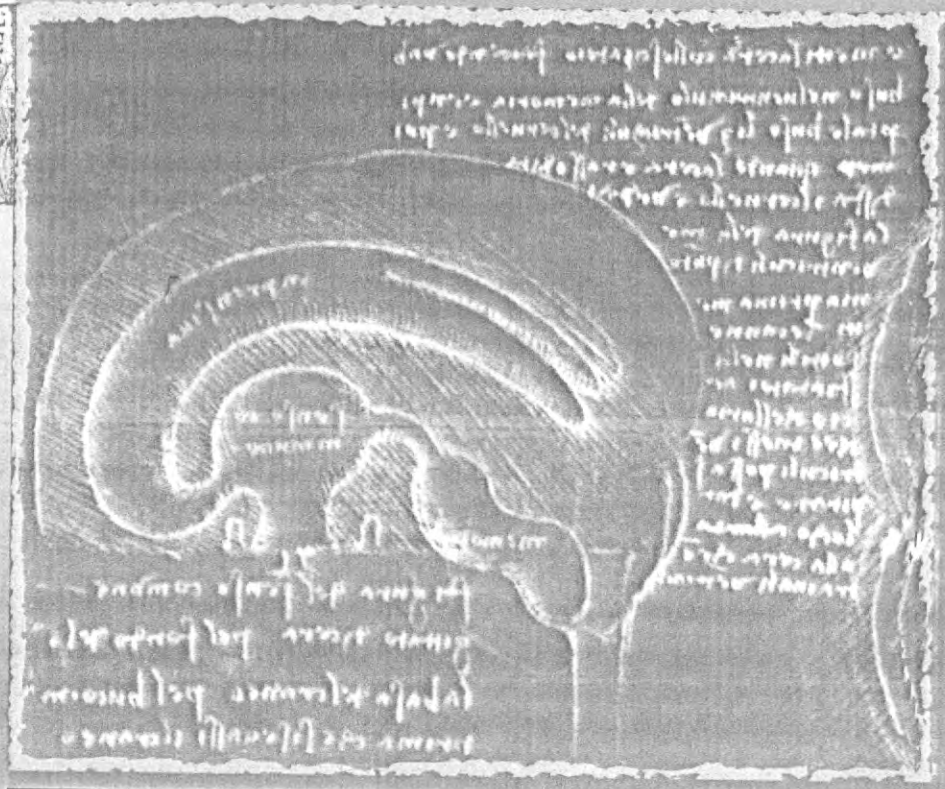

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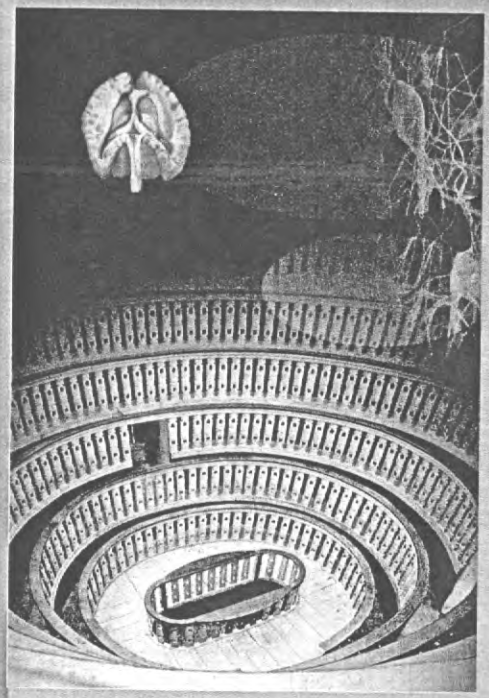

XL Congress of the Italian Neurological Society

ABSTRACTS

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Neurological Sciences

Founded by Renato Boeri
Official Journal of the Italian Neurological Society

Continuation of
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Neurological Sciences

Neurological Sciences is intended to provide a medium for the communication of results and ideas in the field of neuroscience. The journal welcomes contributions in both the basic and clinical aspects of the neurosciences. The official language of the journal is English. Reports are published in the form of original articles, short communications, editorials, reviews and case reports. Original articles present the results of experimental or clinical studies in the neurosciences, while short communications are succinct reports permitting the rapid publication of novel results. Original contributions may be submitted for the special sections History of Neurology, Health Care and Neurological Digressions - a forum for cultural topics related to the neurosciences. The journal also publishes correspondence book reviews, meeting reports and announcements.

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ABSTRACTS

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Background: In the last decades various studies have showed that cognitive and behavioural disorders are frequent in ALS.

Objectives: To delineate a specific cognitive and behavioural profile in patients with early ALS . Materials and Methods: sixteen patients with probable or definitive ALS disease, according to El Escorial (1998) criteria, were tested with an extensive neuropsychological testing. We have included patients with 0–2 years history disease. The patients were recruited in order of diagnosis to avoid an inadequate selection. A behavioural scale (The Neuropsychiatric Inventory) and a depression scale (Hamilton Rating Scale for Depression) were administered.

Results: In the 40% of patients at least four tests, especially frontal tests, were impaired. FAB (The Frontal Assessment Battery) was the more sensible test to diagnose a cognitive impairment. All the patients with cognitive impairment had a mild cognitive impairment and two had a dementia. Apathy was the more frequent behavioural disorder.

Discussion and Conclusions: Cognitive and behavioural disturbances are frequent in patients with early ALS disease. Dysexecutive syndrome is the typical cognitive pattern in patients with ALS. The FAB is a sensible cognitive screening instrument in these patients.

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SOMATOPARAPHRENIA: A CASE REPORT

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Delusional beliefs concerning the controlesional side of the body were termed by Gerstmann "somatoparaphrenia" [1]. This neuropsychological disorder of body awareness has been reported more often in right-brain-damaged patients, with motor and somatosensory deficits and syndrome of unilateral spatial neglect. These findings suggest that the sense of body identity and ownership are largely right-hemisphere based. Only few cases exhibite somatoparaphrenia for the right side of the body, associated with a left-sided lesion [2]. In this report we describe an uncommon case of somatoparaphrenia after left brain damage. MS, a 84-year-old right-handed man, was admitted to our department about 7 days after a left hemispheric ischemic stroke, for rehabilitative treatment. On admission clinical examination revealed fluent aphasia, right spatial extrapersonal and personal neglect, anosognosia, colour agnosia, right homonymus hemianopia, right sensorimotor hemisyndrome with somatosensory impairment for touch and position. Brain MRI demonstrated a subacute ischemic lesion in the territory of the left posterior cerebral artery (medial occipital lobe, with extension to ipsilateral thalamus). No history of confusional state and no documented cognitive impairment were known previously. During the first weeks after stroke, MS developed denial of hemiparesis with confabulations and delusions of disownership of right-sided body parts. He told that his right hand belonging to his wife, sometimes he told to have supernumerary hands (from one to ten). We also observed apparent autonomy of the acts performed by his right hand such as alien hand phenomenon: the unintended movements caused serious lesions on the dorsal face of his right hand. These disorders of representation of the body concern were transitory and recovered over few weeks but slowed down rehabilitative treatment. This case opens the debate about the hemispheric asymmetries and the neural processes concerned with body awerness and suggests a role of the posterior cerebral regions as neural basis of somatoparaphrenia.

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VERY SLOW PROGRESSION OF CLINICALLY DIAGNOSED BEHAVIOURAL VARIANT OF FRONTOTEMPORAL DEMENTIA

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Introduction: The behavioural variant of Frontotemporal dementia (bv-FTD) is a neurodegenerative disease characterised by early and progressive changes in personality and behaviour, such as apathy, reduced empathy and lack of the usual social mores, or dishinibition, agitation, irritability and stereotypical and ritualized behaviours (Hodges and Miller, 2001). Imaging changes in frontotemporal regions are supportive but not mandatory for the diagnosis (Neary *et al.*, 1998). Recently, Kipps *at al.* (YEAR), compared two groups of patient with a clinical diagnosis of bv-FTD. At the onset of disease the first group showed an absence (or mild) brain atrophy and hypometabolism, while the second group showed a definite frontotemporal hypometabolism in FDG-PET analysis. These patient were followed for 6 years, and a lack of progression was observed in the first group. In contrast, patients with significant frontal atrophy of hypometabolism at onset showed a clear-cut deterioration over time. The authors suggest that these findings support the existence of a non-degenerative phenocopy of the disease.

Material and methods: We describe three patients who met the criteria for bv-FTD (Neary et al. , 1998); no patient had clinical or neuroradiological evidence of cerebrovascular disease, or a positive hystory of psychitaric disorder or alcohol abuse. CT scan of all patients at the onset of the disease was normal, while SPECT imaging showed a significant hypoperfusion in frontotemporal regions. Neuropsychological evaluation showed a profile characterized by executive function deficits in all patients. Memory and visuo-spatial abilities were also impaired, possibly as a consequence of attentional and executive disorders. All patient were regularly followed over a period of 10 years (2000–2009).

Results and conclusions: Neuroimaging studies showed a significant progression of atrophy and hypoperfusion in frontotemporal areas. Neuropsychological evaluation showed a mild and aspecific progression of the deficits. The clinical and functional status was stable over time. In particular, the behavioural disturbances remained relatively stable in the type and frequency of symptoms, while activities of daily living were spared. These findings suggest the existence of a variant of bv-FTD characterised by very slow clinical progression.

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