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The role of functional MRI in the diagnosis and prognosis of patients with severe chronic disorders of consciousness

B. Wutzl¹, C. Florea², M. Seidl², A.B. Kunz²,
K. Schwenker², R. Nardone², E. Trinka³,
F. Gerstenbrand⁴, S. Golaszewski²

¹Neurology, Paracelsus Medical University Salzburg, Salzburg, Austria, ²Salzburg, Austria, ³Department of Neurology, Paracelsus Medical University Salzburg, Salzburg, Austria, ⁴Vienna, Austria

Background and aims: Accurate diagnosis of patients with severe chronic disorders of consciousness (scDOC) following brain damage is essential for clinical and rehabilitative care as well as decision-making and a rate of 43% of misdiagnosis is evident. Neurobehavioral tests relying on the patients' intellectual and motor ability to communicate are the most widely used diagnostic tools, since their advantage over clinical assessment has been validated. However, with the emergence of modern neuroimaging methods, especially fMRI, objective physiological markers for assessing the state of consciousness are available but the benefits still have to be determined.

Methods: 21 patients clinically and neurobehaviorally diagnosed as "Apallic-Syndrome (AS)" and 6 patients as "Minimally Conscious State (MCS)" after brain damage of different etiologies were examined with different fMRI paradigms testing fundamental functional networks of the brain (proprioceptive, pain, motor, emotion, self-awareness, language, resting state). The findings were compared with the clinical and neurobehavioral diagnosis and it was analyzed whether additional information from fMRI confirmed or questioned the clinical and neurobehavioral diagnosis.

Results: 16 of the 21 AS- and 5 of the 6 MCS-patients show specific brain activation in a special diagnostic battery of fMRI-paradigms suggesting that the AS-patients are in MCS or even better.

Conclusion: Misdiagnosis in scDOC-patients is still a big problem even with well-established diagnostic assessment scales. As long as internationally accepted guidelines for assessing patients with scDOC do not exist, we propose a special diagnostic battery of fMRI-paradigms to minimize diagnostic errors in these patients and to find systematically perceptive channels to approach the patients in neurorehabilitation programs.

Disclosure: Nothing to disclose

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Cerebrospinal fluid anti-Caspr2 antibodies determine a subtype of autoimmune encephalitis with prominent limbic symptoms and seizures

B. Joubert¹, M. Saint Martin², N. Noraz², G. Picard³,
V. Rogemond², F. Ducray⁴, V. Desestret¹, D. Psimaras⁵,
J.-Y. Delattre⁵, J.-C. Antoine⁶, J. Honnorat¹

¹Lyon Bron, France, ²Neuroscience, INSERM, Lyons, France, ³Neurology, Hospices Civils de Lyon, Lyons, France, ⁴Lyons, France, ⁵Paris, France, ⁶CHU Saint-Etienne, Saint-Etienne, France

Background and aims: Autoantibodies against Caspr2 (Caspr2 ab) associate with several neurological syndromes, including neuromyotonia, Morvan's syndrome and limbic encephalitis. We aimed to characterize the clinical and biological presentations of patients with Caspr2 ab in the cerebrospinal fluid (CSF).

Methods: We analysed the clinical presentations, ancillary features and outcomes of all patients with CSF Caspr2 ab detected in our centre between March 2009 and November 2015.

Results: We identified 18 patients (males, 94%; median age, 64.5 years). 3 patients (17%) had a history of cancer (prostate, haematological, thyroid). Symptoms of limbic encephalitis were seen in all patients, including memory disorders in 17/18 (94%) and seizures in 16/18 (89%). Extra-limbic signs were observed in 12/18 patients (67%) including cerebellar ataxia in 6 cases (33%). Brain MRI displayed temporo-limbic T2-weighted abnormalities in 14/15 (93%) patients. CSF analysis was abnormal in 9/12 (75%) patients. 16 patients (89%) were followed-up at least 6 months (median, 34 months). Relapses occurred in 6 of them (37.5%). 15 patients (94%) improved. CSF IgG4 autoantibodies were found in all the patients, along with IgG1 autoantibodies in 10/17 patients (59%). CSF Caspr2 ab targeted the laminin G1 and discoidin domains of Caspr2 in all patients, along with other Caspr2 epitopes in 10/18 (56%) cases.

Conclusion: CSF Caspr2 ab associate with a subtype of autoimmune encephalitis with prominent limbic involvement and seizures that rarely associate with cancer. IgG4 autoantibodies targeting the discoidin and laminin G1 domains of Caspr2 are constant and might have a functional effect on Caspr2.

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HEAD OFFICE: Breite Gasse 4/7
1070 Vienna, Austria

PHONE: +43 1 889 05 03
FAX: +43 1 889 05 03 13
E-MAIL: headoffice@eaneurology.org
WEB: www.eaneurology.org

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