The Locked-In-Plus-Syndrome

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Background and aims

Recently, an intense discussion started about the nomenclature and examination protocols of patients with disorders of consciousness and related syndromes as the locked-in-syndrome (LIS). As the number of publications finding brain responses to different stimuli using fMRI or Evoked Potentials in these patients increases, we are in need of a diagnosis scheme which best fits to describe these patients. LIS plus (LIPS) is proposed as the diagnostic category for patients who show typical signs of LIS combined with disorders of consciousness.

Methods

We collected clinical and instrumental data of these patients to start with the development of a new classification for patients. 10 patients with pontine and other brain lesions following vascular injuries were examined clinically, with standardized behavioral assessment scales, MRI and fMRI.

Results

All 10 patients showed different degrees of arousal, consciousness, and other neurological and behavioral symptoms. Additionally the extent of structural brain damage and brain response in fMRI was found to be variable in spread.

Conclusion

The main clinical differences between LIS and LIPS comprise a variety of additional extrapontine brain lesions and frequently occurring symptoms as hypersonic syndromes, frontal release signs, thalamic posturing of hand and/or feet. Rarely an akinetic mutism may be present in LIPS. Extra-pontine brain lesions may frequently occur in mesencephalic, thalamic and cerebellar brain structures. Also an involvement of occipital and temporal brain regions is possible, depending on varieties of the vertebro-basilar artery blood supplying system. Due to the heterogeneity of the collected data a new diagnostic category should be implemented in clinical practice. World Federation for NeuroRehabilitation presents its Biennial Congress



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