Epileptic Disord 2019; 21 (Suppl. 1): S1-S2

Linking epilepsy, sleep disruption and cognitive impairment in Encephalopathy related to Status Epilepticus during slow Sleep (ESES)

"Encephalopathy related to Status Epilepticus during slow Sleep (ESES)" was described more than forty-five years ago in a small group of children with learning disabilities who displayed a peculiar EEG pattern consisting of apparently "sub-clinical" spike-and-wave discharges, that occurred almost continuously during sleep for a variable length of time (months to years). Later on, the condition of a protracted "status epilepticus during sleep" (SES) in the developmental age was proposed to be the factor leading to the appearance of severe cognitive and psychic disturbances. Indeed, the extreme activation of epileptic activities during NREM sleep still stands as the electroencephalographic hallmark of a condition that, if prolonged, causes the appearance of a clinical picture that has been acknowledged to be an encephalopathy related to SES. From a broader perspective, SES may be responsible not only for cognitive dysfunctions, such as for instance acquired aphasia, *i.e. Landau-Kleffner syndrome*, but also (and often concomitantly) for other dysfunctions, such as severe behavioral disorders and motor impairment (*i.e.* apraxia and negative myoclonus). Etiology of ESES can be heterogeneous as well, in fact it has been reported in children with organic brain lesions as well as in children with an epilepsy of benign evolution - whether idiopathic or cryptogenic. After hundreds of observations and comprehensive reviews on the subject (including a monography on the Venice Symposium edited by Beaumanoir *et al*. in 1995), it became clear that SES, cognitive

Figure 1. From left to right: Alexis Arzimanoglou, Rikke S. Møller, José Serratosa, Guido Rubboli, Gaetano Cantalupo, Pierre Szepetowski, Carlo Alberto Tassinari, Sandor Beniczky, Tobias Loddenkemper, Michael Siniatchkin, Philippe Paquier, Edouard Hirsch, Reto Huber, J. Helen Cross, Patrick van Bogaert, Floor E. Jansen.

doi:10.1684/epd.2019.1049

impairment and behavioural disturbances evolve in parallel, and in fact when these latter disorders recover, SES tends to disappear (or it is already over). However, in spite of the wealth of data accumulated over decades, the clinical spectrum of ESES and its boundaries, the diagnostic criteria, the pathophysiology and the therapeutic management are still a matter of debate.

In recent years, clinical observations, neurophysiological and imaging investigations, and genetic studies have renewed interest in ESES. In addition, experimental findings from sleep research have opened fascinating perspectives on some possible pathophysiological mechanisms involved in this condition. These issues have been discussed at the 1st Dianalund International Conference on Epilepsy on "*Encephalopathy related to Status Epilepticus during slow Sleep. Linking epilepsy, sleep disruption and cognitive impairment*" that was organized by Guido Rubboli, Marina Nikanorova, and Carlo Alberto Tassinari on March 14-15, 2014 in Sorø (Denmark). Clinicians, neurophysiologists, sleep physiologists and geneticists (*figure 1*) who have studied ESES gathered with the aim to create a better overall understanding of this special syndrome in the light of recent research. Based on the scientific and educational structure of the meeting, the speakers and session chairs were later invited to contribute with an updated review of their topics. All these contributions are acknowledged in this Supplement with the aim to provide an updated overview of the current knowledge on ESES. □

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