

SLEEP DISTURBANCE IN RETT SYNDROME

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Sleep problems are extremely common in Rett syndrome, with a significant burden for the health and wellbeing of both those with Rett syndrome and their families.

A recent study showed, in a sample of more than 300 cases followed over 12 years, that the prevalence of any sleep disturbance was very high (more than 80%) and decreased with age (less common in individuals older than 18 years). Night laughing and night screaming occurred in 77 and 49%, respectively, when younger. Those with a large deletion had a higher prevalence of night laughing, which often occurred frequently. Sleep medications such as melatonin and chloryl hydrate were associated with a 1.7% reduction in risk of further sleep problems.

Among 21 patients followed by our centre in the last two years, 21,7% (5) reported sleep disturbances, including nocturnal awakenings, bruxism and sleep talking. Two patients were treated with niaprazine, one with periciazine, one with melatonine and all of them had good response.

Chronic sleep deprivation compromises quality of life of both children and families and is associated with poorer developmental outcome, overweight, and behavioral disturbances. Despite the widespread use of pharmacological treatment, the lack of well designed, controlled studies concerning the efficacy, tolerability, dosage, and safety profile of hypnotic medications in children raise the need of further research.