

Are there distinctive sleep problems in Angelman syndrome?

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Angelman syndrome is a neurogenetic condition characterized by developmental delay, absence of speech, motor impairment, epilepsy and a peculiar behavioral phenotype that includes sleep problems. It is caused by lack of expression of the UBE3A gene on the maternal chromosome 15q11-q13. Although part of the diagnostic description, 'sleep problems' are not well characterized. A pattern emerges from the available reports. It includes reduced total sleep time, increased sleep onset latency, disrupted sleep architecture with frequent nocturnal awakenings, reduced rapid eye movement (REM) sleep and periodic leg movements. Poor sleep does not significantly interfere with daytime alertness and sleep problems commonly diminish by late childhood, with continuing improvement through adolescence and adulthood. Sleep problems in Angelman syndrome reflect abnormal neurodevelopmental functioning presumably involving dysregulation of GABA-mediated inhibitory influences in thalamocortical interactions. Management may be difficult, particularly in young children; it primarily involves behavioral approaches, though pharmacological treatment may be required. The relationship between sleep and seizure disorder, and between sleep and learning raises critical questions, but more studies are needed to address these relationships adequately.

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