Pediatric Neurology: Chapter 87. Pediatric narcolepsy: clinical and therapeutical approaches (Handbook of Clinical Neurology)

Lecendreux Michel

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Narcolepsy occurs during childhood in combination with cataplexy in one-third of the subjects. Symptoms may develop rapidly over a few weeks or months, with excessive daytime sleepiness and cataplexy being the most dramatic and observable symptoms. It can be secondary to brain tumors or several rare diseases, but in most cases narcolepsy with or without cataplexy is a primary condition, better explained by the selective loss of hypocretin neurons in posterolateral hypothalamus. A specific HLA allele, HLA-DQB1*0602, is involved and the disease is thought to be autoimmune. Tribbles 2-specific antibodies have been identified. Recent medications have greatly improved the symptoms and wellbeing of young patients. However, these treatments are delivered off-label in the pediatric population. There is an absolute necessity for well-conducted clinical trials in order to improve treatment in children and adolescents with narcolepsy and to evaluate clinical efficacy and good tolerance of medications. Nonpharmacological approaches are certainly helpful and should be promoted systematically, especially in very young children. Narcolepsy of recent onset in children or adolescents should be considered a therapeutic emergency, even though immunotherapy is still controversial.

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