MOVEMENT DISORDERS

MOVEMENT DISORDERS AT ONSET OF NARCOLEPSY

Researchers at University of Bologna; Niguarda Hospital, Milan; and University of Rome, Italy; Montpellier, France; Stanford University, CA, USA; and Institute of Neurology, London, UK analyzed motor features in 39 children with narcolepsy with cataplexy in comparison with 25 age- and sex-matched healthy controls. Motor phenomena were "negative" and "active" (positive). Negative motor phenomena, including head drop and falls, ptosis and tongue protrusion, facial hypotonia, and generalized hypotonia, were significantly more frequent in patients compared to controls. Active motor phenomena, including eyebrow raising, tongue movements, facial grimacing, trunk swaying, stereotyped and dyskinetic-dystonic movements, were significantly more common in patients than controls. Neck extension while watching videos and puppet-like choreic movements occurred in 24% and 13%, respectively. Both negative and active motor phenomena increased significantly with emotional stimuli. Patients with narcolepsy and cataplexy (n=17) showed higher negative and active composite scores with the presence of hypotonic features on neurological examination, higher scores for active motor phenomena in the facial area (raising eyebrows, facial grimacing) and swaying of head or trunk, and more frequent neck extension and puppet-like movements. Age at onset of narcolepsy with cataplexy was inversely related to negative composite scores, and disease duration was inversely related to both negative and active composite scores. Patients with anti-streptolysin O levels of >400 showed higher composite scores for active motor features (facial and tongue movements) at baseline and during emotional stimulation, whereas negative motor features were not increased. (Piazzi G, Pizza F, Palaia V, et al. Complex movement disorders at disease onset in childhood narcolepsy with cataplexy. **Brain** Dec 2011;134:3477-3489). (Respond: Dr Giuseppe Piazzxi, Dipartimento di Scienze Neurologiche, Via Ugo Foscolo 7, 40123, Bologna, Italy. E-mail: Giuseppe.piazzi@unibo.it).

COMMENT. Cataplexy, pathognomonic of narcolepsy with cataplexy, is complicated by frequent awakenings during nocturnal sleep, periodic and non-periodic nocturnal limb movements, restless legs syndrome and rapid eye movement (REM) sleep behavior disorder. Narcolepsy with cataplexy is characterized by a loss of hypocretin-producing neurons in the postero-lateral hypothalamus, involved in the regulation of sleep and wakefulness. Childhood cataplexy presents abruptly as emotionally-triggered episodes (eg. watching cartoons, tickling), as spontaneous falls to the ground, or as generalized hypotonia with facial involvement. The choreic movements and high anti-streptolysin titers in some patients resemble Sydenham's chorea and PANDAS, autoimmune movement disorders linked to streptococcal infection. (Cardoso F et al. **Lancet Neurol** 2006;5:589-602).