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MENTAL RETARDATION SYNDROMES

SMITH-MAGENIS SYNDROME: SELF-HUGGING BEHAVIOR

A self-hugging behavior is described in 11 patients with Smith-Magenis syndrome (SMS) examined at Elwyn, PA, and the Alfred I du Pont Institute, Wilmington, Delaware. Five were children ages 7 - 16, and 6 were adults of 19 - 51 years. All had chromosome deletion 17p11.2, characteristic of SMS, and were mentally retarded, with IQs of 40-50. The involuntary, tic-like movements consisted of crossing both arms across the chest and tensing the body or clasp the hands and squeezing the arms to the sides. The spasms lasted a few seconds and occurred in series or flurries, generally accompanied by facial grimacing and occasional grunting. They were expressions of happiness, affection or positive excitement and were most pronounced during transition periods, at the beginning or end of a class, in gym or during soccer. They were not observed in temper tantrums, at times of emotional upset, or during visits to the doctor's office. In contrast to the frequent negative, aggressive, and self-injurious behaviors characteristic of SMS, self-hugging was perceived as a favorable personality trait and a behavior of value as a diagnostic marker. (Finucane BM et al. The spasmodic upper-body squeeze: a characteristic behavior in Smith-Magenis syndrome. Dev Med Child Neurol Jan 1994;36:70-83). (Respond: BM Finucane, Elwyn Inc, Elwyn, PA 19063).

COMMENT. Smith-Magenis syndrome has specific physical as well as behavioral features. These include brachycephaly, midface hypoplasia, ear malformations, and brachydactyly. Mental retardation is usually moderate. Aggression and self-mutilation, head-banging and hand-biting, and sleep disorders are the most typical behaviors, causing management problems. Self-hugging is a more benign behavior and a welcome contrast to the self avulsion of fingernails, and insertion of foreign objects into bodily orifices (*polyembolokoilomania*), as sometimes described in patients with SMS and other mental retardation syndromes (Ped Neur Briefs Jan 1994;8:10). Self-hugging was considered distinct from the stereotypical hand movements of autism and Rett syndrome and a pathognomonic sign of Smith-Magenis syndrome.

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