COMMENT. The prevalence of night wakings and sleep-onset problems is high in preschool children but symptoms lessen by 6 years of age. Parasomnias are also prevalent in early childhood and are associated with separation anxiety. Sleep terrors are associated with somnambulism, somniloquy, and frequent night wakings.

Night terrors and nocturnal frontal lobe epilepsy (NFLE). The differentiation of parasomnias and NFLE may be difficult, and the two diagnoses may co-exist. Night terrors in early childhood are sometimes followed by NFLE in school-age children. The Frontal Lobe Epilepsy and Parasomnias (FLEP) scale (Derry CP et al. **Epilepsia** 2006;47:1775-1791; Derry CP et al **Arch Neurol** 2006;63:705-709) is considered reliable in distinguishing these disorders. A video-polysomnographic recording may be necessary to confirm the diagnosis in some cases (Tinuper P et al. **Epilepsia** 2007;48:1033-1034).

CONGENITAL DEVELOPMENTAL ANOMALIES

CONGENITAL OCULAR MOTOR APRAXIA

The clinical and neuroradiological findings, and long-term intellectual prognosis in 10 patients (4 boys and 6 girls) with congenital ocular motor apraxia (COMA) are reviewed by researchers at Tottori University, Yonago, Japan. Age at first examination was 4 months to 5 years, and follow-up ranged from 4 to 37 years (mean 17.3 years). COMA was diagnosed by impaired voluntary, saccadic eye movements, and head thrusting movements in the horizontal direction. A representative patient, a boy examined at the age of 7 months and sitting had head thrust movements. He looked at objects obliquely, with eyes deviated laterally. He walked late at 22 months, but speech developed normally. At 5 years of age, he was examined because of persistent unsteady gait and abnormal eye movements. Apart from the tandem ataxia, impaired eye movements, and a left external strabismus, the remainder of the neurologic exam was normal. Brain CT showed dilatation of the fourth ventricle and hypoplasia of the cerebellar vermis. At follow-up at age 7 years, tandem gait had improved, but titubation of the head and signs of OMA, including head thrust and excessive blinking at visual tracking, persisted. At 37 years old, he is an independent, working adult.

Typical oculomotor findings of COMA (locking-up during manual spinning and impaired optikokinetic nystagmus) were elicited in all patients, excessive blinking in 5, strabismus (7), and oblique lateral glance in 8. Atypical signs, including rotatory or jerky nystagmus and limitation in upward gaze, were present in 4 patients, and were often accompanied by intellectual disabilities. Delay in walking occurred in all patients, 4 at 2 years of age and 6 walked later. Ataxia was common in the later-walking group, and was complicated by speech problems and intellectual retardation. Neuroimaging revealed dilatation of the fourth ventricle in 8, hypoplasia of the cerebellar vermis in 6, and 'molar-tooth' sign at the midbrain in 3. None had a family history of COMA, but 3 had family members with mental retardation. (Kondo A, Saito Y, Floricel F et al. Congenital ocular motor apraxia: Clinical and neuroradiological findings, and long-term intellectual prognosis. **Brain Dev** August 2007;29:431-438.) (Respond: Dr Akiko Kondo, Division of Child Neurology, Institute of Neurological Sciences, Tottori University, 36-1 Nishi-Cho, Yonago 680-8504, Japan).

COMMENT. First described by Cogan DG in 1952, congenital ocular motor apraxia (COMA) is a paresis of conjugate purposive gaze, with retained random movements, and compensatory jerking movements of the head. Optokinetic nystagmus is abnormal, with absence of the fast, refixation phase. The patient is unable to follow a fast moving object but can follow a slow moving object horizontally to either side. Jerking of the head redirects the eyes into the desired position. Repetitive blinking helps in correcting fixation. MRI is normal or may reveal agenesis of the corpus callosum, hypoplasia of the cerebellar vermis, and brainstem dysgenesis. In addition to congenital cases, the syndrome may be a sign of brain tumor, Gaucher's disease, ataxia-telangiectasia, and Joubert syndrome. Joubert syndrome and COMA are allelic for the NPHP1 gene for juvenile nephronophthisis (Betz R et al. **J Pediatr** 2000;136:828-831). The congenital type, originally described as benign, is now known to be associated often with gait ataxia, speech apraxia, behavioral and learning disabilities, and intellectual delay. The present series of COMA cases demonstrates a correlation between delay in walking ability and subsequent gait ataxia, speech problems, and intellectual retardation. Structural abnormalities of the cerebellum and brainstem are demonstrated in more than half the cases.

NEUROLOGIC ABNORMALITIES IN WILLIAMS SYNDROME

The neurologic features of 47 cases of Williams syndrome were determined and a follow-up study was performed on a subgroup of 27 subjects at the Neurorehabilitation Unit, IRCCS Eugenio Medea, Bosisio Parini, Italy. All the patients showed some neurologic deficit, but none was major, and none affected the cranial nerves or peripheral nerves. Proximal hypotonia was found in all but one (98%), and mild rigidity in 6 (12.8%). Hyperreflexia was elicited in 20 patients (42.6%), and Babinski responses in 2 (4.3%). Soft cerebellar signs were frequent: dysmetria (31.9%), dysdiadochokinesia (95.7%), tandem dyspraxia (93.6%), braking (59.6%), and gait ataxia (6.4%). Extrapyramidal signs were mild and included choreiform movements in 46.8%, dystonia in 42.6%, other involuntary movements in 36.2%, and facial grimacing in 55.3%. Cerebellar signs showed no consistent change whereas extrapyramidal signs, especially dystonia, tended to increase with age from 8 years to 14+ years, during the 4-year follow-up ($P<0.01$). The extrapyramidal abnormalities are linked to a dysfunction in the nigrostriatal dopaminergic system that increases with the accelerated ageing process, a characteristic of Williams syndrome. (Gagliardi C, Martelli S, Burt MD, Borgatti R. Evolution of neurologic features in Williams syndrome. **Pediatr Neurol** May 2007;36:301-306). (Respond: Chiara Gagliardi MD. E-mail: chiara.gagliardi@bp.Inf.it).

COMMENT. Williams syndrome is caused by a deletion in chromosome 7q11.23 and affects multiple systems, especially cardiac. The patient has an elfin-like facial appearance, infantile hypercalcemia that resolves with age, supravalvular aortic stenosis and hypertension, hyperacusis, and cognitive disorders. Nonverbal functions, visual-motor and spatial perceptions, are weak, whereas expressive language, musical abilities, and facial recognition are relative strengths. The older patient has a "cocktail personality." Less attention has been given to neurological and behavioral symptoms. Some patients with Williams syndrome (WS) have ADHD and have been treated with methylphenidate (Bowden