COMMENTARY. Childhood onset SCAs and their clinical features in addition to ataxia include dentatorubropallidoluysian atrophy (chorea, dystonia, seizures, dementia), SCA13 (mental retardation), SCA with tremor, cognitive defects, and facial dyskinesia, and Friedreich ataxia variant [1]. Testing for SCA21 is recommended in families with early onset mild or slowly progressive ataxia, particularly when associated with moderate to severe cognitive impairment. The causative gene in SCA21, TMEM240, is highly expressed in the cerebellum, dentate gyrus, putamen and caudate nucleus. The authors note that another transmembrane protein (TMEM237) is involved in Joubert syndrome-related disorders, characterized by midbrain malformation with hypoplasia of the cerebellar vermis [2].

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COGNITIVE IMPAIRMENTS IN ATAXIA-TELANGIECTASIA

Investigators from Massachusetts General Hospital, Boston, and centers in Frankfurt, Germany, examined 22 patients with the classic phenotype of ataxia-telangiectasia for neurocognitive features, and compared patients with early stage cerebellar disease (group AT-I) versus those with late stage cerebrocerebellar disease (group AT-II). Group AT-I patients scored low average compared with standard norms on all tests and were significantly impaired compared with healthy controls for verbal IQ, vocabulary and comprehension, processing speed, visuospatial processing, and working memory. Group AT-II patients scored below average on all tests for attention, working memory, and abstract reasoning. Comprehension scores were lower for patients in AT-II than in AT-I, whereas vocabulary scores showed no difference between groups. Cognitive impairments in ataxia-telangiectasia present early, coinciding with cerebellar pathology and are characteristic of the cerebellar cognitive affective syndrome. Cognitive impairments worsen in later stages of ataxia-telangiectasia, and correlate with development of supratentorial, noncerebellar pathology. (Hoche F, Frankenberg E, Rambow J, et al. Cognitive phenotype in ataxia-telangiectasia. **Pediatr Neurol** 2014 Sep;51(3):297-310).

COMMENTARY. The cerebellar cognitive affective syndrome (CCAS) associated with acquired cerebellar lesions is characterized by cognitive impairment, disorders of executive and visuospatial function, and expressive language and affective disorders. The behavioral developmental profile of patients with congenital cerebellar malformations is variable but similar to the CCAS. Malformations affecting the cerebellar vermis induce affective and social disorders, evolving to an autistic symptomatology, whereas malformations of cerebellar hemispheres are associated with selective neuropsychological deficits involving executive functions and visuospatial and linguistic abilities [1]. Patients with ataxia-telangiectasia, a neurodegenerative disorder, display a cerebellar motor phenotype during their first to third year of life and later, with involvement of noncerebellar or cerebrocerebellar circuits, the progression of cognitive and behavioral disorders is apparent. Functional neuroimaging studies during the phase of

mutism following posterior fossa tumor resection show SPECT perfusion deficits in the supratentorial areas subserving language dynamics, syntax, naming, executive functioning, affective regulation, and behavior. The clinical remission of mutism parallels improvement of frontal perfusion deficits. A theory of cerebello-cerebral diaschisis is proposed, reflecting the impact of a cerebellar lesion on supratentorial cognitive and affective functions [2].

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SEIZURE DISORDERS

MANIFESTATIONS OF ICTAL FEAR

Investigators from Okayama University and Medical Center report five pediatric patients with ictal fear followed from Jan 2003 to Dec 2012. The age of epilepsy onset ranged from 8 months to 9 years and 10 months. The ictal symptoms were sudden fright, clinging to someone nearby, and subsequent impairment of consciousness, often accompanied by complex visual hallucinations and psychosis-like complaints. In four patients, ictal fear was perceived as a nonepileptic disorder by their parents. Ictal EEG of ictal fear was obtained in all patients. Three showed frontal onset, while two showed centrotemporal or occipital onsets. Two patients were seizure-free at last follow-up, while seizures persisted in the other three. Development was normal in three and two were mentally retarded. One patient with seizure onset during infancy had a favorable outcome and was considered to have benign partial epilepsy with affective symptoms. (Akiyama M, Kobayashi K, Inoue T, Akiyama T, Yoshinaga H. Five pediatric cases of ictal fear with variable outcomes. **Brain Dev** 2014 Oct;36(9):758-63).

COMMENTARY. Although considered rare as a manifestation of epilepsy, ictal fear is mentioned in 83 references cited by PubMed since 1959. Temporal lobe epilepsy (TLE) and pathology of the amygdala are frequently involved. In surgical procedures and stimulation of the amygdala, ictal fear is frequently associated with a rising epigastric sensation, palpitations, mydriasis, and pallor [1]. Twelve (36%) of 33 patients with TLE reported ictal fear at the onset of seizures and 11 of these were seizure free postoperatively. In contrast, only 11 of 21 patients without ictal fear had a favorable outcome. Results of MR spectroscopy revealed significantly more anteriorly pronounced metabolic changes in the hippocampus of patients with, than in those without, ictal fear [2]. Ictal fear is more common in females than in males as adults, but not as children [3].

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