Fragile X-associated tremor/ataxia syndrome (FXTAS) is described in 2 sisters (Berry-Kravis E et al. Ann Neurol Jan 2005;57:144-147). The sisters were premutation carriers who developed postural tremor at 75 and 83 years of age, progressing to further disabilities with ataxia in the younger one, whose MRI showed increased signal in the cerebellar white matter and middle peduncle, and in subcortical white matter. The older patient had 2 children with FXS and cognitive disability. The father of the 2 sisters and his brother had parkinsonian tremor. Both sisters had nonrandom X-inactivation that dictated symptom severity. Symptoms of FXTAS in the women were milder than those in men in the family. This report demonstrates the association of cerebellar pathology and ataxia with a mental retardation genetic developmental syndrome.

ATTENTION DEFICIT DISORDERS

EEG THETA ACTIVITY, COGNITIVE DISORDERS AND ADHD

The relation between resting EEG theta activity and poor performance in attention-demanding cognitive tasks in 46 unmedicated children/adolescents with attention deficit hyperactivity (ADHD) was studied at Westmead Hospital, University of Sydney, NSW, Australia; and Heinrich-Heine University of Dusseldorf, Germany. Accuracy and reaction time during an auditory oddball signal detection test and a visual continuous performance test were recorded. EEG power spectral analyses revealed an increase in frontal (primarily left frontal) theta activity for ADHD subjects compared to controls (F=4.18, p<0.05). Compared with control subjects, the ADHD group had significantly delayed reaction times and decreased accuracy in both cognitive tasks. In the ADHD group, correlation analyses revealed a significant relation between accuracy on the oddball task and left frontal theta, and in controls, a significant relationship between reaction time in the continuous performance test and right posterior theta. Resting theta is interpreted as an index of inattention, and spatial neurophysiologic deficits in ADHD may be related to disturbed signal detection and inattention. (Hermens DF, Soei EXC, Clarke SD et al. Resting EEG theta activity predicts cognitive performance in attention-deficit hyperactivity disorder. Pediatr Neurol April 2005;32:248-256). (Respond: Dr Hermens, The Brain Dynamics Centre, Acacia House, Westmead Hospital, Hawkesbury Road, Westmead, NSW, 2145, Australia).

COMMENT. This report confirms the finding of increased theta activity in patients with ADHD, and suggests that the evidence may be predictive of errors in signal detection and attention. The authors comment that the localization of the increase in theta to the left frontal brain is consistent with reports of neuroimaging data showing a reduction in left frontal cerebral white matter. These data also refer to reduction in volume of right frontal gray matter (Mostofsky SH et al. Smaller prefrontal and premotor volumes in boys with attention deficit/hyperactivity disorder. Biol Psychiatry 2002;52:785-794). Previous MRI studies have shown a decreased volume of the prefrontal cortex, caudate nucleus, and globus pallidus on the right side, pointing to a dysfunction of right-sided prefrontal-striatal systems in ADHD children (Castellanos FX et al. Arch Gen Psychiatry 1996;53:607-616; Ped Neur Briefs Aug 1996).