COMMENT. Autistic spectrum disorders are characterized by impaired social interaction, restricted interests, and repetitive behaviors. Risk of ASD development in children with TSC is increased with early persistent seizure activity in the left temporal lobe, the brain region responsible for social perception and communication. Interictal epileptiform discharges in the temporal lobe, and cyst-like tubers on MRI may provide markers for ASD in TSC.

Commonly missed signs of TSC, and presenting symptoms and signs according to age group are reported from the Massachusetts General Hospital study of 243 patients with TSC (Staley BA et al. Pediatrics Jan 2011;127:e117-e125). Average age at diagnosis was 7.5 years, 81% before age 10 years (range, birth to 73 years). The most common presenting features were new onset seizures, infantile spasms, family history of TSC, cardiac rhabdomyomas, and hypopigmented macules. Missed symptoms and signs, most commonly seizures (including infantile spasms) and dermatological signs, were reported in 39% of patients. Patients with a TSC2 genetic mutation were diagnosed on average 9 years before patients with a TSC1 mutation, and were more likely to present with infantile spasms, developmental delay, or angiofibromas. Patients with a TSC1 mutation were more likely to present with a family history of TSC or hypopigmented macules. Patients with no mutation identified were more likely to present with renal angiomyolipomas. Infantile spasms are a risk factor for slow cognitive development in TSC. Immediate diagnosis of infantile spasms by EEG hypsarrhythmia and early treatment are required to reduce seizure frequency and risk of cognitive impairment. Vigabatrin is recommended for infantile spasms secondary to TSC.

MOVEMENT DISORDERS

METABOLIC BRAIN NETWORKS IN TOURETTE SYNDROME

Researchers at the Feinstein Institute for Medical Research, Manhasset, NY, studied metabolic brain networks associated with Tourette syndrome (TS) and comorbid obsessive compulsive disorder (OCD) using PET imaging and spatial covariance analysis in 12 unmedicated patients and 12 age-matched controls. An abnormal TS-related spatial covariance pattern was characterized by reduced resting metabolic activity of the striatum and orbitofrontal cortex associated with relative increases in premotor cortex and cerebellum. In TS/OCD patients, a second metabolic pattern correlated with OCD and was characterized by reduced activity of the anterior cingulate and dorsolateral prefrontal cortical regions associated with relative increases in primary motor cortex and precuneus. The OCD pattern in individual subjects was correlated with severity of OCD. Different clinical manifestations of TS are associated with 2 distinct abnormal metabolic brain networks of potential value as biomarkers for assessing response to therapy. (Pourfar M, Feigin A, Tang CC et al. Abnormal metabolic brain networks in Tourette syndrome. Neurology March 15, 2011;76:944-952). (Respond and reprints: Dr Andrew Feigin,
Center for Neurosciences, The Feinstein Institute for Medical Research, 350 Community Drive, Manhasset, NY 11030. E-mail: afeigin@nshs.edu)

COMMENT. Metabolic brain network patterns differentiate subjects with TS from controls, as well as a second pattern that differentiates TS subjects with OCD from those without OCD. The brain networks involve regions associated with motor activity as well as those regions associated with behavioral changes (anterior cingulate and prefrontal cortex).

In an editorial, Dr Katie Kompoliti of Rush Med Sch, Chicago comments that this study identifies TS-related abnormal network patterns that encompass multiple interacting nuclei instead of isolated regions, a view of the whole “elephant,” not just the trunk or tail. (Neurology March 15, 2011;76:938-939). She remarks that the study is limited by differences in gender mix of subjects (mainly male) and controls (mainly female). Other limitations include the age of subjects (all adults whose tics are usually severe), and the absence of additional TS comorbidities such as ADHD that might influence results.

AZITHROMYCIN-ASSOCIATED CHOREOATHETOSIS

Researchers at Women and Children’s Hospital of Buffalo, NY, report an 11-year-old boy with a history of developmental delay who developed transient agitation and choreoathetoid movements of upper extremities in temporal relation to treatment of influenza A and B respiratory infections with oral azithromycin on 2 occasions. Symptoms improved with brief administration of clonazepam or lorazepam, and they resolved within 36-48 hours of discontinuation of azithromycin. The association of agitation and movement disorder with azithromycin is previously unreported. A causal relation was considered probable, based on a score of 6 on the Naranjo adverse drug reaction probability scale. (Farooq O, Memon Z, Stojanovski SD, Faden HS. Azithromycin-induced agitation and choreoathetosis. Pediatr Neurol 2011;44:311-313).

(Respond: Dr Farooq, Women and Children’s Hospital, Division of Pediatric Neurology, 219 Bryant Street, Buffalo, NY 14222. E-mail: osmanfarooq@yahoo.com).

COMMENT. Macrolide antibiotics, especially clarithromycin, are known to cause acute psychoses when given with amoxicillin. A syndrome known as “Höigne syndrome” or “antibiomania” consisting of delusions, paranoia, and hallucinations is reported. One patient developed catatonia during azithromycin treatment. In the present case-report of choreoathetosis with azithromycin, the association of influenza viral infection is a possible factor in etiology.

NEUROPATHIES

PEDIATRIC SCIATIC NEUROPATHIES

Prospective review of the incidence, cause, and prognosis of pediatric sciatic neuropathy (SN) in a 30-year experience of 53 patients is reported from the Department