epilepsy developing is higher in those with FS occurring before age 1 year or after age 3 years, and with complex FS); and 10) clinical factors associated with a poor prognosis include a neurodeficit, poor response to initial therapy, and some epilepsy syndromes. (Shorvon SD, Goodridge DMG. Longitudinal cohort studies of the prognosis of epilepsy: contribution of the National General Practice Study of Epilepsy and other studies. **Brain** 2013 Nov;136(Pt 11):3497-510). (Response: Professor Simon Shorvon, E-mail: S.shorvon@ucl.ac.uk).

COMMENT. Ethics committee approval had been obtained at the beginning of the study, which did not require patient consent for collection of anonymized prognostic data. In 2007 a government appointed committee ruled that, with some exceptions, the patient's consent was required for collection of personal data from the general practitioner. The reduction in available data that followed has resulted in the premature termination of the study. The authors comment that this ill-advised decision by government appointed committees has harmed large-scale epidemiological studies in Britain, and specifically resulted in loss of 30-year follow-up of a large cohort of patients with epilepsy and reasons for a persistently high mortality.

INTRACRANIAL TUMORS

DIAGNOSIS OF INTRACRANIAL GERM CELL TUMORS

Investigators from the Massachusetts General Hospital, Boston, studied the manifestations and time to diagnosis of 70 children with germ cell tumors (GCTs) treated between 1998 and 2012. The median duration of symptoms before diagnostic MRI was 6 months (range, 2 days to 72 months). Diagnosis was delayed (>6 months) in 38 (54%). The delay increased the risk of disseminated disease. Thirty patients (43%) had nongerminomatous tumors (NGGCTs) and 40 (57%) were diagnosed with pure germinomas (PGs). The majority of primary tumors were located in the suprasellar region (28% of NGGCT and 40% of PG) followed by the pineal region (23% of NGGCT and 33% of PG). All isolated pineal region tumors occurred in male patients; suprasellar tumors occurred in females in 61%.

Symptoms of GCT were headache (69%), nausea and vomiting (50%), polyuria and/or polydipsia in 59%, double vision (34%), visual field cuts or impaired visual acuity in 27%, poor growth (17%), and premature puberty in 14%. Pineal tumors presented with symptoms of hydrocephalus, whereas suprasellar tumors caused endocrinopathies. Ophthalmic symptoms occurred in all patients: pineal located tumors caused diplopia and Parinaud syndrome symptoms in 61%, and suprasellar tumors caused visual acuity and/or visual field limitations in 29%. Patients with GCT were evaluated by a broad spectrum of pediatric specialists, and patients with delayed diagnosis were seen by 2 or more physicians and subspecialists: a neurologist in 17%, ophthalmologist 27%, or endocrinologist 34%. An endocrinopathy, especially diabetes insipidus (in 50%), was diagnosed before the diagnosis of brain tumor. Progressive enlargement of the infundibulum led to biopsy, and diagnosis was confirmed by abnormal levels of human chorionic gonadotropin in the CSF and elevated serum alpha-fetoprotein. (Sethi RV, Marino R, Niemierko A, Tarbell NJ, Yock TI, MacDonald SM. Delayed diagnosis in

children with intracranial germ cell tumors. **J Pediatr** 2013 Nov;163(5):1448-53). (Reprints: Shannon M MacDonald MD, Dept. of Radiation Oncology, MGH, 55 Fruit St, Yawkey 112, Boston, MA 02114. E-mail: smacdonald@partners.org).

COMMENT. **Bifocal germ cell tumors: synchronous tumors or metastases?** Bifocal germ cell tumors in the suprasellar and pineal regions are reported in 23 (12.8%) of 181 patients with intracranial GCTs treated at Seoul National University Children's Hospital, Korea (Phi JH, Kim SK, Lee J, et al. **J Neurosurg Pediatr** 2013 Feb;11(2):107-14). Eleven patients (47.8%) presenting with bifocal GCTs exhibited tumor seeding, compatible with bifocal lesions. Patients with bifocal germinomas show significantly shorter survival than those with germinomas from a single site. Bifocal GCTs may result from the metastatic spread of suprasellar or pineal GCTs and are a sign of disseminated disease and poor prognosis.

NEUROMUSCULAR DISORDERS

SPASMODIC MUSCLE CRAMPS AND WILSON DISEASE

Investigators at Ann & Robert H. Lurie Children's Hospital of Chicago report a case of Wilson disease (WD) in a 10-year-old-boy presenting with 3 months of increasingly severe spasmodic muscle cramps and weakness in lower extremities, upper extremities, and cramps in face and chest. Calf palpation was tender, and hyperpigmented flat lesions were present over ankles, knees, and elbows. Eye exam showed Kayser-Fleischer rings and sunflower cataracts. Creatine kinase, aspartate aminotransferase, and alanine aminotransferase were elevated, hemoglobin was low, and urinalysis revealed myoglobinuria. MRI of muscle and muscle biopsy were negative, serum ceruloplasmin was low, 24 h urine copper was elevated, and liver biopsy showed fibrosis and positive staining for copper. Rhabdomyolysis developed after the operation, attributed to the use of succinylcholine. Brain MRI showed symmetric changes in the basal ganglia. Following trientene chelation therapy for WD, symptoms and laboratory abnormalities resolved. (Rosen JM, Kuntz N, Melin-Aldana H, Bass LM. Spasmodic muscle cramps and weakness as presenting symptoms in Wilson disease. **Pediatrics** 2013 Oct;132(4):e1039-42). (Response: John M Rosen MD, Children's Mercy Hospitals and Clinics, 2401 Gillham Road, Kansas City, MO 64108. E-mail: jmrosen@cmh.edu).

COMMENT. The clinical presentation of WD or hepatolenticular degeneration is variable and a number of different syndromes are recognized in addition to the classical syndrome described by Wilson in 1912. The classical syndrome, in children of 10 to 15 years, presents with bulbar symptoms. Rigidity of skeletal musculature follows, and an extrapyramidal type of hypertonus resembling that of the Parkinsonian syndrome is associated with a constant tremor. Tendon reflexes are usually normal. Any stimulus provokes spasms, and a diagnosis of tetanus may be entertained. Myotonia is also described. (Ford FR. **Diseases of the Nervous System. In Infancy, Childhood and Adolescence.** 4th ed. Springfield, IL: Charles C Thomas; 1960. p. 756-62.). The Rosen, Kuntz et al case-report describes a WD syndrome with muscle cramps.