VACCINE-RELATED ENCEPHALOPATHY REDIAGNOSED AS DRAVET SYNDROME

Five patients with a history of seizures in the first year of life, occurring within 24 hours of DPT vaccination, were subsequently tested positive for *SCN1A*, and with support of genetic testing were diagnosed with Dravet syndrome, in a report from San Antonio Military Medical Center; Northwestern University School of Medicine, Children's Memorial Epilepsy Center, Chicago; Baylor College of Medicine, and Texas Children's Hospital, Houston, TX. Recurrent seizures in the first year of life were febrile and afebrile, generalized and unilateral, and often prolonged. After 1 year, seizures were often partial, myoclonic, and atypical absence. They were intractable to medication. Later, patients were often developmentally delayed, cognitively impaired and sometimes autistic. MRI was negative. The case report is intended to raise awareness of Dravet syndrome and aid in the early recognition and diagnosis. (Reyes IS, Hsieh DT, Laux LC, Wilfong AA. Alleged cases of vaccine encephalopathy rediagnosed years later as Dravet syndrome. **Pediatrics** Sept 2011;128:e699-e702). (Respond: David T Hsieh MD, Division of Child Neurology, San Antonio Military Medical Center, 3851 Roger Brooke Dr, Fort Sam Houston, TX 78234. E-mail: david.hsieh@us.af.mil).

COMMENT. Dravet syndrome is a rare genetic epileptic encephalopathy, recognized by ILAE as severe myoclonic epilepsy of infancy, and associated with mutations in *SCN1A* gene (neuronal sodium channel). Seizure onset is in the first year of life, and is often provoked by fevers. In the above cases, the close temporal relation of the pertussis vaccination to the first seizure may indicate a triggering effect (McIntosh AM et al. **Lancet Neurol** 2010;9(6):592-598). Fever alone is an unlikely precipitating factor, since the first seizure was afebrile in 2 of the 5 patients. Patients with seizures that have a close temporal relation to vaccination and are followed by frequent recurrences should be tested for *SCN1A* mutation. Mutations in the *SCN1A* gene are found in up to 80% of cases of Dravet syndrome. (Korff C et al. **J Child Neurol** 2007;22(2):185-194).

METABOLIC DISORDERS

MRI FINDINGS AND OUTCOME IN MOLYBDENUM COFACTOR DEFICIENCY

Researchers at Great Ormond Street Hospital, London, UK examined the clinical, brain MRI, biochemical, genetic, and EEG features and outcome in 8 children with a diagnosis of molybdenum cofactor deficiency seen over a 10-year period. Six neonates had an early (classical) onset with predominantly epileptic encephalopathy and neonatal seizures, and 2 had a late (atypical) onset with global developmental impairment. Four were boys. Neonatal history was normal except in one infant with presumed HIE who required ventilatory support for 3 days. Age at presentation ranged from 1 day to 24 months. Clinical features leading to diagnosis included progressive microcephaly, dysmorphisms, progressive pyramidal and extrapyramidal signs, axial hypotonia, kyphoscoliosis, verbal dyspraxia, and visual impairment due to cortical damage or lens