1.35/1000 person-years. (see Ped Neur Briefs Jan 1995;9:1 for review and commentary). Cardiac causes for idiopathic seizures and a normal EEG, a possible explanation for some cases of SUD, especially in adolescent males, should always be considered when AEDs are ineffective.

INFANTILE SEIZURES

INFANTILE SPASMS IN DOWN SYNDROME

The clinical characteristics, EEG abnormalities, response to therapy, and outcome of 14 patients with infantile spasms and Down syndrome were studied at the Hopital Saint Vincent de Paul, Paris (9 cases); Universita Degli Studi de Pisa, Italy (2 cases); and Hopital de La Timone, Marseille, France (3 cases). None had antecedent cardiopathy or perinatal hypoxia. Spasms began between 4 and 18 months (mean 8 months), development was delayed before seizure onset, and visual contact deteriorated after seizure onset. Interictal EEGs showed typical hypsarrhythmia with no focal abnormality. Hydrocortisone (15 mg/kg/day for 2 weeks, and discontinuation over 2 weeks) in 10, and vigabatrin, valproate, or pyridoxine in 4 patients, controlled spasms and hypsarrhythmia within 6 months. Five with relapses within 2 months responded to further treatments. Seven remained seizure-free, and 7 developed other types of seizures resembling idiopathic generalized epilepsies, including myoclonic jerks, absences, or generalized atonic or tonic-clonic seizures, most responding readily to a combination of valproate, ethosuximide, and diazepam. None developed Lennox-Gastaut syndrome or other chronic refractory seizure disorder. Autistic features persisted in 2. (Silva ML, Cieuta C, Guerrini R, Plouin P, Livet MO, Dulac O. Early clinical and EEG features of infantile spasms in Down syndrome. Epilepsia Oct 1996;37:977-982). (Reprints: Dr O Dulac, Hopital Saint Vincent de Paul, 82 Ave Denfert Rochereau, 75674 Paris Cedex 14, France).

COMMENT. Infantile spasms in Down syndrome have the ictal and interictal EEG characteristics of idiopathic West syndrome, they respond relatively well to therapy, and do not generally evolve into Lennox-Gastaut or other chronic epilepsy syndrome. A delay in diagnosis may contribute to a worsening of cognitive dysfunction, and parents of Down syndrome children should be alerted to the possible development of spasms in the first year.

A case of West syndrome as the initial manifestation of congenital unilateral perisylvian cortical dysplasia is reported from University Children's Hospital, Badajoz; and Galicia General University Hospital, Santiago de Compostela, Spain (Vaquerizo-Madrid J, Eiris-Punal J, Gomez-Martin H et al. Acta Neuropediatr 1996;2:132-138). The child had left hemiatrophy and paresis and was developmentally delayed. Later, he had refractory epilepsy, with complex partial, atypical absence, and atonic seizures. The interictal EEG during sleep showed right sided epileptogenic activity with contralateral spread.

PYRIDOXINE-DEPENDENT SEIZURES AND MRI ABNORMALITY

An infant with pyridoxine-dependent seizures and MRI, PET, and EEG evidence of diffuse structural or functional brain disease is reported from the University of New Mexico Health Sciences Center, Albuquerque, NM, and the UCLA School of Medicine, Los Angeles, CA. Seizures began at 10 weeks, and status epilepticus occurred four times between 3 and 7 months of age. Trials of AEDs and ACTH were partially effective, but he became encephalopathic and