

## SEIZURE DISORDERS

### **ABSENCE EPILEPSY WITH FAST RHYTHMIC ATYPICAL EEG**

The medical files of 31 patients with absence epilepsy (AE) were reviewed at La Timone University Hospital, and Henri Gastaut/Saint Paul Hospital, Marseilles, France. Five having an atypical EEG pattern with fast rhythmic spikes (10-15 Hz) during slow-wave sleep were selected for further study. Age at onset of absence seizures was 3 to 12 years. Four developed generalized tonic clonic seizures with onset at 11 to 16 years. All had borderline intelligence, with social and learning handicaps. Neuroimaging was normal. AE was classified as juvenile absence epilepsy in 2. EEG recordings during absence seizures showed generalized spike or polyspike waves of 3-4 Hz. Sleep EEG showed fast rhythmic discharges during stage II slow-wave sleep, without clinical manifestations on video. Seizures were mostly refractory to valproate and lamotrigine, and one patient died a sudden unexplained death at 3 year follow-up. (Guye M, Bartolomei F, Gastaut JL, Chauvel P, Dravet C. Absence epilepsy with fast rhythmic discharges during sleep: an intermediary form of generalized epilepsy? *Epilepsia* March 2001;42:351-356). (Reprints: Dr F Bartolomei, Service de Neurophysiologie Clinique et Unite d'Epileptologie Clinique, Chu Timone, 264 rue Saint Pierre, 13385 Marseille CEDEX 05, France).

**COMMENT.** Children presenting with absence epilepsy associated with fast rhythmic spikes in sleep EEG are at risk of a relatively poor outcome, with development of generalized tonic clonic seizures, resistance to antiepileptic drugs, and learning handicaps. The authors classify these cases as a separate clinical entity, intermediary between idiopathic and cryptogenic/symptomatic generalized epilepsies, and different from the Lennox-Gastaut syndrome that also exhibits fast discharges during slow-wave sleep in the EEG. Previous reports have recognized the risk of poor outcome of idiopathic generalized epilepsies associated with polyspike waves and fast rhythmic discharges in slow wave sleep. (Gibbs FA, Gibbs EL, 1952; Lennox WG, 1960; Lugaesi E et al, 1974; Degen R, Rodin E, 1991; Michelucci R et al, 1995).

### **EEG IN WOLF-HIRSCHHORN/PITT-ROGERS-DANKS SYNDROMES**

A characteristic electroclinical pattern is described in a child with Pitt-Rogers-Danks syndrome (PRDS) and in 14 reports of Wolf-Hirschhorn syndrome (WHS) reviewed at the Division of Medical Genetics, Centre Hospitalier Universitaire Vaudois, Lausanne, Switzerland. Both syndromes are caused by deletions of the short arm of chromosome 4.

A 13 month-old female child presented with persistent growth failure and developmental delay. She could not sit and exhibited stereotyped hand-wringing movements. Dysmorphic features included a triangular face, prominent eyes, hypertelorism, and micrognathia. A relative macrocephaly (50th percentile) contrasted with a height and weight below the 3rd percentile. At reevaluation at 3 years, the developmental level was at 18 months, and she had tonic and generalized myoclonic seizures, controlled by valproate. In video-EEG recordings, 2-3 Hz high-voltage spike-wave bursts were associated with myoclonic jerks, and some without clinical manifestations. The bursts were localized over the centro-occipital area and became diffuse and generalized in sleep. The parents karyotypes were normal. (Zanki A, Addor M-C, Maeder-Ingvar M, Schorderet DF. A characteristic EEG pattern in 4p-syndrome: case report and review of the literature. *Eur J Pediatr* Feb 2001;160:123-127). (Respond: Dr Andreas Zanki, Institut fur