

PEDIATRIC NEUROLOGY BRIEFS

A MONTHLY JOURNAL REVIEW

J. GORDON MILLICHAP, M.D., F.R.C.P., EDITOR

Vol. 22, No. 2

February 2008

SEIZURE DISORDERS

CHILDHOOD OCCIPITAL EPILEPSY OF GASTAUT

The electroclinical features and evolution of childhood occipital epilepsy of Gastaut (COE-G) are analyzed in a study of 33 patients identified and followed between 1990 and 2007 at the Hospital Nacional de Pediatría, Buenos Aires, Argentina. In comparison, over the same 16-year period, 201 children with Panayiotopoulos syndrome (PS) and 418 with benign childhood epilepsy with centrotemporal spikes (BCECTS) were registered. Age at onset of COE-G ranged from 4 to 16 years, with a mean of 8.5 yrs. Febrile seizures occurred in 5 patients (15%), migraine in 3 (9%), and BCECTS in 2 (6.5%). Family history was positive for epilepsy in 21%, febrile seizures (21%), and migraine (12%). Visual hallucinations were the initial seizure manifestation in 27 (82%). Blindness was the presenting symptom in 17 (52%) and the only clinical symptom in 5 patients. Postictal blindness and hemianopsia occurred in 7 patients (21%). Deviation of the eyes and ipsilateral turning of the head followed the visual hallucination in 20 patients (60%), and hemiconvulsions occurred in 45%. Eyelid closure and blinking occurred in 6 (18%). Migraine manifestations were prominent in 16 patients (48%). The duration of seizures was usually brief and <1-2 min. Seizures occurred while awake but sometimes in sleep. Frequency varied from 7 per week to one every 6 months. EEG showed occipital spike-wave paroxysms with eyes closed, disappearing with eyes open. All received antiepileptic treatment with valproic acid (15), carbamazepine (8), or oxcarbazepine (4). Seizures remitted within 2 to 7 years (mean, 4 yr) after onset in 80%, while EEG abnormalities persisted in 38%. AEDs were discontinued without relapse after 2-4 years in 54%. (Caraballo RH, Cersosimo RO, Fejerman N. Childhood occipital epilepsy of Gastaut: a study of 33 patients. *Epilepsia* Feb 2008;49(2):288-297). (Reprints: Dr Roberto H Caraballo, Department of Neurology, Hospital de Pediatría, Prof Dr Juan P Garrahan, Combate de los Pozos 1881, CP 1245,

PEDIATRIC NEUROLOGY BRIEFS (ISSN 1043-3155) © 2008 covers selected articles from the world literature and is published monthly. Send subscription requests (\$68 US; \$72 Canada; \$75 airmail outside N America) to **Pediatric Neurology Briefs - J. Gordon Millichap, M.D., F.R.C.P.-Editor**, P.O. Box 11391, Chicago, Illinois, 60611, USA. The editor is Pediatric Neurologist at Children's Memorial Hospital and Professor Emeritus, Northwestern University Medical School, Chicago, Illinois.

PNB is a continuing education service designed to expedite and facilitate review of current scientific information for physicians and other health professionals. Fax: 312-943-0123.

Buenos Aires, Argentina. E-mail: rhcarballo@amct.com.ar.

COMMENT. Childhood occipital epilepsy (COE) of Gastaut is manifested by brief seizures, mainly visual hallucinations, illusions or amaurosis, followed by hemiconic seizures while awake, postictal migraine headaches, mean age at onset of 8.9 years, and interictal occipital spike-wave EEG paroxysms that attenuate when eyes are opened. Prevalence is estimated at 0.2-0.9% of epilepsies, and 2-7% of benign childhood focal seizures. Gastaut reported the syndrome in 1982, and it was accepted as an entity by the ILAE in 1989. Despite some recent loss of recognition, the present authors consider the syndrome as a rare but well-defined entity within the group of idiopathic simple partial (focal) epilepsies of childhood. Differential diagnoses include symptomatic occipital epilepsy, migraine with aura, and basilar migraine. Among the idiopathic COEs, the Gastaut type is of late onset and associated with visual symptoms, whereas Panayiotopoulos syndrome is of early onset and characterized by autonomic symptoms (ictal vomiting). The EEG findings alone are not diagnostic of COE. Not all children with occipital spikes develop seizures. Those with COE should be differentiated from patients with occipital spikes occurring both with eyes open and closed, and unassociated with clinical seizures. Occipital spikes also occur in children with myoclonic, absence, and photosensitive epilepsies. (Browne TR, Holmes GL Handbook of Epilepsy. Philadelphia. Lippincott, 2004;86-87).

RISK FACTORS FOR FEBRILE SEIZURE RECURRENCE

Factors that predict recurrence of febrile seizures (FS) were determined in a prospective study of 260 children age 3 months to 6 years followed for a median of 4.3 years after the first FS at Ippokratio Hospital, Aristotle University of Thessaloniki, Greece. The median age of patients at onset of study was 16.5 months (range 3 months to 5.8 years). Final reevaluation was at a median of 6.0 +/- 1.5 years. The sex ratio was 139 boys to 121 girls (1.15 : 1), and recurrence was not higher among boys than girls. Overall recurrence rate was 40.4%. Cumulative recurrence was 24.2% at 6 months, 34.2% at 12 months, 38.1% at 18 months, and 40.4% at 6 years. EEG abnormalities in 12 children at baseline were not significantly associated with FS recurrence. Low age at onset and positive family history of FS, especially maternal, were strong predictors of recurrence. Other risk factors included abnormal perinatal history with low Apgar score and NICU care for >3 days, low temperature (38.9C or below) and short (<12 hrs) duration of fever before initial FS, a history of frequent febrile illness ($p < 0.0001$), focal FS, and recurrence within the same febrile illness. Duration >15 min of first FS was not a factor. Two or more recurrences occurred in 48%; 28 had only 2 and 22 (44%) had 3 or more recurrences. Factors predisposing a child with one recurrence to a second or more are young age at onset and family history of FS ($p < 0.001$). Multiple recurrences were correlated with low temperature elevation ($\leq 38.9C$) before initial FS. By multivariate analysis significant risk factors were early age at onset, complex first FS, and family history of FS ($p < 0.05$). (Pavlidou E, Tzitiroidou M, Kontopoulos E, Panteliadis CP. Which factors determine febrile seizure recurrence? A prospective study. *Brain Dev* Jan 2008;30:7-13). (Respond: Dr Christos P Panteliadis, Department of Paediatric Neurology, Ippokratio Hospital, Aristotle University of Thessaloniki, Greece. E-mail: cpantel@hol.gr).