

FAMILIAL OCCURRENCE IN NEWLY DIAGNOSED EPILEPSY

The familial occurrence of epilepsy among 462 children with newly diagnosed multiple unprovoked seizures was studied at the Leiden University Medical Centre, and other hospitals in the Netherlands. Epilepsy was classified as generalized in 57% of the probands, localization-related in 42%, and undetermined in 1%. Compared to the total group, generalized epilepsies were more common (77%) and localization-related epilepsies less common (23%) in the 10% of cases found to be familial. Epilepsy occurred in 58 first-degree and 21 other relatives. The majority (83%) of first degree relatives with idiopathic or cryptogenic epilepsy had the same seizure type as the proband. (Callenbach PMC, Geerts AT, Arts WFM, et al. Familial occurrence of epilepsy in children with newly diagnosed multiple seizures: Dutch study of epilepsy in childhood. Epilepsia March 1998;39:331-336). (Reprints: Dr OF Brouwer, Department of Neurology, Leiden University Medical Centre, PO Box 9600, 2300 RC Leiden, The Netherlands).

COMMENT. Epilepsy is familial in 10% of newly diagnosed cases in childhood. Those with a positive family history are most likely to have generalized, idiopathic or cryptogenic seizures. The relatives of familial cases usually had the same seizure type as the newly diagnosed child. The role of genetic factors in the pathogenesis of idiopathic generalized childhood epilepsy is supported by this study.

LAMOTRIGINE-INDUCED AGGRESSIVE BEHAVIOR

The response and behavioral profiles of 19 intellectually impaired adults, aged 17-54 years, following treatment with lamotrigine (LTG) as adjunctive therapy for uncontrolled epilepsy are reported from the Epilepsy Research & Services, Chatswood, Australia. Aggressive behavior and violence were provoked in 9 (47%) patients within less than one month of the introduction of LTG, and 5 required complete withdrawal of the drug. Changes in behavior ranged from shouting, slamming doors, or knocking over furniture to attacks on staff and other residents of the institutions. (Beran RG, Gibson RJ. Aggressive behaviour in intellectually challenged patients with epilepsy treated with lamotrigine. Epilepsia March 1998;39:280-282). (Reprints: Dr RG Beran, Epilepsy Research & Services, Suite 5, 6th Floor, 12 Thomas St, Chatswood NSW 2167, Australia).

COMMENT. The behavior of intellectually impaired patients with epilepsy should be closely monitored when lamotrigine is used as adjunctive therapy for uncontrolled seizures. Violent aggression may be provoked and may require withdrawal of the drug.

MENTAL RETARDATION SYNDROMES

ANGELMAN SYNDROME WITHOUT CHROMOSOME ANOMALY

The clinical manifestations of Angelman syndrome (AS) in 12 patients without a cytogenetic or molecular defect and 28 with a deletion were compared at the Department of Neurology, Leiden University Medical Center, and other hospitals in the Netherlands. Two minor differences were uncovered: mandibular prognathism was present in 100% of patients with a defect and 58% of those without; truncal hypotonia occurred in only 54% of the group with a genetic defect and in 91% of those without. All other characteristics, including seizures and EEG abnormalities, occurred with the same frequency in both groups. The