Phenytoin may exacerbate myoclonus whereas treatment with benzodiazepines and valproate is most effective. (Andermann E et al. Classification of progressive myoclonus epilepsies and related disorders. Marseille Consensus Group, Ann Neurol July 1990; 28:113-116).

COMMENT Myoclonus and its relation to monoamine, GABA and other receptors is reviewed from the Departments of Neurology and Pediatrics,University of Southern California School of Medicine, Los Angeles, CA. (Snodgrass SR. FASEBJ July 1990; 4:2775-2788). The term myoclonus was first used in 1881 by Freidreich with reference to a progressive movement disorder of a gradual onset in a middle-aged man. By 1891 Unverricht had written a book on myoclonus, describing a family with myoclonus and progressive deterioration. Snodgrass, in the present article, classifies myoclonus in four categories: a) stimulus-sensitive myoclonus, b) stimulus-insensitive myoclonus, c) sleep myoclonus, and d) asterexis and negative myoclonus. Distinction between epileptic and nonepileptic myoclonus is often difficult. Posthypoxic action myoclonus (Lance-Adams syndrome) is associated with reduced CSF levels of 5-HIAA, the serotonin metabolite, and 50% respond to treatment with 5-HTP. Benzodiazepines and valproic acid are the most useful drugs for patients unresponsive to HTP. Various animal models of myoclonus are described. (Gundlach AL. FASEB J July 1990; 4:2761-2766).

"Severe myoclonic epilepsy of infancy" is discussed by Hurst DL (Epilepsia July/August 1990; 31:397-400) from the Department of Medical and Surgical Neurology, Texas Tech University, Lubbock, Texas. The central features of this syndrome include: 1) normal development before the onset of seizure activity, 2) repeated prolonged febrile seizures, 3) later onset of mixed/myoclonus epilepsy, 4) developmental slowing with onset of seizure activity, and 5) evolving EEG abnormalities. The incidence of the syndrome is estimated at one in 40,000 children. Dravet, who first described the syndrome in 1978, found a 30% prevalence in a group of 142 children with myoclonic epilepsy. A more vigorous, rather than conservative, approach to the management of complex febrile seizures might be suggested by this report.

RASMUSSEN ENCEPHALITIS

Immunological abnormalities associated with chronic encephalitis, epilepsy, and progressive hemiplegia in a three-year, nine-month old girl are reported from the University of Utah School of Medicine and the Primary Children's Medical Center, Salt Lake City, Utah. The child developed simple partial seizures refractory to medications and associated with a contralateral hemicerebral atrophy. Immunologic abnormalities were indicated by elevated antinuclear antibody, CSF oligoclonal bands and elevated immunoglobulin G (IgG). A right subtotal hemispherectomy resulted in control of the seizures. Pathological study showed widespread cerebral vasculitis and severe

cortical atrophy with marked neuronal loss. (Andrews JM et al. Chronic encephalitis, epilepsy and cerebrovascular immune complex deposits. Ann Neurol July 1990; 28:88-90).

COMMENT: Immunofluorescence microscopy revealed granular accumulation of IgG, IgM, IgA within the cerebral vessels of this patient suggesting an immune complex disease as the basis for Rasmussen's syndrome.

SERUM HORMONES AND ANTICONVULSANTS

Circulating sex and thyroid hormones were assessed in 63 male young adults with epilepsy in the Departments of Neurology and Clinical Chemistry, University of Oulu, Finland. All therapeutic regimens that included carbamazepine and/or phenytoin were associated with low levels of circulating thyroxine (T4), free thyroxine (FT4), and dehydroepiandrosterone sulfate, and low values for the free androgen index. Phenytoin, alone or combined with carbamazepine, was associated with high serum concentrations of sex hormone - binding globulin. Hormone values were unaffected by valproate monotherapy, but the combination of carbamazepine plus valproate had the most marked effect on serum thyroid hormone levels and the free androgen index. Serum T3 concentrations were unaffected by any of the medications or combinations. Serum thyrotropin concentrations were not elevated despite low serum thyroid hormone levels. (Isojarvi JIT et al. Serum hormones in male epileptic patients receiving anticonvulsant medication. Arch Neurol June 1990, 47:670-676).

COMMENT This study supports the hypothesis that an increased metabolism of thyroid hormones in the liver is the main reason for decreased T4 and FT4 serum levels in epileptic patients receiving carbamazepine and/or phenytoin treatment. The combination of valproate and carbamazepine has the most marked effect on thyroid hormone balance and both drugs are highly bound to serum proteins. Valproate displaces T4 from its binding sites on plasma proteins thereby leading to a larger amount of T4 subject to the liver enzyme inducing properties of carbamazepine. Thyroid supplements are usually not required in patients with a low serum T4 associated with anticonvulsant drug therapy. If the free T3 and T4 are normal thyroid supplements should be withheld.

To avoid criticism of a gender bias, a reference is included to "Serum steroid hormones and pituitary function in female epileptic patients during carbamazepine therapy" (Iojarvi JIT. Epilepsia Aug 1990; 31:438-445). In 13 female epilepsy patients receiving long-term carbamazepine (CBC) monotherapy, serum sex hormone binding globulin levels increased and dehydroepiandrosterone sulfate levels decreased during CBC treatment. Increased metabolism of steroid hormones caused by liver enzyme inducing proprerties of CBC and a direct inhibitory