PEDIATRIC NEUROLOGY BRIEFS A MONTHLY JOURNAL REVIEW

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

Vol. 12, No. 1

January 1998

INFECTIOUS DISORDERS

CHRONIC PROGRESSIVE ENCEPHALITIDES

An excellent review of chronic progressive encephalitides of childhood (CPEC), with particular emphasis on the prototypical disease, subacute sclerosing panencephalitis (SSPE), is presented from the Institute for Research in Childhood Neurodegenerative Diseases, and University of South Alabama, Mobile, AL. Causes of CPEC include viral, fungal, protozoan, and prion infective agents. They are classified as follows: 1) slow virus diseases: 2) latent childhood-activated subtypes: 3) opportunistic viral encephalitides; 4) uncertain causes of presumed viral origin. Clinical, etiological, and therapeutic aspects of SSPE are discussed, and the four stages outlined in detail: Stage I, behavioral changes, including hyperactivity and attention deficit (weeks to years); stage II, myoclonia (3 to 12months); stage III, choreoathetosis and dementia (3 to 18 months); stage IV, vegetative state, and rapid weight loss. Progression may be acute, subacute or chronic. Diagnosis is confirmed by CSF examination for elevated measles antibody, and >1:8 complement fixation. MRI may be normal in stages I and II, and shows diffuse atrophy in later stages. EEG generalized pseudoperiodic S/W complexes every 5 to 10 seconds are characteristic of stage II. Interferon by intraventricular injection, sometimes combined with isoprinosine, has some reported benefits but much neurotoxicity. An international multicenter trial is in progress. (Dyken P, Philippart M, Maertens P. The chronic progressive encephalitides of childhood. Neurological Infections and Epidemiology 1997;2:145-158), (Respond: Dr Paul Dyken, Institute for Research in Childhood Neurodegenerative Diseases, PO Box 70191, Mobile, AL).

COMMENT. Due to effective immunization against measles, SSPE is relatively rare in the USA, most cases presenting in immigrants from countries lacking the mandatory infant and preschool vaccination. In 1987, less than 5% of children diagnosed with SSPE in the UK had received measles immunization. (see <u>Progress</u> in <u>Pediatric Neurology I</u>, PNB Publishers, 1991;pp420-421).

THALAMIC SYNDROME AND MEASLES INFECTION

Four young children who presented with acute thalamic syndrome within a

PEDIATRIC NEUROLOGY BRIEFS (ISSN 1043-3155) © 1998 covers selected articles from the world literature and is published monthly. Send subscription requests (558 US; S60 Canada; \$68 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.

PNB is a continuing education service designed to expedite and facilitate review of current scientific information for physicians and other health professionals.

The editor is Pediatric Neurologist at Children's Memorial Hospital and Northwestern University Medical School, Chicago, Illinois.