

lipidosis. **Folia Neuropathologica** 2012;50(4):330-45). (Response: Harvey Sarnat, MD, FRCP, Alberta Children's Hospital, 2888 Shaganappi Trail, NW, Calgary, AB T3B 6A8, Canada. E-mail: Harvey.sarnat@albertahealthservices.ca).

COMMENT. Hemimegalencephaly is a hamartomatous dysgenesis, sometimes associated with other neurocutaneous syndromes, especially epidermal nevus and Proteus syndromes. Infants with Proteus syndrome [alt: Wiedemann syndrome] are normal at birth and develop skin tumors and bone growths with increasing age, especially involving the skull and soles of feet. Neurological involvement in rare reports of Proteus syndrome includes HME, Ohtahara syndrome, syringomyelia, arachnoid cyst, craniocutaneous lipomatosis, vascular malformation, and meningioma. (Bastos H, da Silva PF, de Albuquerque MA, et al. **Seizure** 2008 Jun;17(4):378-82). (Opitz JJ. Hamartoma syndromes, exome sequencing, and a protean puzzle. **N Engl J Med** 2011 Aug 18;365(7):661-3). Proteus syndrome is caused by a mutation in AKT1, and is classed as a genetic mosaicism. A knowledge of neurological syndromes helps in the selection of diagnostic tests and our understanding of the cause of refractory epilepsies.

MORPHOLOGICAL VARIATIONS OF HIPPOCAMPAL FORMATION IN EPILEPSY

Researchers at Hospital Sao Paulo and other centers in Brazil compared the hippocampal formation (HF) morphology of healthy asymptomatic individuals (n=30) with that of patients with mesial temporal lobe epilepsy and hippocampal sclerosis (MTLE-HS)(n=68), of patients with malformations of cortical development (MCD)(n=34), and of patients with morphological HF variations without other structural signs (pure MVHF)(n=12). Morphological variations of HF were significantly more frequent in patients with MCD than in patients with MTLE-HS or in normal individuals. Febrile seizures occurred only in patients with MTLE-HS, supporting the hypothesis that febrile seizures cause the MTLE-HS. Refractory epilepsy is more associated with abnormalities like hippocampal sclerosis or malformations of cortical development than with variations of the hippocampal formation itself. Patients with pure morphological variations of the hippocampal formation showed higher incidence of extratemporal seizure onset. (Hamad APA, Carrete H Jr, Bianchin MM, et al. Morphological variations of hippocampal formation in epilepsy: Image, clinical and electrophysiological data. **Epilepsy & Behavior** 2013 Jan;26(1):67-70). (Response: Dr Hamad. E-mail: anahamad@gmail.com).

COMMENT. The authors conclude that morphological variations of hippocampal formation (HF) are rare in patients without seizures and that hippocampal formation malrotation is probably pathological. The development of HF is complete after 18 weeks gestation and is similar to the adult HF after 30 weeks gestation. Some authors suggest that febrile seizures might lead to hippocampal sclerosis or to hippocampal formation abnormalities but not to epilepsy. (Auer T, Barsi P, Bone B, et al. History of simple febrile seizures is associated with hippocampal abnormalities in adults. **Epilepsia** 2008 Sep;49(9):1562-9). The relationship between HF dysmorphism, febrile seizures and hippocampal sclerosis is still unclear.