hypertension may be an unrecognized risk factor for cerebrovascular disease. In a Chinese study only 4 of 251 patients with stroke had hypertension. Hypertension is an identified factor in stroke with sickle cell disease, but with this exception, there is no definite consensus for an association of hypertension and ischemic stroke in children. Hemorrhagic stroke with hypertension was reported in 15 patients with sickle cell disease, and 6-8% of hemorrhagic stroke in children were associated with hypertension. *Posterior reversible encephalopathy syndrome* (PRES) arises with acute severe hypertension and is characterized by headaches, blindness, confusion, and seizures, and evidence of reversible posterior leukoencephalopathy with edema on MRI. The cause is a breakdown of autoregulation and endothelial dysfunction.

Chronic effects of hypertension include cognitive dysfunction affecting attention, learning and memory, reversible with antihypertensive treatment. Hypertension might predispose to endothelial damage and atherosclerosis, as suggested by carotid ultrasound measurements of intima-media thickness, a biomarker of hypertensive vascular damage. Retrospective review of neurological symptoms in 409 children with hypertension aged 7-17 years, and compared with 150 healthy controls, showed 54% had significantly more complaints of pain, sleep disturbance, fatigue, or lack of concentration. Headache incidence of 42% before antihypertensive treatment fell to 6-9% after treatment. (Croix B, Feig DI. Childhood hypertension is not a silent disease. **Pediatr Nephrol** 2006;21:527-532). Future studies should standardize the definition and monitoring of hypertension in children, investigate transcranial doppler changes correlated with cognitive decline, and investigate the value of MRI in definition of subtle infarcts and small vessel disease. (Sharma M, Kupferman JC, Brosgol Y et al. The effects of hypertension on the paediatric brain: a justifiable concern. **Lancet Neurol** Sept 2010;9:933-940). (Respond: Prof SG Pavlakis, Maimonides Infants and Childrens' Hospital, Brooklyn, NY 11219. E-mail: spavlakis@maimonidesmed.org).

COMMENT. The authors find pediatric hypertension is underdiagnosed and except for hypertensive encephalopathy, the neurological effects are under-recognized. With the increased incidence of obesity and diabetes type 2 in children, hypertension has become more prevalent. Patients at risk of hypertension should receive more frequent monitoring, so that chronic effects of hypertension on cognition and memory may be avoided.

## SEIZURE DISORDERS

## INFANTILE SPASMS TREATED WITH THE KETOGENIC DIET

Researchers at Johns Hopkins Hospital, Baltimore, MD have evaluated the efficacy of the ketogenic diet in the treatment of 104 consecutive infants with infantile spasms (IS) and hypsarrhythmia on EEG. The cohort included 23 of initial patients seen, 13 new-onset IS, and 68 additional patients. The etiology was symptomatic in 74 (71%), and previous therapy had included a mean of 3.6 anticonvulsants, with corticosteroids or vigabatrin in 71%. Diet efficacy was assessed through patient clinic visits at 3, 6, 9, 12, and 24 months, and by telephone and e-mail contact. Developmental progress was based on parental reports and clinical examination. Follow-up EEG was obtained after 6-12

months in children with intractable IS and 2-4 weeks in those with new-onset IS. Mean age at diet onset was 1.2 years, and diet duration was a mean of 1.3 years. An initial 3.1:1 or 3.5:1 ratio of fat to carbohydrate and protein was used in 68 (65%) patients; a 4:1 ratio was started in 16 of older age patients. All patients fasted prior to diet initiation. Parents kept patient records of daily seizure frequency, weekly weight, and biweekly urine ketones. Intent-to-treat analysis showed >50% control of spasms in 64% at 6 months and 77% after 1-2 years. Complete control for at least 6 months within a median of 2.4 months was achieved in 38 (37%). Improvement in development occurred in 62%, EEG improvement in 35%, and reduction in current anticonvulsants in 29%. Adverse effects developed in 33%, with diminished linear growth in 6%. Older age at onset of IS and fewer prior anticonvulsants were associated with >90% spasm reduction at 6 months. In this prospective trial, two thirds of patients with IS treated with the ketogenic diet were benefited, and the diet is strongly recommended for patients who have failed to respond to corticosteroids and vigabatrin. (Hong AM, Turner Z, Hamdy RF, Kossof EH. Infantile spasms treated with the ketogenic diet: Prospective single-center experience in 104 consecutive infants. **Epilepsia** Aug 2010;51(8):1403-1407). (Respond: Eric H Kossof MD, The Johns Hopkins Hospital, Baltimore, MD 21287. E-mail: ekossof@jhmi.edu).

COMMENT. Nordli DR Jr and associates at Children's Memorial Hospital, Chicago, have previously demonstrated the efficacy and safety of the ketogenic diet in the treatment of 32 infants with intractable seizures, particularly for infantile spasms/myoclonic seizures. (Nordli DR Jr et al. **Pediatrics** 2001;108(1):129-133). Fifty five percent of infants had >50% reduction in seizure frequency (19% were seizure-free). Improvements in behavior, attention/alertness, activity level and socialization were also reported. The diet was well tolerated, and 96% maintained appropriate growth. Adverse events were all reversible.

## EFFECT OF PRICE ON USE OF ACTH FOR INFANTILE SPASMS

The effect of the 14-fold price increase on Aug 27, 2007 of ACTH treatment of infantile spasms in the US was evaluated at University of Colorado, Denver, in 97 patients treated 2007-2009. Before the price increase, patients were more likely to be treated with ACTH as first choice, and were hospitalized for 2.2+/-0.5 SD days for initiation. After price increase, oral AEDs were a more likely first treatment ($P<0.002$), and those selected for ACTH were hospitalized significantly longer (5.1+/-0.6 days, SD $P<0.001$). ACTH is considered the most effective treatment for infantile spasms and before 2007, 88% of child neurologists in the US used ACTH as initial therapy. In late 2009, vigabatrin became the only approved drug for infantile spasms in the US. UK studies in 2004 and 2005 found that hormonal treatment more often stopped spasms and improved outcomes than vigabatrin. Since the studies, treatment practices in the UK changed in 2008 in favor of steroids. Whereas the natural form of ACTH is used in the US, accounting in part for the price increase, a synthetic form is used elsewhere. Pricing and availability are influencing the choice of treatment and hospitalization for infantile spasms. (Wray CD, Benke TA. Effect of price increase of adrenocorticotropic hormone on treatment practices of infantile spasms. **Pediatr Neurol** Sept 2010;43:163-166). (Respond: E-mail: carterwraymd@gmail.com).