cysts and hydrocephalus. An ocular cyst should be ruled out before treatment with albendazole that may exacerbate inflammation. (American Academy of Pediatrics **Red Book**, 27th ed, Elk Grove Village, IL, AAP, 2006, pp 646-7).

LYME NEUROBORRELIOSIS AND ALICE IN WONDERLAND SYNDROME

Pediatric neurologists at the Floating Hospital, Tufts University, Boston, MA report a 7-year-old boy with Alice in Wonderland syndrome associated with Lyme disease. He initially awakened with a nightmare, scared and screaming, saying that "his mind was running fast" and he heard "baseball fans cheering." He vomited the following day but had no headache. Three nights later he awoke, went down stairs, and was pale and scared. He had repetitive swallowing and lip-smacking and said "my head is running fast." Next evening while reading, the book appeared to be a distance away, and he said the letters were becoming smaller.

During 36 hours of video EEG recording he had 3 events of distorted perception, a feeling of becoming smaller and the book print farther away, none associated with evidence of seizure. A Lyme disease test at the 6th day, performed at the insistence of the mother, was positive. CSF revealed lymphocytes 22/mm3, protein 23/mg/dl, and glucose 63 mg/dl. Lyme disease Western blot immunoglobulin M on 10th day tested positive in serum and CSF. Lyme PCR on CSF was negative and MRI was normal. He was treated with IV ceftriaxone for 21 days, and symptoms resolved after 3 days. He had no rash and no recurrence at 12-month follow-up. No personal or family history of migraine or epilepsy was elicited. Alice in Wonderland syndrome, or metamorphopsia, should be added to the clinical spectrum of Lyme neuroborreliosis, and a high index of suspicion is required in diagnosis. (Binalsheikh IM, Griesemer D, Wang S, Alvarez-Altalef R. Lyme neuroborreliosis presenting as Alice in Wonderland syndrome. **Pediatr Neurol** March 2012;46;185-186). (Response: Dr Binalsheikh. E-mail: alsheikh55@yahoo.com).

COMMENT. "Curiouser and curiouser!" said Alice, when she grew and expanded like a telescope after eating the cake. Distortions of form, size, movement, color, or sense of time are characteristic of metamorphopsia that may occur as a sensory aura during migraine or frontal lobe epilepsy. A PubMed literature search lists reports of several infectious agents associated with Alice in Wonderland syndrome, including Epstein-Barr virus, varicella, coxsackievirus B1, H1N1 influenza virus, and infectious mononucleosis. Lyme disease in the Tufts' case presents with visual and other illusions without systemic manifestations of neuroborreliosis. Alice in Wonderland syndrome is a benign disorder of childhood caused most frequently by migraine or Epstein-Barr virus infection. (Losada-Del Pozo R et al. **Rev Neurol** 2011;53(11):641-648).

CEREBRAL MALFORMATIONS

HEMORRHAGE RISK OF CAVERNOUS MALFORMATION

Researchers at the Mayo Clinic, Rochester, MN reviewed the records and radiograph data of 292 patients (47.3% male) seen between 1989 and 1999 with a

diagnosis of intracerebral cavernous malformation (ICM). The mean age at diagnosis was 45.8 years (range 3.5-88.9 years). Seventy-four patients presented with hemorrhage, 108 with seizure or focal deficit (symptoms not related to hemorrhage), and 110 were asymptomatic. The overall annual rate of hemorrhage at follow-up in the patients grouped according to presenting symptoms was 6.19%, 2.18%, and 0.33%, respectively. The median length of follow-up was 7.3 years (range 0-25 years); 68 patients underwent surgical excision. Patients who presented initially with symptomatic hemorrhage were at higher risk for future hemorrhage ($p<0.001$), and hemorrhage risk decreased with time; 81% were free of prospective hemorrhage at 10.6 years. Of 32 patients with prospective symptomatic hemorrhage, 19 (59%) initially presented with hemorrhage. The annual statistically significant risk factors for prospective hemorrhage included younger age ($p=0.02$) (without adjustment for gender etc.), male gender ($p=0.02$), infratentorial location ($p=0.015$), initial presentation as hemorrhage ($p<0.001$), and multiplicity of ICMs ($p=0.01$). Pregnancy was not a risk factor. The median time from first to second hemorrhage was 8 months. (Flemming KD, Link MJ, Christianson TJH, Brown RD Jr. Prospective hemorrhage risk of intracerebral cavernous malformations. **Neurology** Feb 2012;78:632-636). (Response: Dr Flemming. E-mail: flemming.kelly@mayo.edu).

COMMENT. In an editorial, Salman and Murray question whether a risk of re-bleeding of 10-20% in the first 2 years of follow-up of cavernous malformations warrants early neurosurgical excision. (Salman RA, Murray GD. **Neurology** 2012;78:614-615). This question is addressed in the following studies in Switzerland.

In a multicenter study of 79 pediatric patients with cerebral cavernous malformation (CCM) treated by surgical resection at University Hospital, Zurich, 77.3% became seizure free. Resection was the treatment of choice if lesions caused medically refractory epilepsy or other persistent symptoms. (Hugelshofer M, et al. **J Neurosurg Pediatr** 2011;8(5):522-525). Mean age at presentation was 9.7 years, and mean age at operation was 11.3 years. One-quarter of all CCMs affect children.

In a study of outcome of 20 children with CCM treated in Berne, Switzerland, average age at presentation was 8.5 years (range 7 months to 16 years). Presentation was acute hemorrhage in 17 (85%), seizures in 9 (45%), focal neurologic symptoms in 5 (25%), and headache only in 3 (15%). Location was supratentorial in 15 (75%). Treatment was conservative in 10 and surgical in 10. Neurological sequelae at follow-up (0.5-10 years) occurred in 6 (30%) patients. (Bigi S et al. **Eur J Pediatr Neurol** 2011;15(2):109-116).

A study of the natural history of CCM in 92 children and young adults at the University of Michigan, Ann Arbor, found the imaging prevalence of CCM increased with advancing age ($p=0.002$). Multiple CMs occurred in 28 (30%) patients. Thirty patients presented with hemorrhage, and the hemorrhage rate was 8% per patient-year in the symptomatic group. Symptomatic hemorrhage after long-term follow-up was associated with initial acute hemorrhage ($p=0.02$). (Al-Holou WN et al. **J Neurosurg Pediatr** 2012;9(2):198-205).

A prospective, Scotland, population-based cohort study of 139 adults with CCM, radiologically validated, found the risk of recurrent intracranial hemorrhage or focal neurological deficit from a CCM is greater than the risk of a first event, greater for women than for men ($p=0.01$), and declines over 5 years from 9.8% in year 1 to 5.0% in

year 5. (Salman R A-S et al. **Lancet Neurol** March 2012;11:217-224). The increased risk in women in this study of adults only is different from that in the Mayo Clinic study showing a preponderant risk in males.

SECKEL SYNDROME WITH HOLOPROSENCEPHALY

A case of Seckel syndrome (SS) accompanied by semilobar holoprosencephaly and arthrogryposis is reported from Erciyes University, Kayseri, Turkey. Seckel syndrome is a rare autosomal recessive disorder characterized by prenatal and postnatal growth retardation, microcephaly, and "bird-like" face with prominent, beak-like nose and micrognathia. A 1-day-old female newborn was admitted with dysmorphic features and feeding difficulties. The parents were consanguineous. In addition to typical features of SS, the baby had arthrogryposis, and cranial MRI showed semilobar holoprosencephaly, lissencephaly/pachygyria, dilated occipital horn of the lateral ventricle, hypoplasia of the frontal horn, non-cleavage of the basal ganglia and frontal lobe, and dysgenesis of the corpus callosum. Neuronal migration disorders should be investigated in infants born with facial characteristics of SS. (Sarici DS, Akin MA, Kara A, Duganay S, Kurtoglu S. Seckel syndrome accompanied by semilobar holoprosencephaly. **Pediatr Neurol** March 2012;46:189-191). (Respond: Dr Sarici. E-mail: drdilekcoban@yahoo.com.tr).

COMMENT. Holoprosencephaly (HP) is presented as a new associated feature of SS. Based on grades of severity, this case is a semilobar form of HP. The mechanism of the facial dysmorphism in SS may be attributable to defective mesencephalic neural crest tissue formation (Sarnat HB, Flores-Sarnat L. **J Child Neurol** 2001;16;918-931).

LEARNING DISABILITIES

TRACTOGRAPHY NEUROANATOMICAL STUDY OF DYSLEXIA

Researchers at Catholic University of Leuven, Belgium used diffusion tensor imaging tractography, a structural MRI technique, to assess the integrity of white matter tracts involved in reading. Group comparisons of 20 adults with dyslexia and 20 typical reading adults showed a significantly reduced fractional anisotropy (i.e. an index of the amount of anisotropy) in the left arcuate fasciculus of dyslexics, reflecting reduced myelination. Performance on phoneme awareness and speech perception was specifically related to the integrity of the left arcuate fasciculus (dorsal phonological route underlying grapheme-phoneme decoding), whereas orthographic (direct word) processing was related to fractional anisotropy values in the left inferior fronto-occipital fasciculus (ventral orthographic route). Structural anomalies found in the left arcuate fasciculus of dyslexics corroborate the hypothesis of dyslexia as a disorder of network connections. (Vandermosten M et al. **Brain** 2012;135:935-948).(Response: M Vandermosten. E-mail: maaike.vandermosten@ppw.kuleuven.be).

COMMENT. Dyslexics have reduced white matter integrity in the component of the left arcuate fasciculus that links Wernicke's to Broca's area.