

headache. A higher body mass index in children with nonspecific headache predisposes to sleep-disordered breathing and leads to sleep disorders. Polysomnography may help to clarify the association of headache and sleep disturbances.

## **CEREBRAL BLOOD FLOW AND MIGRAINE**

Blood flow in the basilar and internal carotid arteries and diameters of middle meningeal, carotid, and cerebral arteries were measured using 3Tesla magnetic resonance angiography, at baseline, during infusion of nitroglycerin or placebo, and during a provoked attack, at 6 hrs after infusion, in 32 migraineurs, aged 18-55 years, in a study at Leiden University Medical Centre, the Netherlands. Migraine headache was provoked in 20/27 (74%) migraineurs who received nitroglycerin, but in none of 5 patients who received placebo. Nitroglycerin caused a transient vasodilatation of all blood vessels. Blood vessel diameters were no different during a provoked migraine attack compared to baseline, nor between headache and non-headache sides. Blood flow in the basilar and internal carotid arteries was unchanged during nitroglycerin infusion and migraine headache. (Schoonman GG, van der Grond J, Kortmann C, van der Geest RJ, Terwindt GM, Ferrari MD. Migraine headache is not associated with cerebral or meningeal vasodilatation – a 3T magnetic resonance angiography study. *Brain* Aug 2008;131:2192-2200). (Respond: GG Schoonman MD, Department of Neurology (KS-Q), Leiden University Medical Centre, PO Box 9600, 2300 RC Leiden, The Netherlands. E-mail: [g.g.schoonman@lumc.nl](mailto:g.g.schoonman@lumc.nl)).

COMMENT. Contrary to current theory of migraine mechanisms, vasodilatation of cerebral or meningeal blood vessels is not of primary importance in the pathophysiology of the migraine headache.

## **NEUROCUTANEOUS SYNDROMES**

### **SYMPTOMS AND COURSE OF XERODERMA PIGMENTOSUM**

Sixteen Finnish patients with xeroderma pigmentosum (XP) were followed for up to 23 years, and their neurological symptoms and course determined in a study at Turku University Central Hospital, Finland; Erasmus University, Rotterdam, The Netherlands; University of Brighton, and University of Sussex, UK. Severe sunburn with minimal sun exposure in early infancy was the first sign of the disease in all cases, only 2 cases being diagnosed at that time. XP patients are assigned in 8 complementation groups, XP-A and XP-C being the most common groups in Europe. Neurological symptoms occur most often in XP-A patients. In XP-C patients, skin problems are severe, but neurological symptoms are rare.

Seven of the 16 Finnish patients were classified as XP-A. All had short stature and microcephaly. They developed normally until age 2 years, but neurological and cognitive dysfunction was apparent in childhood, before the age of 8 years. Cerebellar ataxia was recognized before age 4-16 years, followed by sensory motor neuropathy with areflexia, and sensorineural deafness. Cognitive problems were associated with an unusual tendency to weep and to be frightened. In early adulthood, 8 of 11 patients had developed choreoathetoid

involuntary movements. Corticospinal involvement appeared in the third decade, progressing to spastic tetraplegia and dysphagia. Two patients in the XP-C group had normal neurological findings, but they developed severe skin and ocular malignancies in pre-school years. The one XP-G patient had sensorineural hearing loss, laryngeal dystonia and peripheral neuropathy. Neurological disease was associated with failure of fibroblasts to recover RNA synthesis following UV irradiation. Dermatological symptoms included freckling, poikiloderma with hyper- and hypo-pigmentation and skin atrophy, in areas exposed to sun. Eye signs included nodular tumors in the eyelids, conjunctivitis, and keratopathy. Seven patients with severe neurological signs died at a median age of 33 years (range, 29-40 years). Cause of death was pneumonia. (Anttinen A, Koulu L, Nikoskelainen E, et al. Neurological symptoms and natural course of xeroderma pigmentosum. *Brain* Aug 2008;131:1979-1989). (Respond: Anu Anttinen MD, Department of Neurology, Turku University Central Hospital, PB 52, 20521 Turku, Finland. E-mail: [anu.anttinen@tyks.fi](mailto:anu.anttinen@tyks.fi)).

COMMENT. Xeroderma pigmentosum is a rare autosomal recessive disease that presents in early childhood with unusual skin sensitivity to sun exposure. Dermatological manifestations are complicated in later childhood by progressive neurological, cognitive and ocular manifestations. Patients are classified by complementation analysis in 8 groups, some (group A) being particularly susceptible to neurological symptoms. Early diagnosis and protection from exposure to sunlight result in improved prognosis with minimal skin problems and slower neurological deterioration. UV penetrates only the skin, and the nature of the DNA lesion and mechanism of neurological degeneration are not precisely understood. Dr AMR Taylor of the University of Birmingham, UK, comments that the neurodegeneration is most likely related to some form of oxidative damage (*Brain* Aug 2008;131:1967-1968).

## SEIZURE DISORDERS

### EVALUATION OF CHILDREN FOR EPILEPSY SURGERY

To assess the possibility of streamlining the decision process for epilepsy surgery in children with intractable epilepsy, the value of MRI, video EEG, and SPECT was investigated retrospectively in a study of 353 patients at the Lingfield Epilepsy Centre, Great Ormond Street Hospital, and Institute of Child Health, London, UK). Of 238 children offered resective surgery, 215 (92%) had a unilateral localized lesion on MRI, 20 (8%) had bilateral imaging abnormalities, and 3 had normal imaging. In the group with unilateral localized structural abnormalities, EEG telemetry did not affect a decision to operate. In children with bilateral MRI abnormalities or normal scan, the probability of resective surgery was 78% in those with EEG-localized ictal onset compared to 9% with nonlocalized EEG ( $p<0.001$ ). SPECT did not affect a decision to operate in any group. Children with medically intractable epilepsy and localized lesions on MRI may not need ictal EEG recordings or SPECT in the evaluation for epilepsy surgery. The value of EEG telemetry in selected cases requires further investigation. (Patil SG, Cross H, Chong WK, et al. Is streamlined evaluation of children for epilepsy surgery possible? *Epilepsia* Aug 2008;49:1340-1347). (Respond: Dr Rod C Scott, Institute of Child Health, The Wolfson Centre, Mecklenburgh Square, London WC1N 2AP, UK. E-mail: [r.scott@ich.ucl.ac.uk](mailto:r.scott@ich.ucl.ac.uk)).