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).

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).

COMMENT. The clinical and MRI findings are usually sufficient evidence on which to base a decision to offer resective surgery in the majority of children with medically intractable epilepsy. Ictal EEG video recordings provide confirmatory evidence of focal lesions, but in situations with limited resources, they may be reserved for children with bilateral MRI changes or normal MRI. SPECT findings do not influence a decision to operate. The role of ictal EEG in estimating prognosis following surgery requires further study. Psychological assessment is an additional factor in the decision making process for epilepsy surgery and in the evaluation of its benefits.

Extratemporal ictal clinical features in hippocampal sclerosis are most frequent in cases with severe hippocampal atrophy but do not affect surgical outcome, in a study at the University of Verona, Italy; and National Hospital, London, UK. (Borelli P, Shorvon SD, Stevens JM, et al. **Epilepsia** Aug 2008;49:1333-1339).

SPIKE FREQUENCY AND SURGICAL OUTCOME IN TLE WITH UNILATERAL HIPPOCAMPAL ATROPHY

Prognostic implications of absolute spike frequency over the affected temporal lobe and relative spike distribution between the two temporal lobes for postoperative seizure control were assessed in 55 adult patients with medically refractory mesial temporal lobe epilepsy (MTLE), in a study at General Hospital Hietzing, Vienna, Austria. Age at seizure onset was 11.2 +/- 9.3 years, and age at evaluation was 34.7 +/- 8.1 years. Presurgical evaluation included prolonged video-EEG monitoring. One year following surgery, 4 of 14 patients (28.6%) in the frequent spike group (>60/hr over the affected temporal lobe) were completely seizure free compared to 33 of 41 patients (80.5%) in the nonfrequent spikes group (<60/hr) (p=0.001). In the unitemporal group (>90% spikes over the affected temporal lobe), 25 of 35 patients (71%) were completely seizure free compared to 12 of 20 patients (60%) in the bitemporal spike group (p=0.282). Relative spike distribution over the two temporal lobes was not significantly related to postoperative outcome. (Krendl R, Lurger S, Baumgartner C. Absolute spike frequency predicts surgical outcome in TLE with unilateral hippocampal atrophy. **Neurology** Aug 2008;71:413-418). (Respond: Dr C Baumgartner. E-mail: christoph.baumgartner@wienkav.at).

COMMENT. Absolute spike frequency over the affected temporal lobe, but not relative spike distribution between the two temporal lobes, is a strong predictor of postoperative seizure control in young adults with medically refractory mesial temporal lobe epilepsy. TLE patients with a low spike frequency over a unilateral hippocampal atrophy should be excellent candidates for epilepsy surgery.

NOVEL LOCUS FOR GENERALIZED EPILEPSY WITH FEBRILE SEIZURES PLUS

Linkage analysis was conducted in 5 French families with generalized epilepsy with febrile seizures plus (GEFS+), with at least 7 affected members with autosomal dominant transmission, in an attempt to localize a new gene for GEFS+, at Hospitalier Pitie-Salpetriere,