reported in two unrelated boys with consanguineous parents from the Centre de Génétique Médicale, Service de Pediatrie Générale and Radiologie, Hôpital d'Enfants de la Timone, Marseille, France. An autosomal recessive mode of inheritance is suggested and echographic survey of further pregnancies is advised. Clinical manifestations included macrocephaly, bulging forehead, antimongoloid slant of eyes, broad short nose, posteriorly rotated ears, herniae, polydactyly, cardiac defect, mental retardation and corpus callosal agenesis. (Philip N et al. The acrocallosal syndrome. Eur J Pediatr Feb 1988;147:206-208).

COMMENT. Only 12 cases of the syndrome have been reported. Schinzel, who described the syndrome in 1979 and has at least 5 publications on the subject, deserves the eponym.

FRAGILE-X SYNDROME

A characteristic epileptogenic EEG pattern is described in five of 12 male subjects with fragile-X syndrome evaluated at the Instituto Oasi, via C. Ruggero, Troina, Italy, and Clinica Neurologica, II Universita Roma and Bologna, Italy. Focal paroxysmal temporal spikes, at times multifocal, occurred in sleep in one non-epileptic and four epileptic patients with mental retardation and fragile-X syndrome, but not in subjects with mental retardation, with or without epilepsy but without the fragile-X chromosome. (Musumeci SA et al. Fragile-X syndrome: A particular epileptogenic EEG pattern. Epilepsia Jan/Feb 1988;29:41-7).

COMMENT. The authors believe that epilepsy must be considered an important clinical feature of fragile-X syndrome, occurring in an average of 26% of reported cases. Karyotyping is advised in mentally retarded patients with epilepsy, even in those without typical clinical features or positive family history and especially in children who frequently lack the characteristic facial dysmorphisms and macro-orchidism (see Ped Neur Briefs 1987;1:41).

INTRACRANIAL TUMORS

NEUROFIBROMATOSIS AND ACOUSTIC NEUROMAS

The criteria for diagnosis, treatment, family counseling and advances in genetics of neurofibromatosis are reviewed by a neurosurgeon and epidemiologist at the Massachusetts General Hospital, Boston, and the National Institute of Neurological Disorders, Bethesda, MD.

The neurofibromatoses consist of two distinct disorders, a peripheral and a central type, with genes located on separate chromosomes. The diagnosis of neurofibromatosis 1 (NF1, von Recklinghausen's neurofibromatosis or VRNF in Europe) requires two or more of the following: 6 or more cafe au lait macules, 2 or more neurofibromas, axillary or inguinal skin freckles, optic glioma, Lisch iris nodules, osseous lesion and familial occurrence. Neurofibromatosis 2 (NF2, bilateral acoustic neurofibromatosis or BANF in Europe) requires one of the following for diagnosis: a) bilateral eighth nerve tumors, or b) a positive family history plus a unilateral eighth nerve tumor or two of the following: neurofibroma, meningioma, glioma, Schwannoma, or lenticular opacity. For patients with NF2 there is a 50% risk of transmission to any offspring, and close relatives should be screened for cafe au lait spots or neurofibromas, acoustic nerve tumors and lens opacities. Acoustic neuromas commonly become symptomatic during or soon after puberty and