diet restricted in isoleucine, valine, methionine, and threonine resulted in improvement and normal growth and development up to 2 yrs of age when she had an acute "metabolic stroke" precipitated by otitis media. Subsequently, she developed dystonia, dysarthria, dysphagia, and spastic quadriplegia. A CT scan showed bilateral symmetric lucencies of the globus pallidus and internal capsules. The lesions were thought to result from deranged organic acid metabolism and accumulation of toxic metabolites in the brain. (Heidenreich R et al. Acute extrapyramidal syndrome in methylmalonic acidemia: "Metabolic stroke" involving the globus pallidus. J Pediatr Dec 1988;113: 1022-7).

COMMENT. Lesions of the basal ganglia have been noted in other inborn errors of organic acid metabolism, including propionic acidemia, and glutaric aciduria type 1, in Leigh syndrome and the mitochondrial encephalomyopathies including Kearns-Sayre and MELAS syndromes, and in several other hereditary diseases involving the CNS.

PEROXISOMAL DISORDERS

The biochemical and clinical diagnostic characteristics of peroxisomal disorders are reviewed by authors from the Department of Pediatrics, Medical University of South Carolina, Charleston, SC. Two major groups are recognized: 1) those that represent a diffuse peroxisomal dysfunction due to (a) a reduction in number of peroxisomes (Zellweger's syndrome, neonatal adrenoleukodystrophy, infantile Refsum's disease, and hyperpipecolic acidemia); and (b) normal numbers but reduced activities of multiple enzymes (rhizomelic chondrodysplasia punctata); and 2) those disorders with specific single peroxisomal enzymatic defects (childhood adrenoleukodystrophy-X-linked, adult Refsum's disease, hyperoxaluria, acatalasemia, and pseudo-Zellweger's syndrome).

Peroxisomal disorders should be considered in any infant with hypotonia and delays in psychomotor development, and especially in those with facial dysmorphisms (high and prominent forehead), hepatomegaly, cataracts, retinitis, calcific stippling, short limbs, failure to thrive, seizures, and hearing deficit. In childhood, loss of motor skills, progressive dementia, and skin bronzing should suggest the diagnosis. Measurement of very long chain fatty acids is used to confirm the biochemical defect, and other tests including bile acid, phytanic acid, and plasmalogen are included for specific diagnoses. (Singh I et al. Peroxisomal disorders. Biochemical and clinical diagnostic considerations. AJDC Dec 1988;142:1297-1301).

COMMENT. Peroxisomal disorders are rare but important because clinical sequelae can be related to specific biochemical deficits and some may be identified prenatally and their recurrence prevented. Dietary restriction of very long chain fatty acids and plasmapheresis are helpful

in treatment by controlling the accumulation of toxic metabolites in some peroxisomal disorders, and the addition of glycerol trioleate, a lipid containing unsaturated fatty acid, is a promising new therapy that reduces the synthesis of C22-26 fatty acids. For further information on peroxisomal disorders, refer to a special article by Moser HW Neurology Oct 1988;38:1617. A family with Refsum's disease (heredopathia atactica polyneuritiformis) in whom 4 out of 6 siblings were affected is reported from the Department of Neurology, Westminster Hospital, London (Britton TC, Gibberd FB. JR Soc Med Oct 1988;81:602-3). Retinitis pigmentosa was the presenting diagnostic sign in the index case, and other affected members of the family were detected by screening for raised plasma phytanic acid levels. Early diagnosis is important because dietary treatment will prevent the development of neuropathy, ataxia, cardiac arrhythmias, and ichthyosis. Retinitis pigmentosa, anosmia, and ataxia should suggest the diagnosis.

CNS TUMORS

POSTERIOR FOSSA DERMOIDS

Three children with dermoid cysts of the posterior fossa are reported from the Assaf Harofeh Medical Center, Zerifen, and Hadassa Medical Center, Jerusalem, Israel. Two presented with acute meningitis at 1 and 2 yrs of age, and the third patient had hydrocephalus treated by ventriculo-peritoneal shunt at 7 mos and complicated by meningitis and cerebellar abscess at 9 mos of age. CT scans with enhancement and bone window setting revealed the midline bony defect and low density lesion with ring enhancement in the posterior fossa. (Starinsky R et al. Dermoids of the posterior fossa. Case reports and review. Clin Pediat Dec 1988;27:579-582).

COMMENT. Recurrent meningitis or brain abscess in an infant or young child should prompt a search for a sinus, fistula, and bone defect in the occipital area. Dermoid cysts of the cerebellum and posterior fossa account for 2% of intracranial tumors in children. Astrocytoma, medulloblastoma, brain stem glioma, and ependymoma of the IVth ventricle are the most frequently encountered IC tumors.

LIPOMA OF CORPUS CALLOSUM

A lipoma of the corpus callosum diagnosed by CT at 7 mos and mistaken for hemorrhage in a premature infant is reported from the Medical College of Pennsylvania, Philadelphia, PA. The Apgar scores were 3 at 1 and 5 min, and the infant had hyaline membrane disease that progressed to bronchopulmonary dysplasia. Cranial ultrasound at 10 hrs demonstrated a subependymal hemorrhage with unchanged appearance at 17 days. Seizures