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

associated with bilateral temporal polyspike and sharp-wave EEG activity were controlled with phenobarbital. His head circumference was at the 75th percentile and disproportionately large. A repeat ultrasound at 7 mos showed increased echogenicity in the midline with normal ventricles, interpreted as blood in the third ventricle with calcification. CT showed a large midline area of decreased density extending into the lateral ventricles, consistent with a lipoma of the corpus callosum and calcifications. At 18 mos the head is large and development is delayed at the 12 mo level. (Imaizumi SO et al. Lesion mistaken for hemorrhage in a premature infant: Lipoma of corpus callosum. Pediatr Neurol Oct 1988;4:313-6).

COMMENT. Blood, fat, and calcium have similar echogenicity by cranial ultrasound, and all three media are hyperechoic when compared to the moderate echogenicity of cerebral white matter, low echogenicity of gray matter, and absent echogenicity of fluid-filled ventricles or cysts. Lipomas are characterized by hyperechoic densities on cranial sonograms.

SPINAL CORD HEMANGIOBLASTOMA

A 6-month-old infant with a spinal cord hemangioblastoma located in the conus medullaris is reported from the University of Washington School of Medicine, Children's Hospital, Seattle, WA. At birth the physical examination was normal except for a lumbosacral dimple. By 1 mo the dimple had deepened and a hemangioma developed at the site. CT and CT metrizamide myelography revealed a posterior filling defect at the level of the conus medullaris. At laminectomy, a congenital dermal sinus tract, cutaneous capillary hemangioma and cord hemangioblastoma were resected. There were no neurologic deficits either postoperatively or following removal of the tumor. (Michaud LJ et al. Hemangioblastoma of the conus medullaris associated with cutaneous hemangioma. Pediatr Neurol Oct 1988;4:309-12).

COMMENT. Spinal cord hemangioblastoma rarely presents in infancy and is usually manifest from the third to the fifth decades. It may be associated with a variety of cutaneous and other lesions, including von Hippel-Lindau and Cobb syndromes. Von Hippel-Lindau disease is an autosomal dominant condition characterized by hemangioblastomas of the cerebellum, medulla and spinal cord; angiomas of the retina, liver, and kidney; pheochromocytomas, adenomas, or cysts of the kidney and epididymis; and pancreatic cysts. In Cobb syndrome, cutaneomeningospinal angiomatosis, spinal cord arteriovenous malformations are associated with cutaneous vascular lesions in corresponding dermatomes. These syndromes were not found in the above case-report.

CHEMOTHERAPY FOR MEDULLOBLASTOMA

The efficacy of adjuvant chemotherapy for patients with