of purine metabolism characterized by hypotonia followed by spasticity, chorea, athetosis, dystonia, growth and mental retardation, self-mutilatory behavior, hyperuricemia, and nephrolithiasis (Lesch M, Nyhan WL. AM J Med 1964;36:561). The phenotypical expression is the result of a deficiency of hypoxanthineguanine phosphoribosyl-transferase enzyme, the gene located on the long arm of the X chromosome. Response to medications is variable but self-mutilation can sometimes be ameliorated (Herman BH et al. Naltrexone decreases self-injurious behavior. Ann Neurol 1987;22:550).

INTRACRANIAL TUMORS

NEONATAL CRANIOPHARYNGIOMA

A craniopharyngioma detected in utero using ultrasound is reported from the Depts of Neurosurgery, Neonatology, Pathology, and Obstetrics and Gynecology, New York University Medical Center, New York, NY. The obstetric ultrasound study performed because of premature labor at 35 weeks showed polyhydramnios and macrocephaly secondary to an intracranial, calcified lobulated mass. A CT scan performed at 2 hrs. of life demonstrated a calcified intracranial mass extending to the base of the skull. The infant suffered cardiorespiratory arrest and died at the second day of life. At postmortem examination, the suprasellar mass was a granular tumor with multiple cysts filled with green mucoid material. Microscopically, the tumor showed branching cords and pallisading of epithelial cells, focal calcifications, and an inner zone of stellate cells, an appearance diagnostic of craniopharyngioma. The authors uncovered only four other reports of congenital intracranial neoplasms diagnosed prenatally using ultrasound. Three were teratomas and one a dysplastic mass. Calcifications have been observed in teratomas and meningiomas in utero and are not pathognomonic of craniopharyngioma. Low set ears and polydactyly are reported as associated congenital anomalies. Polyhydramnios occurred in all five cases of neonatal brain tumors diagnosed antenatally by ultrasound. Radical excision of the tumor in the neonatal period was not advised. (Freeman TB et al. Neonatal cranipharyngioma. NY State J Med Feb 1988;88:81-83).

COMMENT: Craniopharhngiomas comprise about 3% of all intracranial tumors at all ages and 9% in children. Only 10 cases of neonatal craniopharyngioma were culled from the literature. Radical excision for cases presenting in infancy is recommended by the following authors reporting their experiences with 50 cases in Paris.

TREATMENT OF CRANIOPHARYNGIOMA IN INFANCY

A retrospective analysis of the outcome of 50 cases of craniopharyngioma treated in infancy by radical or subtotal surgical excision and irradiation is reported from the Services of Neurosurgery and Endocrinology, Hopital des Enfants Malades, 149, reu de Sevres, 75743 Paris Cedex 15, France. The authors concluded that 1) radical excision is the treatment of choice; 2) if radical excision is not possible, surgery should be followed by irradiation to lower risk of recurrence; and 3) radiotherapy should be delayed as long as possible because of hazards to

growing brain and used only when tumor recurrence has been demonstrated. After radical excision, the rate of recurrence was lowest, with a 10 year recurrence-free survival rate of 88%. After subtotal removal, the recurrence-free survival rate, 10 yrs post-op, was 37%; this rate was significantly higher (72%) when subtotal removal was followed by irradiation, but deafness and severe neuropsychological and intellectual sequelae were frequent complications of irradiation. Post-operative mortality was low in pre-chiasmatic cases and high in retro-chiasmatic tumors. Surgical statistics may improve with newer techniques. (Pierre-Kahn A et al. Traitement des craniopharyngiomes de l'enfant. Analyse retrospective de 50 observations. Arch Fr Pediatr Mars 1988;45:163-167).

COMMENT: The neuropsychological deficits ascribed to irradiation in this report may be explained in part by the location of the tumor. Cognitive defects have been correlated with frontal lobe abnormalities seen on MRI in 4 patients with craniopharyngioma (Stelling MW et al. Am J Dis Child 1986;140:710).

INFECTIOUS DISEASE

TREATMENT OF AIDS ENCEPHALOPATHY

A 3-year old boy who had acquired HIV infection transplacentally and developed AIDS encephalopathy is reported from the Depts of Paediatrics and Immunology, Newcastle General Hospital, Newcastle upon Tyne, England. During hemophilus influenza pneumonia at 26 months his speech regressed to expressive aphasia and he developed spastic diplegia with inability to walk. CT scan showed cerebral atrophy. CSF showed no cells and normal glucose and protein; IgG antibodies to HIV were increased. Treatment with intravenous gammaglobulin 300 mg/kg and oral zidovudine (Retrovir-Wellcome) 100 mg/m^2 4x daily every 4 weeks for 8 months led to considerable clinical improvement and an almost normal CT. Spasticity regressed allowing him to run unaided and his speech in single words became articulate. (Matthews J et al. AIDS encephalopathy with response to treatment. Arch Dis Child May 1988;63:545-547).

COMMENT: AIDS encephalopathy may be acute and rapidly progressive (15%), subacute but progressive (18%), and static with cognitive deficits (28%). A plateau course is apparent in many. The reported case was subacute in onset and without treatment further progression might have been expected. (See Ped Neur Briefs 1988;2:1).

PSYCHOGENIC DISORDERS

HYSTERICAL GAIT

In a study of the clinical features of conversion disorder in 52 children admitted to the Royal Alexandra Hospital for Children, Camperdown, New South Wales, Australia, hysterical gait disturbance was the main complaint in 71%, and pain, paresthesia or anesthesia in 77%. So called classical conversion symptoms such as blindness and globus were relatively rare. The disorder was rare below 8 years of age and girls outnumbered boys three to one. Spring and summer (Sept-Nov and Jan-Mar in Australia) accounted for 75% of admissions, coinciding with the end of year exams and the beginning of the new school year. Only 6 children had