hemorrhage. A mild tentorial laceration was considered likely in the cases reported. An inherited protein C deficiency may be manifested by massive venous thromboses in the newborn (Seligsohn U et al. N Engl J Med 1984;310:559), an etiology to be considered in the absence of a history of brain trauma.

TRANSPOSITION OF GREAT VESSELS AND MOBIUS SYNDROME

The vascular theory of embryopathogenesis for Mobius syndrome is proposed in a case report of a 3-month-old boy from the University of New Mexico School of Medicine, Albuquerque, NM. At birth, he had bilateral facial and abducens palsies, left acheiria (congenital absence of the hand), and transposition of the aorta and pulmonary artery. He was lethargic, cyanotic, and in respiratory distress, and he expired after an arterial switch procedure with closure of a septal defect. The authors cite 2 reports of Mobius syndrome with cardiac anomalies, both presenting with dextrocardia and the Poland anomaly (unilateral hypoplasia or absence of pectoral muscles, nipple, and upper limb). An intrapartum insult during the fourth to seventh week of gestation is consistent with the vascular theory of embryopathogenesis. (Raroque HG, Hershewe GL, Snyder RD. Mobius syndrome and transposition of the great vessels. Neurology Dec 1988;38:1894-5).

COMMENT. Congenital facial diplegia and abducens palsy, Mobius syndrome, has been explained as either a primary hypoplasia of cranial nerve nuclei or a primary deficiency of the muscles derived from the first two branchial arches. A dysgenesis of both neural and muscular tissue has been proposed in some cases. In the above case report, the concomitant occurrence of the vascular anomaly supports the theory of impaired cranial nerve nuclear development due to interruption of vascular supply at or around the sixth intrauterine week.

DEVELOPMENTAL DISORDERS AND LEARNING

VON RECKLINGHAUSEN NEUROFIBROMATOSIS

A population-based study in southeast Wales and reported from the Institute of Medical Genetics and Section of Neurology, University of Wales College of Medicine, Cardiff, has identified 135 patients with neurofibromatosis type 1 (NF-1), a prevalence of approximately 1/5000. The major clinical features were multiple cafe-au-lait spots, dermal neurofibromas, Lisch nodules in the iris (93%), freckling in the axilla (67%) or groin (44%), macrocephaly (45%), and short stature (34%). Complications included plexiform neurofibromas in 40 (30%) patients, mental retardation in 13 (10%) severe in only 1, epilepsy 6 (4%), severe scoliosis 6, visceral and endocrine tumors 6, optic glioma 2, spinal neurofibroma 2, aqueductal stenosis 2, delayed puberty 2, and congenital glaucoma 1. No cases of acoustic neuroma were seen. The frequency of CNS and malignant tumors was 5%. The authors recommend regular biannual examinations during childhood, with particular attention to intellectual