

according to this theory. (McLone DG, Knepper PA. The cause of Chiari II malformation: A unified theory. Pediatr Neurosci 1989; 15:1-12).

COMMENT. Chiari described three types of cerebellar malformation which he had found in cases of congenital hydrocephalus. In type I the medulla was displaced downwards into the spinal canal and was covered by peg-like processes arising from the cerebellar hemispheres, their lower ends opposite the origin of the third cervical nerve roots. This anomaly was found in a girl of 17 in whom the malformation had caused no symptoms during life. Type II was a similar malformation of the lower parts of the cerebellum associated with an elongation of the fourth ventricle which extended into the spinal canal in a six month old baby with hydrocephalus. There were heterotopic nodules of gray matter in the walls of the lateral ventricles, the cerebellum was small, the pons was elongated, the medulla was entirely in the spinal canal, and the lower cranial nerves were elongated. Hydromyelia and meningocele were associated. Chiari type III was a single case of cervical spina bifida in which there was herniation of the cerebellum through the bony defect, a form of occipital meningoencephalocele. Arnold's description of a case of meningocele without hydrocephalus was less detailed than the earlier description by Chiari. Schwalbe and Gredig gave the name Arnold-Chiari malformation to type II which was always associated with meningocele. For a full description of the historical and pathological aspects of Chiari malformations see Greenfield's Neuropathology, Baltimore, Williams and Wilkins Co. The section on malformations of the nervous system was written by Dr. R. M. Norman.

CORTICOSPINAL TRACT IN NEWBORNS

The maturation and function of the corticospinal and corticobulbar tracts in the human newborn are reviewed from the Departments of Paediatrics, Pathology and Clinical Neurosciences, University of Calgary Faculty of Medicine, Calgary, Alberta, Canada. The myelination of these tracts begins in late gestation but is not complete until two years of age. Functions attributed to these descending pathways in the full term human newborn include the following: 1) Development of passive muscle tone and resting postures; 2) Enhancement of suck and swallow reflexes; 3) Relay of cortical epileptic discharges; and 4) Inhibition of complex stereotyped motor reflexes ("subtle seizures"). The antagonistic balance between flexors and extensors are controlled by the subcortical spinal and corticospinal pathways. If the corticospinal tract is impaired as with perinatal asphyxia the infant assumes distal flexion and proximal extension postures which reflect the influence of subcortical pathways when corticospinal tract antagonism is lacking. The "tonic seizures" of preterm infants with intraventricular hemorrhages are probably not true epileptic phenomena but rather episodes of decerebration. A weak suck, poor feeding, and impaired coordination

of swallowing are findings common to cerebral cortical disturbances in the newborn caused by hypoxia, meningitis, hemorrhage or hydrocephalus. Most human neonatal seizures are focal or multifocal and arise in the cerebral cortex and are relayed by the corticospinal tract to spinal motor neurons and to facial and hypoglossal nuclei by the corticobulbar tract. Some neonatal seizures are subcortical in origin. The corticospinal tract probably does not influence muscle maturation because muscle shows histochemical differentiation at 20-28 weeks gestation, long before myelination of the corticospinal tract begins. Cerebellar influence on muscle tone and coordination is mediated mainly by the corticospinal and corticobulbar tracts. Hypotonia is among the most constant clinical findings in infants with cerebellar hypoplasia. (Sarnat HB. Do the corticospinal and corticobulbar tracts mediate functions in the human newborn? Can J Neurol Sci 1989; 16:157-160).

COMMENT. Dr. Sarnat's research concerning the development of the corticospinal tract in the newborn aids the clinician in his understanding of reflexes, seizures, and muscle tone and posture. In another recent study, acridine orange, a fluorochromic stain of nucleic acids, was used to study neural maturation in human brains during development. The increase in cytoplasmic RNA of neurons coincided with the onset of neurotransmitter synthesis, and the presence of orange fluorescence in heterotopic nerve cells served as a marker of the state of maturity and degree of migration. (Sarnat HB. Rev Neurol (Paris), 1989; 145:127-133).

DEGENERATIVE DISORDERS

MOTOR DISORDERS IN RETT SYNDROME

The motor and behavioral findings in 32 patients with Rett syndrome aged 21 months to 30 years, are reported from the Departments of Neurology and Pediatrics, Baylor College of Medicine, Houston, TX. Hand stereotypies and gait abnormalities were present in all patients. Clapping, wringing, and clenching were the most common, followed by washing, patting, and rubbing movements. Gait ataxia was present in 31%, a broad based gait in 13%, and inability to walk in 28%. Bruxism was the next most common involuntary movement (97%) and occurred only when awake. Drooling occurred in 75%. Other motor disturbances included ocular deviations (63%), parkinsonian rigidity (44%), bradykinesia (41%), dystonia (59%), sometimes focal and sometimes associated with scoliosis (50%). Myoclonus, choreoathetosis and intention tremor also occurred. Hyperkinetic disorders were prominent in younger patients and bradykinetic disorders occurred more frequently in older patients. (FitzGerald PM et al. Extrapyramidal involvement in Rett's syndrome. Neurology Feb 1990; 40:293-295).

COMMENT. The number of cases and the plethora of published reports on Rett syndrome add credence to the viral infectious cause postulated by Hagberg (see Ped Neur Briefs 1989; 3:44). A toxic environmental cause might also be considered and pursued further.