and 3) spastic pseudobulbar form (12). Nerve and muscle biopsies showed neurogenic atrophy in the peroneus brevis muscle and minor changes in the superficial peroneal nerve. Infantile and juvenile ALS is usually rare and the frequency in Tunisia may be explained by the high incidence of consanguinity. (Ben Hamida M et al. Hereditary motor system diseases (chronic juvenile amyotrophic lateral sclerosis). Conditions combining a bilateral pyramidal syndrome with limb and bulbar amyotrophy. Brain April 1990; 113:347-363).

COMMENT. Ford FR, in his Diseases of the Nervous System in Infancy, Childhood, and Adolescence, refers to his own experience with hereditary amyotrophic lateral sclerosis as small, but unlike the cases described in the present study, Ford's cases showed a more rapid progression. The hereditary factor in juvenile ALS distinguishes this type from classical ALS or Charcot's disease, which develops in late middle age and which is usually neither hereditary nor familial. Both Ford and the present authors refer to articles by Holmes in 1905 and an autopsy report by Mass in 1911 on a child who had nystagmus in addition to the characteristic features, including degeneration of the pyramidal tracts and the motor cells of the anterior horns. The earliest report of an infantile amyotrophic lateral sclerosis of the familial type was by Brown CH (J Nerv and Ment Dis 1894; 21:707).

ACUTE INFANTILE SPINAL MUSCULAR ATROPHY

Massive muscle cell elimination by apoptosis in an infant who died eight weeks after birth from acute infantile spinal muscular atrophy is described from the Department of Neurology, Medical Academy, Warsaw, Poland, and the Division of Neuropathology, University of Mainz, Mainz, FRG. The classical morphological changes of ISMA included degeneration and loss of motor neurons in the spinal cord, loss of large myelinated fibers in anterior roots, and neurogenic atrophy in muscle. Ultrastructural findings in the muscle showed membrane bound muscle cell fragments or apoptotic bodies. Numerous immature muscle fibers were also observed suggesting a failure in muscle maturation. The authors speculate that in growth retarded muscle the process of muscle apoptosis may also be prolonged or repeated. The resulting protracted muscle cell death may lead to a greater reduction in the number and size of muscle fibers. The removal of the peripheral target of anterior horn cells then results in secondary death of motor neurons. (Fidzianska A et al. Acute infantile spinal muscular atrophy. Muscle apoptosis as a proposed pathogenetic mechanism. Brain April 1990; 113:433-445).

COMMENT. Death of muscle cells by apoptosis has not been demonstrated in infants with SMA previously. The term "apoptosis" was proposed by Kerr et al (1972) for cell death which plays a role in the regulation of animal cell populations. This form of cell death differs from that caused by coagulative necrosis. Apoptosis is responsible for the focal elimination of cells during embryonic development and metamorphosis. The final number of motor neurons in the spinal cord following fetal

development depends on input from muscle for survival. Naturally occurring death of motor neurons is accentuated by the removal of the target muscle. The findings in this case report suggest that motor neuron death in infantile spinal muscular atrophy may be secondary to muscle cell apoptosis.

LEARNING DISORDERS

NEUROLOGICAL TESTS IN PREDICTING LEARNING DISABILITIES

A battery of 12 simple neurological test items that differentiated normal from at-risk children at three and five years of age is described from the Departments of Pediatrics and Education, Wyler Children's Hospital, University of Chicago, IL. A follow-up of the five year olds at age seven showed a significant linear relation between scores on neurological tasks and the Wechsler Intelligence Test for Children. A poor neurological test score at age five correlated with a lower Full-scale IQ at age seven. The correlation for Verbal IQ was -0.42 and slightly lower than that for Performance IQ (-0.48). Both the school system's assessment and the neurological screening test accurately identified nearly all the children who needed special educational help at age seven. The neurological tests of predictive value for learning disabilities in preschool children included walking on toes and heels, tandem gait forward and backward, touch localization, restless movements, downward drift of outstretched hands, rapid alternating movements of forearms, hopping, alternate tapping of the fingers, and complex tapping. The percentages of at-risk children failing each neurological task were significantly lower than the normal group in almost all categories. (Huttenlocher PR et al. Discrimination of normal and at-risk preschool children on the basis of neurological tests. Dev Med Child Neur May 1990; 32:394-402).

COMMENT. The emphasis of attention deficits in the evaluation of children with learning disabilities has overshadowed the recognition of subtle or soft neurological signs in the evaluation of children who may need psychological testing and early remedial education. As a pediatric neurologist it is gratifying to review a report that demonstrates the importance of the neurological examination in children with potential learning problems.

Abnormal neurologic signs almost identical to those included in the above test battery have previously been correlated with hyperactive behavior and response to stimulant medication. In a study of 28 hyperactive children with learning disabilities and ADDH, those with the highest incidence of abnormal neurologic signs had the greatest degree of overactivity and were most likely to benefit from methylphenidate. (Millichap JG. Methylphenidate in hyperkinetic behavior: Relation of response to degree of activity and brain damage. In "Clinical Use of Stimulant Drugs in Children". Ed. Conners, CK. Amsterdam, Excerpta Medica 1974; 130-140).