height 65 cm, 50th percentile for age 4.5 mos), dysmorphic features, hypotonia, and developmental delay. Urine acylglycines revealed elevated ethylmalonic acid, suberylglycine, and dodecanedioic acid. Urine organic acids revealed moderate amounts of ethylmalonic and adipic acids, small amounts of methylsuccinic, and sebaric and sebacic acids. Muscle biopsy disclosed atrophic fibers throughout, myophagocytosis, occasional basophilic fibers, inflammatory infiltrates focally around blood vessels, and mild fiber type grouping. Trichrome stain revealed a focal increase in endomysial connective tissue with no ragged-red fibers. Electron microscopy demonstrated a normal myofibrillar architecture with normal number and structure of mitochondria. There was a slight increase in glycogen and no abnormal lipid deposition. Fatty acid oxidation studies in muscle showed that the activity of SCAD was about 30% of controls. EMG was myopathic. SCAD deficiency should be considered in the differential diagnosis of children with failure-to-thrive and nonspecific myopathy.

185. NEUROLOGIC MANIFESTATIONS AND MOLECULAR BASIS OF GROUP A XERODERMA PIGMENTOSUM

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The molecular basis of group A xeroderma pigmentosum (XP) was investigated by northern blot analysis of XPAC messenger RNA (mRNA). XPAC protein was also analyzed by Western blotting of immunoprecipitates. Northern blot analysis of poly(A) + RNA revealed that XPAC mRNAs of the typical group A XP cells were smaller than that of normal controls, and amounts were markedly reduced. Conversely, the XPAC mRNA of 2 atypical groups of A XP, whose neurologic manifestations were mild, were almost normal size and amount. Western blot analysis of XPAC protein revealed that the typical group A XP had no XPAC protein bands, while a smaller truncated XPAC protein was detected from 2 atypical group A XP cells. These results suggest that a correlation exists between neurologic manifestations and both quantity and quality of XPAC mRNA and XPAC protein.

186. BEST TREATMENT FOR CHILDHOOD EPILEPSY: RANDOMIZED TRIALS OF THE LAST 20 YEARS

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Seizure disorders are the most common reason for a child's visit to a neurologist. Ideally, decisions regarding treatment are made by the clinician based on properly designed trials. We reviewed the medical literature of the past 20 years to determine the extent to which antiepileptic drugs have been evaluated for children using randomized, double-blind trials. A Medline search was conducted for 1974 to 1993 abstracts that contained the words *randomized* or *double-blind* and one of the following words or phrases: *seizures, epilepsy, infantile spasms, West syndrome,* and *Lennox-Gastaut*. Bibliographies of textbooks and review articles were also scanned for studies that evaluated treatment options for childhood seizure disorders and epilepsy. We included articles that were limited to children as well as those that enrolled enough children to reach conclusions about them. Thirty-seven studies were found, 29 of which involved nonfebrile seizures and evaluated 849 patients. Only 4 of these studies (3 of them multicenter) enrolled more than 50 patients. The number of patients who had potentially disabling infantile spasms or Lennox-Gastaut syndrome were 93 and 138, respectively. Only 1 study of 33 patients examined partial seizures as a separate group. No randomized trial evaluated the best treatment for neonatal seizures, status epilepticus, or primary generalized tonic-clonic seizures in children. Data regarding optimum treatment of childhood seizure disorders are limited. As new drugs become available, child neurologists should consider increased involvement in randomized trials to determine the best treatments for these conditions. In order to recruit an acceptable number of patients, multicenter studies will be needed.

187. POLYMYOCLONUS AND ATAXIA IN A PATIENT WITH HOMOCYSTINURIA SUCCESSFULLY TREATED WITH BACLOFEN

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A patient with homocystinuria due to cystathionine beta-synthase deficiency developed severe progressive polymyoclonus and ataxia. To our knowledge this is the first time polymyoclonus and ataxia have been reported in association with homocystinuria. Although cerebrovascular thrombosis is usually thought to be responsible for neurologic dysfunction in homocystinuria patients, no infarctions were demonstrated on MRI in our patient. We previously reported that Baclofen dramatically improved polymyoclonus and ataxia in a patient with Unverricht-Lundborg Disease. Baclofen, given to our patient reversed the polymyoclonus, and the ataxia as well. This finding suggests that patients with polymyoclonus and ataxia, no matter what the etiology, may benefit from the use of Baclofen.

188. ULTRASTRUCTURAL CHANGES OF SPINAL CORD OF NEWBORNS WITH NEUROMUSCULAR DISORDERS

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Different neuromuscular disorders are detected quite often in the first days of life in children with cervical spine and the spinal cord trauma. The changes in the spinal cord—above and below the place of injury—were examined. The degenerative, dystrophic changes in neurons, axons, and synapses were disclosed. They were expressed in the dark and light degeneration of synapses, in the change of location and number of synaptic vesicles, lysosomes, and mitochondrial swelling. The study of neurotransmitter metabolism uncovered steadfast promotion of serotonin level in blood serum which pointed out nucleon hyperfunction of the big suture. The neuromuscular disorders developing in newborns can be explained either by initial alteration of afferent spino-cerebellar impulsation, or secondary change of "working matrytsa" of efferent rubro-spinal signal. The rise of double alteration takes place in dystrophically changed Reksed blades at the segmental level. During the pathogenesis of neuromuscular disorders, neurotransmitters also take part which steadfast pro-