the carotid bodies and the carotid body plays an important role in the development of respiratory maturation. The pineal gland influences diurnal rhythm and is significantly reduced in weight in SIDS patients compared to age matched controls; the significance of this reduction is unknown. Brains of SIDS victims born at term were significantly heavier than reference values matched for both age and body length. (Becker LE. Neural maturational delay as a link in the chain of events leading to SIDS. Can J Neurol Sci Nov 1990; 17:361-671).

COMMENT. The authors emphasize a delay of neural maturation of both myelination and synapses in the etiology of SIDS. Other abnormalities such as brainstem astrogliosis may be secondary to hypoxic-ischemia. In Canada, the incidence of sudden infant death syndrome is 1.2 per 1000 live births. The peak age is 1-4 months. The highest incidence is in winter and more boys than girls are affected. A thorough autopsy ruled out SIDS in 10% of sudden unexpected deaths occurring under one year of age. The differential diagnoses included congenital heart disease, myocarditis, central nervous system trauma (child abuse), cardiomyopathy, encephalitis, meningitis, congenital diaphragmatic hernia, and medium chain acyl coenzyme deficiency. If an anatomical cause of death is found, the diagnosis is not SIDS. In SIDS the mechanism of death must be related to a central type of respiratory failure or cardiac dysrhythmia.

CONGENITAL MYASTHENIA AND FACIAL MALFORMATIONS

A new genetic syndrome of congenital myasthenia with distinctive ethnic clustering and associated facial malformations transmitted as an autosomal recessive disorder is reported from the Departments of Neurology and Medical Genetics, Chaim Sheba Medical Center, Tel Hashomer, Sackler School of Medicine, Tel Aviv University, Israel. The syndrome was demonstrated in 14 Jewish patients from ten families of Iraqi or Iranian origin. All patients had bilateral ptosis and predominant facial muscle weakness, 11 had weak masticatory muscles, and 12 had easy fatiguability on prolonged speech. Very mild limb muscle involvement was present in only three cases. The facial malformations included an elongated face, mandibular prognathism with malocclusion and a high arched palate. The course was mild and nonprogressive, the electromyogram showed a decremental response on repetitive stimulation of either the accessory or the facial nerve but myopathic changes were not seen. Antibodies to acetylcholine receptor were absent and all patients had a response to cholinesterase inhibitors and a positive Tensilon test. There was clinical improvement with pyridostigmine. In seven of ten families there was close parental consanguinity. (Goldhammer Y et al. Congenital myasthenia associated with facial malformations in Iraqi and Iranian Jews. Brain Oct 1990; 113:1291-1306).

COMMENT. The distribution of muscle weakness in congenital myasthenia in these cases is compatible with previous reports where extraocular and facial muscle involvement have

predominated (Millichap JG, Dodge PR. Neurology 1960; 10:1007). The authors postulated that the facial abnormalities were secondary to the neuromuscular defect. Congenital myasthenia has been described in association with arthrogryposis, the subject of the following article.

ARTHROGRYPOSIS CONGENITA AND HEPATORENAL ABNORMALITIES

Arthrogryposis multiplex congenita with renal and hepatic abnormalities, demonstrated at autopsy in a two month old child of consanguineous parents, is reported from the Pediatric Hospital, Coimbra, Portugal. Three brothers and eight first cousins had died within the first month, all with jaundice. The brothers of the proband had limb abnormalities and one had polyuria, glucosuria, and metabolic acidosis. The patient was born with flexed knees and joint limitation, cubital deviation of the hands with clenched fingers, and muscular atrophy. During the second week of life the infant became jaundiced and on day 18 she was admitted with cholestatic jaundice and hepatomegaly. Electromyography and muscle biopsy were compatible with neuropathic muscular atrophy. There was hypercalcemia with increased density of the base of the skull, renal tubular degeneration, and biliary stasis with pigmentary deposits. The family pedigree suggested an autosomal recessive inheritance. (Saraiva JM et al. Arthrogryposis multiplex congenita with renal and hepatic abnormalities in a female infant. J Pediatr Nov 1990; 117:761-763).

COMMENT. This syndrome was first described by Nezelhof C et al (J Pediatr 1979; 94:258) who reported four patients with these findings. As found in this case report, arthrogryposis is most commonly associated with neuropathic muscular atrophy. The underlying lesion may be found in the anterior horn cells of the spinal cord, the peripheral nerves, the neuromyal junction, the muscle, and sometimes in the brain.

INFECTIOUS DISEASE

NEUROLOGIC MANIFESTATIONS OF LYME DISEASE

The chronic neurologic symptoms and signs in 27 patients with previous signs of Lyme disease and current evidence of immunity to Borrelia burgdorferi are reported from the Departments of Neurology and Medicine, Tufts University School of Medicine, New England Medical Center, Boston, MA. The median age was 49 years with a range of 25-72 years. Signs and symptoms of chronic neurologic abnormalities included encephalopathy in 89% with memory loss, depression, sleep disturbance, irritability, and difficulty finding words; polyneuropathy in 70% with spinal or radicular pain, distal paresthesia, and sensory loss; leukoencephalitis in 4%; and miscellaneous symptoms including fatigue (74%), headache (48%), hearing loss (15%), fibromyalgia (15%), and tinnitus (7%). Seventeen patients (63%) had abnormalities of both the central and peripheral nervous systems, seven (26%) had encephalopathy alone, two (7%) had polyneuropathy alone, and the remaining patient