patients observed in the clinic at the University of Connecticut Health Center, Farmington, were reviewed for descriptions made during the newborn period. At the time of the chart review the patients ranged in age from eight months to 33 years. All initial examinations were performed at 22 days of age or earlier. Of 13 patients with high resolution chromosome analysis 12 showed the typical deletion of the long arm of chromosome 15. Several of the characteristic features of Prader-Willi syndrome in early infancy were confirmed including hypotonia and genital hypoplasia. Features that have not previously been emphasized included an abnormal cry, disproportionately large head circumference and anterior fontanel, mild micrognathia, mild anomalies of gingivae or alveolar ridges, and changes in the appearance of the skin such as poor color, cyanosis, jaundice, ecchymoses, hirsutism, and foot edema. Hypoplasia of the scrotum was present with a normal appearing penis. Contrary to one previous report the hypotonia was associated with absence of deep tendon reflexes, and hyperreflexia was found in only one. (Aughton DJ, Cassidy SB. Physical features of Prader-Willi syndrome in neonates. AJDC Nov 1990; 144:1251-1254).

COMMENT. Prader-Willi syndrome is a sporadic multisystem disorder characterized after infancy by obesity, acromicria, short stature, hypogonadism, and abnormal cognitive and behavioral functioning. Infants exhibit hypotonia, genital hypoplasia, and feeding problems with failure to thrive. The additional neonatal characteristics described in this paper may aid in early diagnosis and counseling of parents.

NEURAL MATURATIONAL DELAY IN SIDS

The pathological evidence for developmental delay in SIDS is reviewed from the Division of Neuropathology, The Hospital for Sick Children, University of Toronto, Toronto, Canada. Evidence of hypoxic ischemic insult to the brain includes astrogliosis and subcortical leukomalacia. Astrogliosis is most apparent in the region of the tegmentum in SIDS victims, a finding interpreted as hypoperfusion during episodes of bradycardia associated with apnea. Subcortical or periventricular leukomalacia was found in 21.6% of infants who died of SIDS. Term and prematurely born SIDS infants showed persistence of the reticular dendritic spines which suggests a delay of development to a mature, higher level of neuronal function. This delay may reflect a functional impairment of the higher levels of respiratory control in SIDS infants. A number of reports suggest alterations in neurotransmitter levels of catecholamines, beta endorphin, met-enkephalin, and substance P in SIDS. Examination of 36 infants who had died of SIDS showed that the mean number of small myelinated vagal fibers was significantly decreased in the SIDS infants compared with controls, suggesting an abnormal or delayed development of the vagus nerve. This finding was similar to that reported in a two year old infant dying of persistent infantile sleep apnea (Ondine's curse). Elevated dopamine in the carotid bodies of SIDS victims suggests a role in pathogenesis: dopamine inhibits respiration by acting directly on

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