of fatal and benign cytochrome c oxidase deficient myopathies of infancy: an immunohistochemical approach. Neurology Feb 1991; 41:300-305).

COMMENT. The diagnosis of COX-deficient myopathies of infancy relies on histochemical and biochemical evaluation of COX activities in muscle biopsies. These tests fail to distinguish the fatal and benign phenotypes at early stages because both show lack of COX activity. COX is a complex enzyme composed of 13 subunits with three larger subunits (I, II and III) which are synthesized in the mitochondria and the ten smaller subunits manufactured in the cytoplasm. COX VIIa,b is absent in the fatal myopathy and both COX-II and COX VIIa,b are absent in the early stages of benign myopathy. Thus, the immunohistochemistry of COX-II is sufficient for the differential diagnosis.

NEMALINE MYOPATHY: RESPIRATORY FAILURE

A Japanese boy with nemaline myopathy diagnosed at three years of age and complicated by severe respiratory failure at 8 years is reported from the Division of Child Neurology, National Center Hospital for Mental, Nervous, and Muscular Disorders, Kodaira, Tokyo, Japan. The histologic findings of the respiratory muscles obtained during thoracic surgery for pneumothorax showed marked variation in fiber size with increase in fibrous tissue, type II fiber deficiency, elevated acid phosphatase activity, and disorganized intermyofibrillar network. Truncal and biceps muscles showed little variation in fiber size, numerous nemaline bodies and type I fiber predominance. The sudden onset of severe respiratory failure was related to the preferential and progressive involvement of the respiratory muscles. (Saski M etal, Respiratory muscle involvement in nemaline myopathy. Pediatr Neurol Nov/Dec 1990; 6:425-427).

COMMENT. Severe respiratory insufficiency is an uncommon development in nemaline myopathy, but a frequent complication of Duchenne's muscular dystrophy. Miller RG et al from the Children's Hospital of San Francisco have made serial measures of respiratory function in 17 patients with Duchenne's muscular dystrophy who underwent segmental spinal fusion and in 22 patients without operations. Declining respiratory function was observed in both groups, but operated patients showed improved sitting comfort (Neurology Jan 1991; 41:38-40).

SEIZURE DISORDERS

TREATMENT OF STATUS EPILEPTICUS

The drugs used in status epilepticus, primary care in the community, secondary hospital care, and tertiary or intensive care are reviewed from the Royal Hospital for Sick Children, Edinburgh. The two preferred drugs recommended for first line care are rectal diazepam and intramuscular paraldehyde. In second line care at a hospital emergency

room, intravenous diazepam is preferred in a dosage of 0.25 to 0.3 mg/kg with a glass syringe. Third line management in intensive care includes 20% mannitol given over 20 minutes in a dose of 7 ml/kg for cerebral edema. EEG monitoring is essential to demonstrate seizure activity, paradoxical reactions to drugs such as diazepam, and overdosage with drugs, eg barbiturates causing burst suppression. The authors stress the need to consider Hemophilus influenzae and pneumococcal meningitis as an underlying cause of status epilepticus and caution that lumbar puncture must never be done in an unconscious child without a CT scan to exclude signs of brain swelling or edema. The need for tertiary intensive care is usually a sign of failure of early control resulting from inappropriate anticonvulsant medication rather than drug resistance. (Brown JK, Hussain IHMI. Status epilepticus II: Treatment. Dev Med Child Neurol Feb 1991; 33: 97-109).

COMMENT. The primary care of seizures is all important so that status epilepticus of prolonged duration may be avoided. Mortality from status epilepticus is usually the result of the underlying disease with an 8% incidence at the above institution. A paradoxical convulsant response to the anticonvulsant diazepam should be considered in children whose seizures are not rapidly controlled, and an alternative anticonvulsant should be used (Livingston and Brown, 1988).

The home use of rectal diazepam for cluster and prolonged seizures is reported from the Department of Neurology, Hennepin County Medical Center; Department of Pharmacy Practice, University of Minnesota, Minneapolis; and the Department of Pediatric Neurology, Gillette Children's Hospital St. Paul, Minnesota (Kriel RL, Cloyd JC et al. Pediatr Neurol Jan/Feb 1991; 7:13-17). Rectal diazepam was effective in controlling seizures in 85% of patients. Adverse reactions were mild and consisted of drowsiness and/or behavioral changes. Improvements in quality of life associated with the availability of rectal diazepam were observed by 58% of users and 27% of nonusers. In addition to improved management of seizures there was increased flexibility in family activities and less parental anxiety. The diazepam injectable solution in a dose ranging from 0.3 to 0.5 mg/kg was used for rectal administration. It was administered with a needleless lubricated 3 ml plastic syringe inserted 2 to 4 cm into the rectal cavity. Effective serum concentrations are usually reached within ten minutes.

METHSUXIMIDE FOR INTRACTABLE SEIZURES

The use of methsuximide in 25 children with intractable epilepsy is reported from the Department of Neurology and School of Pharmacy, University of North Carolina, Chapel Hill. In 15 patients methsuximide was well tolerated and resulted in a 50% or greater reduction in seizure frequency. The predominant seizure types were tonic, complex partial, secondary generalized and astatic/myoclonic. The EEG showed generalized slow spike and wave in 14 and focal spikes in three. Two