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