Neurology, Pediatrics and Cell Biology, St. Radboud University Hospital, Nijmegen, Netherlands. There was a three month history of weakness in the legs, sudden falling, inability to run and difficulty in climbing stairs. Within two months he developed weakness in his hands, a wobbling gait and a positive Gowers' sign. Muscle tone was impaired and proximal muscles were wasted. Biochemical studies on muscle tissues showed a defect of NADH dehydrogenase (complex I). Dramatic improvement followed oral treatment with L-carnitine 2 g daily and riboflavin 9 mg daily. Seven months later the complex I activity was normal, Gowers' sign was negative, deep tendon reflexes had returned and muscle weakness was limited mainly to the peroneal muscles. Nine further cases of a pure myopathy associated with complex I deficiency are reviewed (Bernsen PLJA et al. Successful treatment of pure myopathy, associated with complex I deficiency, with riboflavin and carnitine. Arch Neurol March 1991; 48:334-338).

COMMENT. The present case report of a mitochondrial myopathy differed from nine other reports. There was no history of excessive fatiguability or exercise intolerance, the serum lactate concentration was normal, and the muscle biopsy did not show ragged red fibers. The dramatic improvement with riboflavin and carnitine and the return to normal of complex I activity were unusual findings. The authors stress the clinical, biochemical and morphological heterogeneity in complex I deficiency. Biochemical studies of mitochondrial metabolism in the muscle are advisable in children with progressive weakness and exercise intolerance when a precise diagnosis is unclear.

FATAL AND BENIGN CONGENITAL MYOPATHIES: DIFFERENTIAL DIAGNOSIS

Muscle biopsies from four infants with fatal myopathy and four with benign myopathy were examined using biochemical, histochemical and immunohistochemical techniques in the Departments of Neurology, Pathology and Genetics, Columbia University, New York; Universita Cattolica del Sacro Cuore, Rome, Italy; and Fachbereich Chemie, Philipps-Universitat, Marburg, Germany. At early stages the clinical picture failed to provide clues for differential diagnosis; both fatal and benign myopathies presented with hypotonia, generalized weakness, lactic acidosis, failure to thrive and severe respiratory insufficiency, often requiring assisted ventilation. Patients with benign myopathy gain strength and show an increase in the number of fibers with COX (cytochrome c oxidase) deficient activity. In contrast, patients with fatal myopathy die before one year of age and muscle histochemistry shows no increase in the number of fibers with COX activity. The subunit composition of COX was studied directly on frozen muscle sections using immunologic probes. The fatal type was characterized by absence of the nuclear DNA-encoded subunit VIIa,b of COX while in the benign myopathy both VIIa,b and the mitochondrial DNA-encoded subunit II were absent. From a practical standpoint, immunohistochemistry of COX-II should suffice for differential diagnosis because this subunit was present in the fatal myopathy but absent in the early stages of the benign myopathy (Tritscler HJ, DiMauro S et al Differential diagnosis

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