Nemaline myopathy (NM) is a heterogeneous disorder defined by the presence of rod-shaped structures known as nemaline bodies or rods. The diagnosis is based on muscle weakness, combined with visualization of nemaline bodies on muscle biopsy. There is no curative treatment for nemaline myopathy. Therapeutic strategies for this condition are symptomatic and empirical.

Herein, we present a newborn with severe respiratory failure and generalized muscle weakness, who was diagnosed as NM by muscle biopsy. The patient experienced remarkable decrease in sialorrhea and improvement of spontaneous movements after L-tyrosine treatment. This case is presented to emphasize the importance of muscle biopsy in the differential diagnosis of severe hypotonia during neonatal period and a possible benefit of L-tyrosine supplementation for decreasing sialorrhea and restoring muscle strength.

**Keywords**: Infant, newborn; L-tyrosine; Muscle hypotonic; Nemaline myopathy.

Sociedad Argentina de Pediatría.