ELECTROMYOGRAPHY OF THE DIAPHRAGM AND PHRENIC NERVE CONDUCTION STUDIES IN PATIENTS WITH ALS

S. Podnar, I. Rigler
Institute of Clinical Neurophysiology, University Medical Centre Ljubljana, Ljubljana, Slovenia

In patients with restrictive ventilation failure of unknown aetiology neuromuscular disorders always need to be considered. In suspected lesions to the phrenic anterior horn neurons or motor axons or to the diaphragm electrodiagnostic investigation, including phrenic nerve conduction studies and needle examination of the diaphragm muscle, can be useful. Phrenic nerve conduction studies are usually done using a supramaximal (80–100 mA, 0.1–0.2 ms long) electrical stimulation applied by a surface electrodes in the supraclavicular fossa (just above the clavicle, between both heads of the sternocleidomastoid (SCM) muscle) (1), and with a pair of self adhesive recording electrodes on the thorax (the active electrode (G1): 5 cm above the xiphoid process, the reference electrode (G2): 16 cm from G1, on the chest margin (2)). Phrenic nerve M-wave parameters describing axonal loss (amplitude (mV), and area (mVms)), or demyelination (latency and duration and (ms)) can be measured. Needle examination of the diaphragm muscle is usually done by a standard concentric EMG needle inserted into the medial recess of the 7th, 8th or 9th intercostal space (3). During slow advancement of the needle electrode through the tissue EMG signal is carefully observed and listened. Rhythmic bursts of low amplitude MUPs during inspirations disclose that tip of the electrode reached the diaphragm (3). At that point EMG activity during normal breathing is observed. In addition, motor unit potential (MUP) parameters can be measured (quantitative MUP analysis), and compared to normative limits (1).

Respiratory electrodiagnostic studies are valuable also in patients with suspected ALS, because some of these patients present with respiratory insufficiency. Furthermore, respiratory failure limits the life span of these patients. However, in this population apart from lower motor neuron (at high cervical or thoracic levels) upper motor neuron involvement may cause of respiratory failure. Phrenic nerve conduction studies and needle examination of the diaphragm muscle, however, evaluate mainly only the lower motor neuron function. Low amplitude or area of the phrenic nerve M-wave point to few remaining functional diaphragm muscle fibres, and therefore to a advanced stage of the disease in the respiratory domain. The needle EMG studies may be of some prognostic value and aid in long-term planning of respiratory management in patients with ALS. Abundant spontaneous denervation activity with a decreased number of relatively normal-sized MUPs points to acute denervation and rapidly developing disease with poor reinnervation. In contrast, little spontaneous denervation activity with a decreased number of large MUPs indicates a more slowly progressive denervation and better reinervation. Respiratory electrodiagnostic findings need to be combined with electrodiagnostic findings obtained in other trunk and limb nerves and muscles to diagnose ALS.

References:

1. Resman-Gasperšič A, Podnar S. Phrenic nerve conduction studies: technical aspects and normative data. Muscle Nerve 2008; 37: 36–41.

2. Chen R, Collins S, Remtulla H, Parkes A, Bolton CF. Phrenic nerve conduction study in normal subjects. Muscle Nerve 1995; 18: 330–5.

3. Bolton CF, Grand'Maison F, Parkes A, Shkrum M. Needle electromyography of the diaphragm. Muscle Nerve 1992; 15: 678–81.