

factor is reduced. Thus in myasthenia gravis, repetitive stimulation, particularly at a rate of between 2 and 5 Hz, may lead to a depression of neuromuscular transmission, with a decrement in size of the response recorded from affected muscles. Similarly, immediately after a period of maximal voluntary activity, single or repetitive stimuli of the motor nerve may elicit larger muscle responses than before, indicating that more muscle fibers are responding. This postactivation facilitation of neuromuscular transmission is followed by a longer-lasting period of depression, maximal between 2 and 4 min after the conditioning period and lasting for as long as 10 min or so, during which responses are reduced in size.

Decrementing responses to repetitive stimulation at 2–5 Hz are common in myasthenia gravis but may also occur in the congenital myasthenic syndromes (Chap. 461). In Lambert-Eaton myasthenic syndrome, in which there is defective release of acetylcholine at the neuromuscular junction, the compound muscle action potential elicited by a single stimulus is generally very small. With repetitive stimulation at rates of up to 10 Hz, the first few responses may decline in size, but subsequent responses increase. If faster rates of stimulation are used (20–50 Hz), the increment may be dramatic so that the amplitude of compound muscle action potentials eventually reaches a size that is several times larger than the initial response. In patients with botulism, the response to repetitive stimulation is similar to that in Lambert-Eaton myasthenic syndrome, although the findings are somewhat more variable and not all muscles are affected.

#### **SINGLE-FIBER ELECTROMYOGRAPHY**

This technique is particularly helpful in detecting disorders of neuromuscular transmission. A special needle electrode is placed within

a muscle and positioned to record action potentials from two muscle fibers belonging to the same motor unit. The time interval between the two potentials will vary in consecutive discharges; this is called the *neuromuscular jitter*. The jitter can be quantified as the mean difference between consecutive interpotential intervals and is normally between 10 and 50  $\mu$ s. This value is increased when neuromuscular transmission is disturbed for any reason, and in some instances impulses in individual muscle fibers may fail to occur because of impulse blocking at the neuromuscular junction. Single-fiber EMG is more sensitive than repetitive nerve stimulation or determination of acetylcholine receptor antibody levels in diagnosing myasthenia gravis.

Single-fiber EMG can also be used to determine the mean fiber density of motor units (i.e., mean number of muscle fibers per motor unit within the recording area) and to estimate the number of motor units in a muscle, but this is of less immediate clinical relevance.

#### **BLINK REFLEXES**

Electrical or mechanical stimulation of the supraorbital nerve on one side leads to two separate reflex responses of the orbicularis oculi—an ipsilateral R1 response having a latency of approximately 10 ms and a bilateral R2 response with a latency in the order of 30 ms. The trigeminal and facial nerves constitute the afferent and efferent arcs of the reflex, respectively. Abnormalities of either nerve or intrinsic lesions of the medulla or pons may lead to uni- or bilateral loss of the response, and the findings may therefore be helpful in identifying or localizing such pathology.