EDX Studies of the Facial Nerve in Peripheral Facial Palsy and Hemifacial Spasm

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CME Information
Product: MM23 - EDX Studies of the Facial Nerve in Peripheral Facial Palsy and Hemifacial Spasm

Course Description
Electrodiagnostic (EDX) assessment is one of the most important aspects in the evaluation of the two most common disorders of the facial nerve: facial palsy and hemifacial spasm. Facial palsy is usually an acute disorder that resolves in a few weeks but, in a number of cases, leads to a postparalytic facial syndrome featuring muscle synkinesis, myokymia, and involuntary mass contractions of muscles on the affected side. Hemifacial spasm is usually a chronic disorder characterized by paroxysms of involuntary, clonic, and synchronous twitching of all facial muscles on the affected side. EDX studies provide information on lesion location and severity, pathophysiology underlying the two disorders, and differential diagnosis between syndromes presenting with abnormal facial muscle activity. This monograph is intended to describe the most relevant EDX findings in the two disorders and the most appropriate timing for the examinations in order to provide useful information for prognosis and therapeutic decision-making.

Intended Audience
This course is intended for Neurologists, Psychiatrists, and others who practice neuromuscular, musculoskeletal, and electrodiagnostic medicine with the intent to improve the quality of medical care to patients with muscle and nerve disorders.

Learning Objectives
Upon conclusion of this program, participants should be able to:
   1. recognize the most relevant electrodiagnostic findings in hemifacial spasm and facial palsy.
   2. recognize the most appropriate timing for the examinations to provide useful information for prognosis and therapeutic decisions.

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ABSTRACT: Electrodiagnostic (EDX) assessment is one of the most important aspects in the evaluation of the two most common disorders of the facial nerve: facial palsy and hemifacial spasm. Facial palsy is usually an acute disorder that resolves in a few weeks but, in a number of cases, leads to a postparalytic facial syndrome featuring muscle synkinesis, myokymia, and involuntary mass contractions of muscles on the affected side. Hemifacial spasm is usually a chronic disorder characterized by paroxysms of involuntary, clonic, and synchronous twitching of all facial muscles on the affected side. EDX studies provide information on lesion location and severity, pathophysiology underlying the two disorders, and differential diagnosis between syndromes presenting with abnormal facial muscle activity. This monograph is intended to describe the most relevant EDX findings in the two disorders and the most appropriate timing for the examinations in order to provide useful information for prognosis and therapeutic decision-making.


ELECTRODIAGNOSTIC STUDIES OF THE FACIAL NERVE IN PERIPHERAL FACIAL PALSY AND HEMIFACIAL SPASM

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The facial nerve is one of the most complex nerves in the human body. Its motor nucleus lies in the caudal pontine tegmentum and its fibers run rostrally to surround the nucleus of cranial nerve VI and then turn caudally to emerge from the pons near the cerebellopontine angle. At its exit from the brainstem, the facial nerve appears to be formed by two roots: the facial nerve itself that carries motor fibers innervating all facial muscles, and the nervus intermedius of Wrisberg that carries preganglionic parasympathetic fibers innervating the submandibular ganglion and the pterygopalatine ganglion. The most relevant structure in the petrous course of the facial nerve is the geniculate ganglion, located at the genu of the facial nerve, between the labyrinthine and the tympanic segments. After its exit from the skull at the stylomastoid foramen, but before entering the parotid gland, the facial nerve issues collateral branches to the posterior auricular muscle, the posterior belly of the digastric muscle, and the stylohyoid muscles. The actual distribution of the terminal divisions of the facial nerve varies among individuals, but zygomatic, buccal, mandibular, and cervical branches are usually identified.

The two most common disorders of the facial nerve are facial palsy and hemifacial spasm. Lesions in the facial nerve can occur at various points along its course. The lesion location can be determined approximately by taking into account the presence or absence of signs revealing involvement of collateral branches.53 The most vulnerable nerve segment is the intrapetrosal segment where a relatively minor inflammatory event may cause nerve compression due to the non-distensibility of the osseous canal. Benign tumors growing in the pontine–medullary–cerebellar angle as well as parotid tumors can also cause compression of the facial nerve. The natural history of facial palsy does not seem to be determined by the cause of the lesion but rather by the extent of axonal damage. Compression from an artery, usually the posterior inferior cerebellar artery, on top of the nerve at the posterior fossa has been implicated as the cause of hemifacial spasm.
Peripheral Facial Palsy

Peripheral facial palsy can be due to many causes, including infections, trauma, vascular lesions, and tumors. Idiopathic facial palsy, or Bell’s palsy, is a relatively benign condition with an incidence rate of about 25 per 100,000.14 It may occur at any age, although it is more frequent in adults than children.13 In a histopathological study performed on a patient who died from another cause 1 week after onset of Bell’s palsy, Liston and Kleid17 found diffuse inflammatory infiltrates with swollen macrophages containing demyelination debris, suggesting an inflammatory process with axonal damage. The finding of an increased titer of antibodies against herpes simplex virus in a number of patients suggests that a localized infection by this virus may be the cause of Bell’s palsy in many patients.82

In idiopathic facial palsy, the first symptoms vary among patients. Some patients notice an inability to retain liquid in the mouth; others are told by relatives or friends that their mouth is displaced to one side. In most instances, peripheral facial palsy is accompanied by severe emotional distress that usually fades away after a thorough explanation of the cause of the paralysis and of its presumed evolution and by the patient’s own appraisal of the nonprogressive character of the lesion. A short time after the onset of facial palsy, corneal irritation and limited eyelid movements may trigger compensatory and adaptive changes apparent only on the side contralateral to the paralysis. This usually presents with an increased frequency of blinking and, on some occasions, muscle hyperactivity.2,5 If the lesion is limited to neurapraxia, Bell’s palsy resolves in a few weeks, leaving no trace of nerve damage. However, if a significant percentage of axons undergo Wallerian degeneration, a variable degree of abnormal regeneration should be expected.24 Regenerating errors in a pure motor nerve such as the facial nerve may be one of two types: (1) the axon generated in a motoneuron previously activating one muscle ends up in a funiculus going to another muscle; and (2) a single axon branches into two or more axons passing to different muscles that may have antagonistic functions. In the case of the facial nerve, regenerating axons may undergo regenerating errors not only among facial motor axons, but also between facial motor axons and parasympathetic axons of the nervus intermedius of Wrisberg. Parasympathetic axons may enter the facial nerve and reach motor end-plates, and motor axons of the facial nerve may enter the parasympathetic funiculi and reach the lacrimal gland or the gustatory receptor. Lacrimation and hemifacial sweating when activating facial muscles are consequences of such reinnervation errors. Reinnervation abnormalities may lead to either subclinical or clinically evident synkinesis. In some instances, hemifacial synkinesis may be so severe as to lead to massive contractions of all the muscles on one side of the face.

Neurophysiological examination is helpful at all stages in patients with facial palsy (Table 1). In the first few days, electrically induced blink reflexes16 or the assessment of transpetrosal facial nerve conduction time provide information on the degree of axonal conduction block. At about 10 days from onset of symptoms, the size of the compound muscle action potential (CMAP) reflects the relative functional loss of axons on the affected side. Approximately 20 days after onset, needle electromyography (EMG) brings an approximate measure of the intensity of denervation. If axonal damage has occurred, axonal regeneration may first be seen in perioral or periocular muscles 3 months after the onset of symptoms. In these patients, subsequent neurophysiological examination may provide evidence of abnormal regeneration and changes in motoneuronal excitability, characteristic of the postparalytic facial syndrome.25 It is convenient, at all of these stages, to perform neurophysiological examinations on both the paralyzed and nonparalyzed sides. The tests to be applied depend on the evolution of the symptoms, not all of which may be informative in all patients.

Electrophysiological Methods. Blink Reflex. The simplest method to obtain the blink reflex is by stimulating the supraorbital nerve with the cathode placed on the supraorbital notch and the anode 2–3 cm away in the course of the supraorbital nerve. Recordings are obtained from the orbicularis oculi of both sides with surface electrodes, the active one in the middle of the lower eyelid and the reference 2–3 cm lateral to it.16 The reflex circuit involves the afferent fibers of the trigeminal nerve, a pontine oligosynaptic circuit for a unilateral short-latency response (R1), a spinal polysynaptic circuit for bilateral long-latency responses (R2 and R2c), and the efferent fibers of the facial nerve.28 A lesion in the facial nerve is characterized by absent (or abnormally small and delayed) R1 and R2 responses on one side, with normal responses on the other, to stimulation of the supraorbital nerve of either side (Fig. 1). An additional observation in most patients with facial palsy is a difference in the size of the R2 and the R2c responses generated in the nonaffected orbicularis oculi to ipsilateral or contralateral stim-
In healthy subjects, the late response to ipsilateral stimuli (R2) is slightly larger than the corresponding response to contralateral stimuli (R2c). However, in patients with facial palsy this correlation is reversed, pointing to an enhanced gain of the blink reflex to inputs from the ophthalmic nerve of the paralyzed side. The electrodes in the lower eyelid also allow for recording of the EMG activity and eyelid movement during spontaneous blinking. This involves activation of the orbicularis oculi and relaxation of the levator palpebrae. Therefore, even with an apparently complete facial palsy, the eyelid may move slightly downwards during spontaneous blinking, a movement that can be recorded as a slow negative potential with the electrodes placed on the lower eyelid if the bandpass frequency filters are

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**Table 1.** Electrophysiological tests at different stages of the evolution of peripheral facial palsy.

**FIGURE 1.** Blink reflex to supraorbital nerve electrical stimulation in a healthy volunteer (left panel) and in a patient with right-sided Bell’s palsy 1 week after onset (right panel). Recordings were taken from the orbicularis oculi of the right (rOOc) and left (lOOc) sides. The recordings show the R1 and R2 responses on the side ipsilateral to the stimulus, and the R2c on the contralateral side. Note the absence of a response in the rOOc in the patient. Also note that the R2c response generated on the IOOc by stimuli applied to the side of the paralysis is larger than the R2 generated in the same muscle by stimuli applied to the nonaffected side. This is not the case in the healthy volunteer, who has R2 responses larger than R2c responses on both sides.
wide open (e.g., 0.5–1000 Hz).

Transpetrosal Conduction Time. The facial nerve can be stimulated at the posterior fossa using a magnetic coil. The coil, usually round and 90 mm in diameter, is positioned tangentially in the parieto-occipital region with a clockwise current orientation for the right side and counterclockwise for the left side. This gives rise to a response in the nasalis muscle, with a latency between 5.1 and 6.3 ms and peak-to-peak amplitude between 0.5 and 1.2 mV in healthy volunteers. The nerve can be stimulated electrically at its emergence from the stylomastoid foramen; the petrosal conduction time is calculated as the latency difference between the two responses. Care must be taken not to induce large artifacts that would prevent a good evaluation of the response onset latency.

Facial Compound Muscle Action Potential. The most informative means for determining the degree of axonal loss early after onset of the lesion is testing of the facial CMAP. The most appropriate muscle is likely the nasalis. The muscle response can be recorded by the active electrode placed over the nasalis of the stimulated side and the reference on the contralateral side. In that way, reversing the polarity with stimulation of the other side would be enough to obtain a clear and objective comparison of CMAP amplitudes. Other muscles may also be used with similar results. However, care must be taken while recording over the orbicularis oculi, as responses can be obtained there from activation of the masseter or temporalis muscles on the side of facial palsy if a high stimulus intensity is used.

Needle Electromyography. Needle recording may provide definitive confirmation of muscle denervation and, consequently, confirmation of the degree of axonal damage. This can be assessed by the amount of fibrillation potentials and positive sharp waves appearing only at a certain timepoint after nerve damage (i.e., beyond 2 weeks). Muscles mostly used for needle EMG recording are the orbicularis oris and the orbicularis oculi. In complete facial palsy only denervation potentials are observed at rest and no motor unit action potentials (MUAPs) appear on attempted muscle contraction. However, at approximately 1 month after a lesion accompanied by complete denervation of facial muscles, some MUAPs are usually obtained in the orbicularis oris with a needle inserted lateral and caudal to the oral commissure. These MUAPs are usually small-amplitude polyphasic potentials that can be activated not by stimulation of the affected facial nerve but by stimulation of the contralateral one. The exact pathophysiology underlying the contralateral innervation of perioral muscles is not completely understood at present.

Needle EMG also allows the first signs of reinnervation from the ipsilateral facial nerve to be recorded at approximately 3 months after onset of the lesion. These are seen in the orbicularis oris as polyphasic potentials that are activated during voluntary or automatic contraction but are difficult to elicit by direct electrical stimulation of the facial nerve axons. Cossu et al. demonstrated that the first responses to facial nerve stimulation are indeed reflex responses, which have similar latencies (between 44 and 132 ms) when stimulating the supraorbital nerves or the contralateral facial nerve. These responses were thought to result from activation of trigeminal cutaneous terminals and synaptic excitation of facial motoneurons. These observations indicate that, at the onset of reinnervation, facial motoneurons are more excitable to transsynaptic inputs than the axons of the facial nerve to electrical current. The findings also indicate that lateral spread of excitation to motoneurons innervating lower facial muscles occurs from the very first time that reinnervating axons reach the orbicularis oris muscle fibers.

Nerve regeneration may take place not only in the affected facial nerve but the contralateral facial nerve as well. This might be due to a widespread induction of regenerating processes, triggered by chemical changes generated at the lesion site, in a process similar to the motoneuronal reorganization that has been shown to occur in distant muscles after local denervation induced by botulinum toxin injections. However, in sphincter-like muscles, such as the orbicularis oris, denervation of circular muscle fibers may be a strong additional stimulus for the contralateral facial nerve to undergo axonal regeneration changes. Perioral muscle fibers denervated on the side of the paralysis and innervated by axons from the contralateral facial nerve may be one explanation for the finding of contralateral innervation.

Simultaneous Recording of Facial Muscles Bilaterally. Simultaneous recording of various facial muscles using surface electrodes is useful to document the possible sequelae of facial palsy and contribute to the differential diagnosis between the postparalytic facial syndrome and hemifacial spasm. Typically, recordings are performed of the orbicularis oris (electrodes at the lateral oral commissure) and the orbicularis oculi (electrodes on the lower eyelid). When at rest, the only activity seen in normal subjects consists of bursts of EMG activity in the orbicularis oculi during spontaneous blinking. In patients with synkinesis of hemifacial muscles, such bursts of EMG
activity in the orbicularis oculi are accompanied by simultaneous activity in the orbicularis oris. The same is seen with voluntary contraction. Although the patient intends to contract an individual muscle, other hemifacial muscles are recruited, leading to an unwanted facial movement. However, a subtle difference can be observed between synkinesis in postparalytic facial syndrome and hemifacial spasm. In postparalytic facial syndrome, spontaneous blinking is always accompanied by activation of the orbicularis oris, whereas in hemifacial spasm there are occasions in which there is no synkinetic activation (Fig. 2).

Hemifacial spasm

Gowers is credited with the first description of the syndrome of hemifacial spasm in 1888. This consists of paroxysms of rapid irregular clonic twitching or phasic contraction. The abnormal activity usually starts in muscles around the eyes and eventually spreads to involve all hemifacial muscles. In rare cases the hemifacial spasm is bilateral and, in this situation, the abnormal activity is not synchronous. In accordance with the prevailing hypothesis, essential hemifacial spasm could be considered, strictu sensu, the consequence of chronic subclinical facial nerve damage. Many different researchers have localized the site of origin of the facial spasms to vascular compression of the nerve at its root exit zone, even though the compressing vessel is not always found.\textsuperscript{1,19,24} Consistent with this hypothesis, patients improve after surgical intervention at the posterior fossa.\textsuperscript{12} However, there is also some proof that facial motoneurons are hyperexcitable in essential hemifacial spasm. It is likely that extrinsic irritation of the facial nerve at the posterior fossa generates an antidromic bombardment of inputs to facial motoneurons, causing excitability changes and spontaneous or reflex firing of motoneurons after a “kindling” effect.\textsuperscript{20,21}

Electrophysiological examination of patients with hemifacial spasm usually shows bursts of high-frequency impulses that can reach up to 150 impulses per second, irregular repetition of the bursts, and synchronization of impulses in various hemifacial muscles and induction of abnormal bursts by antidromic volleys in the facial nerve. The pathophysiological mechanisms underlying such abnormal activity are ectopic generation of discharges, ephaptic transmission, and lateral spread of excitation between facial axons.\textsuperscript{25} The characteristic abnormality seen in hemifacial spasm is the recording of a response in the orbicularis oris to electrical stimuli applied over the supraorbital nerve, when the trigeminofacial reflex should be limited to the orbicularis oculi. There are various possible pathophysiological mechanisms to account for the generation of

FIGURE 2. Surface electromyographic (EMG) recording from left orbicularis oculi and orbicularis oris in a patient with hemifacial spasm (A), and in a patient with facial palsy who complained of muscle tension in the cheek (B). In this case, there are myokymic discharges in the orbicularis oris in the patient with facial palsy. Note the differences between the two cases in that the abnormal synkinetic EMG activity recorded in the orbicularis oris with blinking is not always present in the patient with hemifacial spasm, and in the differences between the type of abnormal activity (high-frequency, highly synchronized, repetitive firing of action potentials in hemifacial spasm and low-frequency, small action potentials firing irregularly in facial palsy).

FIGURE 3. Schematic representation of possible mechanisms for the generation of responses in lower facial muscles by electrical stimulation at the supraorbital region. In normal subjects (straight line), the expected depolarization of the supraorbital nerve will lead to impulses traveling through the trigeminal nerve and brainstem circuits to reach the facial motoneurons. In patients with hemifacial spasm, a zone of ephapsis in the facial nerve could lead to lateral spread of activity to the fibers going to the orbicularis oris, or a secondary change in motoneuronal excitability could allow for activation of orbicularis oris motoneurons. Another possible mechanism (dashed line) is the generation of ephaptic responses in the facial nerve after antidromic activation of facial nerve axon terminals by the electrical stimulus at the supraorbital region.
such an abnormal orbicularis oris response (Fig. 3). In the simplest mechanism, the electrical stimulus would activate the supraorbital nerve afferents that reach the facial motoneurons of the orbicularis oculi muscles as in the normal blink reflex. However, the facial nerve axons to the orbicularis oculi would cause a lateral spread of excitation to the axons directed to the orbicularis oris at the site of the presumed nerve demyelination. A variant of that mechanism is that the supraorbital nerve stimulus activates not only the facial motoneurons directed to the orbicularis oculi but also those of the orbicularis oris, due to increased facial motoneuronal excitability, facial nucleus reorganization, or even interneuronal hyperexcitability. A further possibility is that the electrical stimulus activates not only the supraorbital nerve but also the terminal axons of the facial nerve running in the superficial facial muscles under the electrode. These would carry the volley until the site of presumed demyelination and induce the ephaptic response in the lower facial muscles. It is certainly possible that all three mechanisms contribute in part to the generation of abnormal reflex responses.

Lateral spread is a logical possibility if there is a zone of demyelination, as described by Jannetta and colleagues. Hyperexcitability of facial motoneurons is also a logical phenomenon after continuous antidromic bombardment of inputs. Finally, ephaptic transmission that has been demonstrated by direct activation of main facial nerve fibers is likely to occur from the very terminal facial nerve axons that are activated under the electrode. Montero et al. demonstrated that this is the case by obtaining a reflex response in the orbicularis oris that progressively shortened its latency when the stimulator was moved from the supraorbital nerve toward the malar zone. With such a maneuver, the length of the facial nerve axoaxonal ephaptic arc becomes progressively shorter while the length of the trigemino-facial reflex arc becomes progressively longer. The progressive shortening of response latency favors the belief that ephaptic mechanisms play an important role in the generation of abnormal facial reflex responses in muscles other than the orbicularis oculi. This interesting observation allows for a simpler explanation of reflex responses being induced by stimuli over the supraorbital nerve in patients having undergone hypoglossal–facial nerve anastomosis due to non-regenerating facial palsy. The identification of the ephaptic faciofacial nature of the orbicularis oculi responses renders plastic changes in the brainstem, supposedly connecting the trigeminal and hypoglossal nuclei, as an unlikely hypothesis.

As in patients with facial palsy, facial motoneuronal hyperexcitability is likely to contribute, together with peripheral nerve mechanisms, to clinico-electrophysiological manifestations of essential hemifacial spasm. In patients with essential hemifacial spasm, the elicitation of a larger R2 response on the affected side compared to the contralateral side has been taken as a sign suggesting motoneuronal excitability enhancement. An increased recovery of the blink reflex response to the test stimulus with the paired stimulus technique leading to a shift to the left of the blink reflex excitability recovery curve has suggested brainstem interneuronal excitability enhancement.

Treatment with botulinum toxin markedly alleviates hemifacial spasm and the abnormal activity occurring in patients with postparalytic facial syndrome. However, even though patients experience noticeable improvement, the abnormal muscle contraction does not completely disappear, and electrophysiological signs of hyperexcitability remain. Although hemifacial spasm and the hemifacial mass contractions observed in some patients with facial palsy may have similar clinical expressions and respond to similar treatments, several neurophysiological differences should help in distinguishing between the two syndromes. It is better to avoid using terms such as postparalytic spasm to refer to the mass contractions seen in patients with facial palsy.

REFERENCES