12 Neurophysiological Investigations

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Core Messages

- ✔ Neurophysiological investigations go beyond electromyographic recordings
- ✔ Evoked potentials (motor and sensory) allow for the assessment of spinal fiber tracts
- ✔ Electromyography and nerve conduction studies focus on the peripheral nerves
- ✔ Electrodiagnostics distinguish between acute nerve damage and preexisting neuropathies
- ✔ Neurophysiological reflex studies provide additional information about clinical reflexes
- ✔ Intraoperative monitoring improves neuroprotection in scoliosis surgery
- ✔ Electrodiagnostics predict clinical recovery in spinal cord injury (SCI)
- ✔ Subclinical spinal cord impairment can be objectified by neurophysiological recordings
- ✔ Electrodiagnostics confirm the clinical relevance of spinal cord pathologies exposed by neuroimages (morphological description by CT or MR)

Historical Background

Electrical activity within the muscle is recorded by electromyography

The history of electrodiagnostics started in the 17–18th centuries with the discovery in frogs that stroking a nerve generates a muscle contraction (Jan Swammerdam, 1637–1680) and the development by Alessandro Volta (1745–1827) of the first device to produce electricity and to stimulate muscles (the term "volt" is named in his honor). Luigi Galvani (1737–1798) made the first approaches to neurophysiology by applying electrical stimulation to muscular tissue and recording muscle contractions and force. The proof of electrical activity in voluntary muscle contractions was demonstrated in 1843 by Carlo Matteucci (1811–1868) in frogs and by Emil Du Bois-Reymond (1818–1896) in humans. This was the basis for the term "**electromyography**" (EMG). Following Charles Sherrington's (1857–1952) proposal of the concept of the motor unit in 1925 and the invention of the concentric needle electrode by E.D. Adrian and D.E. Bronk in 1929, the clinical application of electrophysiological observations was developed [23]. Finally, Herbert Jasper (1906–1999) developed the first electromyography machine at McGill University (Montreal Neurological Institute), marking the broad introduction of EMG into clinical practice [3].

Evoked potentials allow for online surveillance of spinal cord function during surgery

The assessment of spinal pathways has been made possible by the introduction of **somatosensory evoked potential** (SSEP) recording since 1970 [the first guidelines for SSEPs by the American Association of Electrodiagnostic Medicine (AAEM) were released in 1984] and **motor evoked potential** (MEP) recording from about 20 years ago. In 1980, P.A. Merton and M.H. Morton published the first study on the stimulation of the cerebral cortex in the intact human subject [28]. Anthony Barker at the University of Sheffield introduced a device for transcranial magnetic stimulation (TMS) as a new clinical tool for non-invasive and painless stimulation of the cerebral cortex [9]. Using the principle that a time-

varying magnetic field will induce an electrical field for the activation of excitatory neurons enables MEPs to be recorded from several muscles.

Intraoperative neuromonitoring started in the late 1970s

In the late 1970s, **intraoperative neuromonitoring** using SSEPs during the correction of scoliosis was introduced, while recording using MEPs due to electrical stimulation was introduced in the mid 1990s [14].

Neuroanatomy

The spinal cord covers upper and lower motoneuron pathways

In spinal disorders, an involvement of the central (CNS) and/or peripheral (PNS) nervous systems has to be considered [35]. While radiculopathies and lesions of the cauda equina exclusively affect branches of the PNS (radicular motor and sensory nerve fibers), spinal disorders inducing spinal cord malfunction almost always compromise both CNS and PNS structures. The **alpha-motoneuron** located in the central part of the spinal cord (ventral horn of the gray matter) represents the most proximal part of the peripheral motor fibers. Motor fibers from the alpha-motoneuron up to the motor endplates in the muscles constitute the secondary motor pathways, and lesions within this system show characteristic (clinical and electrophysiological) findings of a PNS lesion (lower motoneuron), e.g., flaccid weakness with muscle atrophy and signs of neurogenic denervation. In contrast, the peripheral sensory nerve fibers originate at the dorsal root ganglion, which is located outside the spinal canal. Therefore, in contrast to the motor fibers, even severe intramedullary lesions do not affect the peripheral branch of the sensory nerve fibers, and sensory nerve conduction studies remain normal.

Severity of SCI is related to localization, somatotopic extent and completeness of the lesion

The **somatotopic organization** (Fig. 1) of the longitudinal as-/descending spinal tracts (corticospinal, dorsal column, spinothalamic) allows the differentiation of the axial distribution of a lesion affecting more the anterior, posterior or central part of the cord, as well as the hemicord or total cord [24]. The sagittal localization and extension of a lesion are represented in the affection of motor

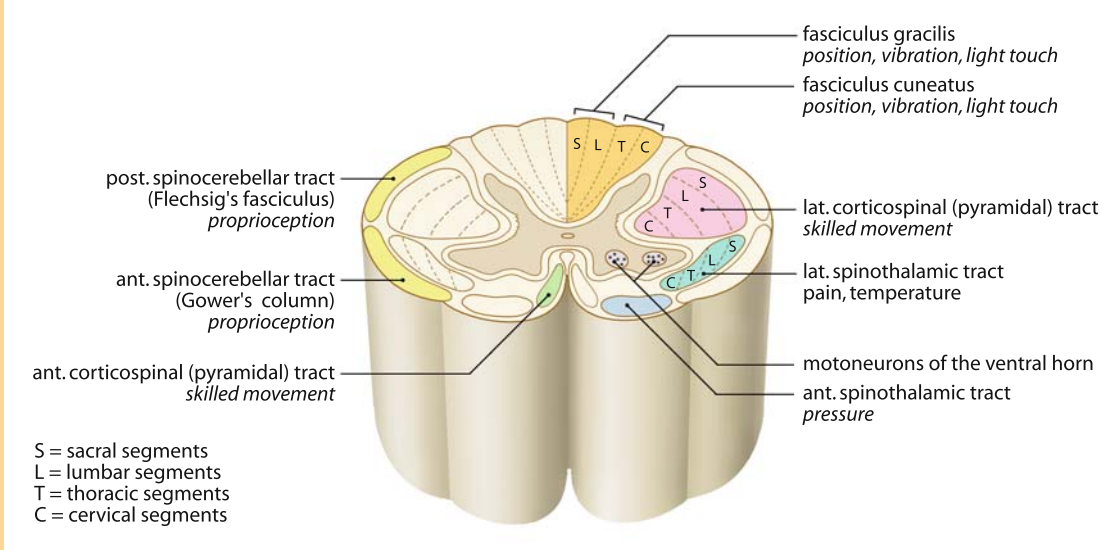

Figure 1. Somatotopic organization of the spinal cord

and sensory segments and can be demonstrated by the affected motor levels (extent of segments with denervation) as assessed by EMG. It has to be acknowledged that the intramedullary segments are more rostrally located than the related nerve roots and the alpha-motoneurons are distributed in columns over several segments.

Neurophysiological Modalities

The purpose of this section is not to provide detailed technical and procedural descriptions but to outline the general indications (strengths) of the specific techniques and their limitations (weaknesses) in answering clinical questions. The section aims to give guidance about the various electrophysiological techniques and enables the correct technique to be chosen for the diagnostic assessment of a spinal disorder with an assumed or obvious neurological affection.

Electromyography

EMG is the modality of choice for the diagnosis of a peripheral nervous lesion

Electromyography (EMG) is one of the most frequently applied electrophysiological techniques in spinal disorders and the term "EMG" is often almost synonymously used when asking for electrophysiological testing. It is the modality of choice for identification of a lesion within the peripheral nervous system affecting the lower motoneuron at any level (from the alpha-motoneuron within the spinal cord down to the distal motor endplates located in the muscle).

Technique

Needle and surface EMG recordings should be distinguished. **Surface EMG recordings** (cup electrodes attached to the skin) are primarily used for kinesiological studies (when investigating to what extent a muscle is activated during a complex motor task, such as walking) (Fig. 2), while **needle EMG recordings** are used to search for lower motoneuron lesions. They are performed with bi- or monopolar needles that have to be inserted into the target muscle. The insertion induces some discomfort comparable to when taking blood. It is an invasive procedure and therefore the specific indications and contraindications (anticoagulation treatment) need to be acknowledged. The EMG records the electrical activity within a muscle and is applied in the resting and activated muscle (some cooperation from the patient is needed). Besides the proof of a neurogenic lesion, myogenic motor disorders (myopathy, myotonic and muscle dystrophic disorders) can also be diagnosed [19, 25, 29].

Indications

Signs of denervation in EMG are temporarily delayed while innervation patterns change immediately

In spinal disorders, EMG is the method of choice for the identification of damage within the **peripheral motor nerve fibers** (highest sensitivity). However, the delay between the time of the actual damage and the first signs of denervation (acute denervation potentials occur after a mean of 21 days) must be considered. Also the activation pattern (complete or reduced interference) assessed during voluntary activation (here the patient needs to cooperate and perform a voluntary activation) can be applied as soon as the very first few days after a lesion to disclose a pathological innervation. The performance of EMG in several muscles allows the specific localization of the nerve damage (somatotopic localization of a lesion) to be indicated and for the differentiation of acute, subacute and chronic axonal damage (denervation). EMG is also the method of choice for the demon-

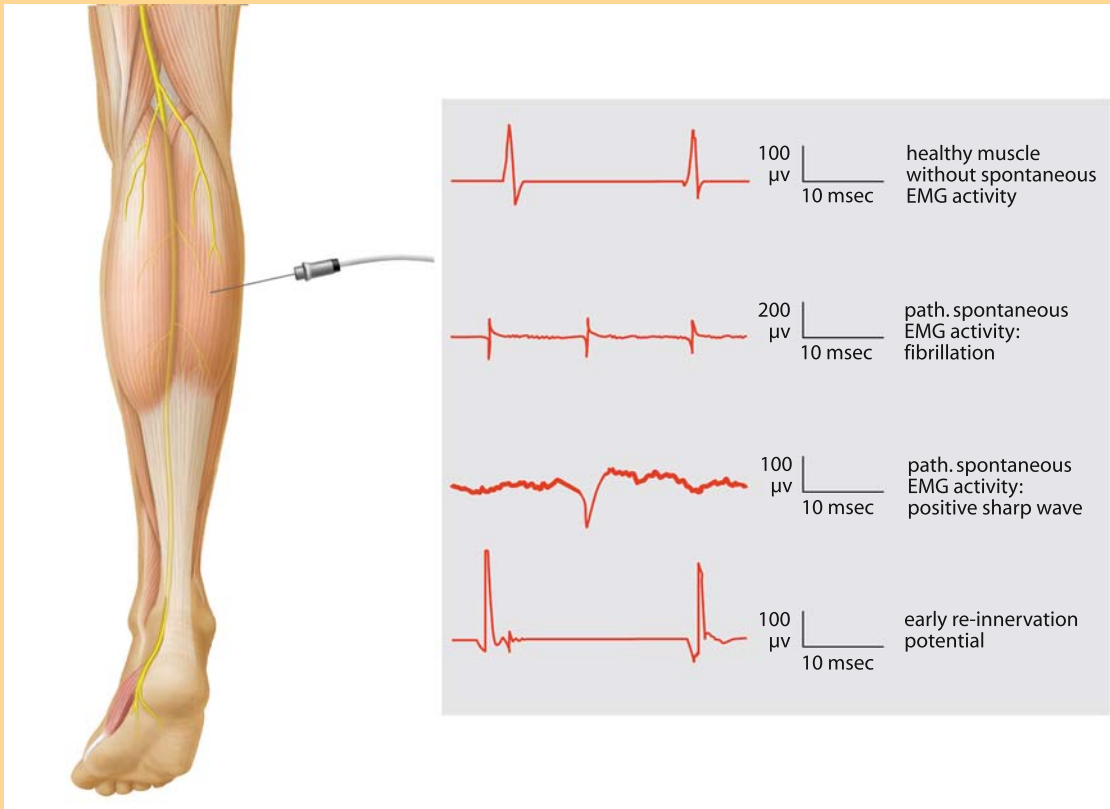

Figure 2. Electromyography

Spontaneous muscle activity is recorded at the target muscles.

stration of neurogenic reinnervation (subacute to chronic reinnervation pattern).

Limitations

The extent of axonal nerve damage and reinnervation is difficult to quantify

Spinal disorders with demyelination of motor nerve fibers (very slowly evolving neural compression as in benign tumor or stenosis) are less assessable by EMG. The extent of axonal nerve damage and reinnervation cannot be easily quantified by EMG. Needle EMG recordings provide some discomfort (which can be painful) for patients.

Nerve Conduction Studies

Motor and sensory nerve conduction studies (NCS) assess the **conduction velocity** (mainly properties provided by the myelination of peripheral nerves) and **amount of impulse transmission** (axonal transport capacity). These parameters distinguish between a primarily axonal and/or demyelinating neuropathy, which cannot be achieved by the clinical examination. Frequently NCS are combined with reflex recordings that provide additional information about changes in nerve conduction.

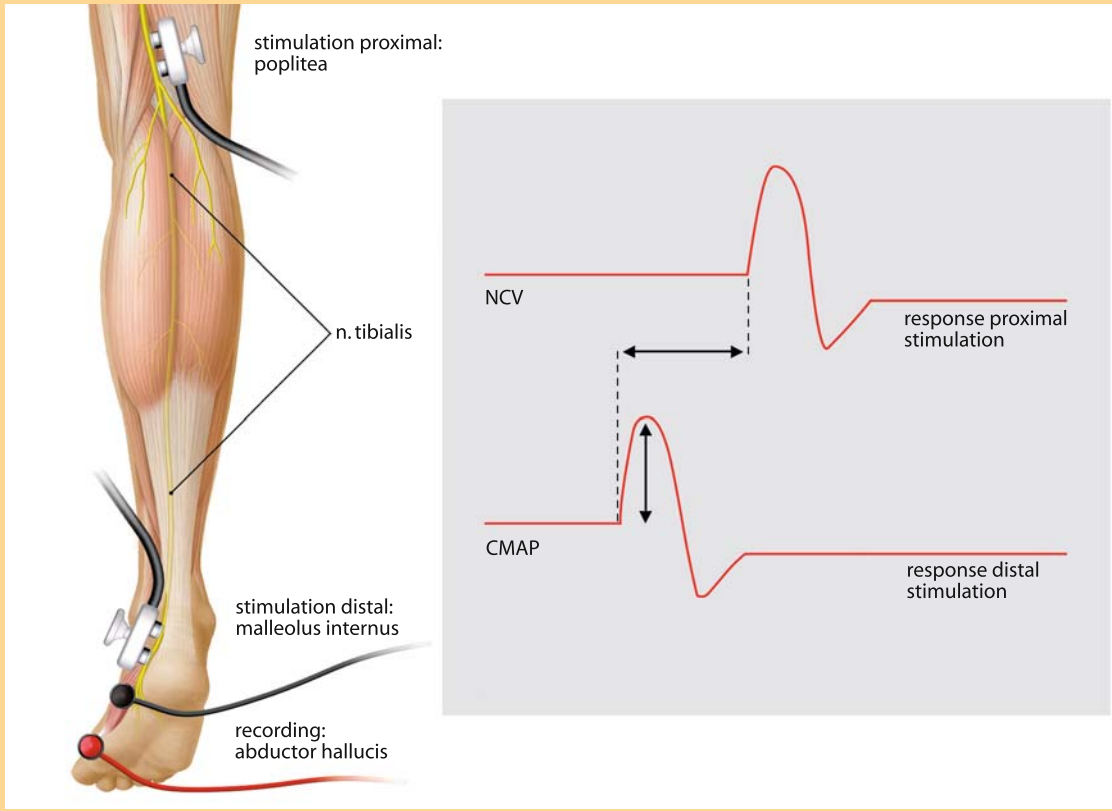

Figure 3. Nerve conduction studies

The nerve conduction velocity (NCV) is calculated dividing the distance between the stimulation points by the conduction time between these points.

Technique

Electrical stimulations (Fig. 3) applied along the peripheral nerve branch (distal to proximal) and recordings by surface electrodes at the distal motor or sensory site allow for the assessment of responses separately and for the calculation of **nerve conduction velocities** (expressed in meters per second) by measuring the distance [8, 20]. The **compound muscle action potential** (CMAP, in millivolts) and the sensory action potential (in microvolts) are calculated to assess the axonal nerve integrity.

Indications

Nerve conduction studies are primarily indicated in conditions assumed to affect the peripheral nerves (damage or disorders of the plexus, peripheral nerves, compartment syndromes, polyneuropathy), while they are not applicable for the diagnosis of a radiculopathy [34]. NCS are the method of choice for the diagnosis of a peripheral neuropathy (e.g., diabetic neuropathy) or nerve compression syndrome (carpal tunnel syndrome). They are very sensitive in demonstrating and **quantifying a conus medullaris and cauda equina lesion** (i.e., when combined with reflex recordings). However, isolated damage of S2–S5 roots can be missed. In **spinal cord injury** (SCI), intramedullary alpha-motoneuron damage induces a reduction of the CMAP of the related peripheral nerves, while the sensory NCS

NCS are indicated for the diagnosis of peripheral neuropathy but not radiculopathy

NCS are used to distinguish between axonal and demyelinating neuropathies

remains normal (a pattern which is able to exclude additional peripheral nerve injury). As sensory NCS in contrast to the motor NCS remain unaffected in spinal cord injuries, they enable the assessment of polyneuropathy in complete cauda and conus medullaris lesions.

Limitations

The characteristic signs of acute nerve damage appear with a delay of about 10 days after damage (however, this is earlier than signs of denervation in the EMG), and single recordings do not enable the acuteness of damage to be demonstrated. Here, the EMG recordings are able to distinguish between an acute and chronic course of nerve damage due to specific denervation potentials, which is not possible by NCS. Changes in NCS allow the **differentiation between primarily demyelinating and axonal neuropathies**, which are typically neuronal complications in medical disorders (e.g., neuropathy due to diabetes mellitus or uremia) but cannot be used to determine the underlying disorder.

F-Wave Recordings

F-wave recordings are not considered to be reflexes since only the motor branches of a peripheral nerve become involved. They are not mediated via a reflex arc where sensory and motor fibers are involved, like the tendon tap that induces an afferent input on the **spindle organ** (stretch of muscle) and an **excitation of motoneurons** in the spinal cord with an **efferent motor response** (the muscle jerk is the reflex response).

Technique

The electrical stimulation of a peripheral nerve induces a bidirectional electrical volley with a **direct motor response** (M-response of the orthodromic volley) (Fig. 4) and an antidromic volley propagating to the alpha-motoneuron, inducing an efferent motor response which travels back on the peripheral motor nerve fibers. This response is called the **F-wave**. The patient should be in a relaxed position without activation of the muscle.

Indications

F-wave recordings assess the alpha-motoneuron excitability and conduction velocity of the peripheral motor branch [10, 22]. The excitability of F-wave responses (expressed as a percentage of F-wave responses to 20 stimuli) can be applied to diagnose the level of spinal shock as they become abolished or reduced. They are sensitive to **demyelinating motor neuropathies** (e.g., diabetes mellitus) and complement NCS.

F-waves are sensitive to spinal cord excitability

Limitations

F-waves cannot assess the extent of intramedullary and peripheral axonal damage

F-waves are not sensitive enough to assess the extent of intramedullary and peripheral axonal nerve damage (no quantification of damage). The responses are not related to spasticity and are recordable only in some motor nerves (ulnar, median, tibial nerves).

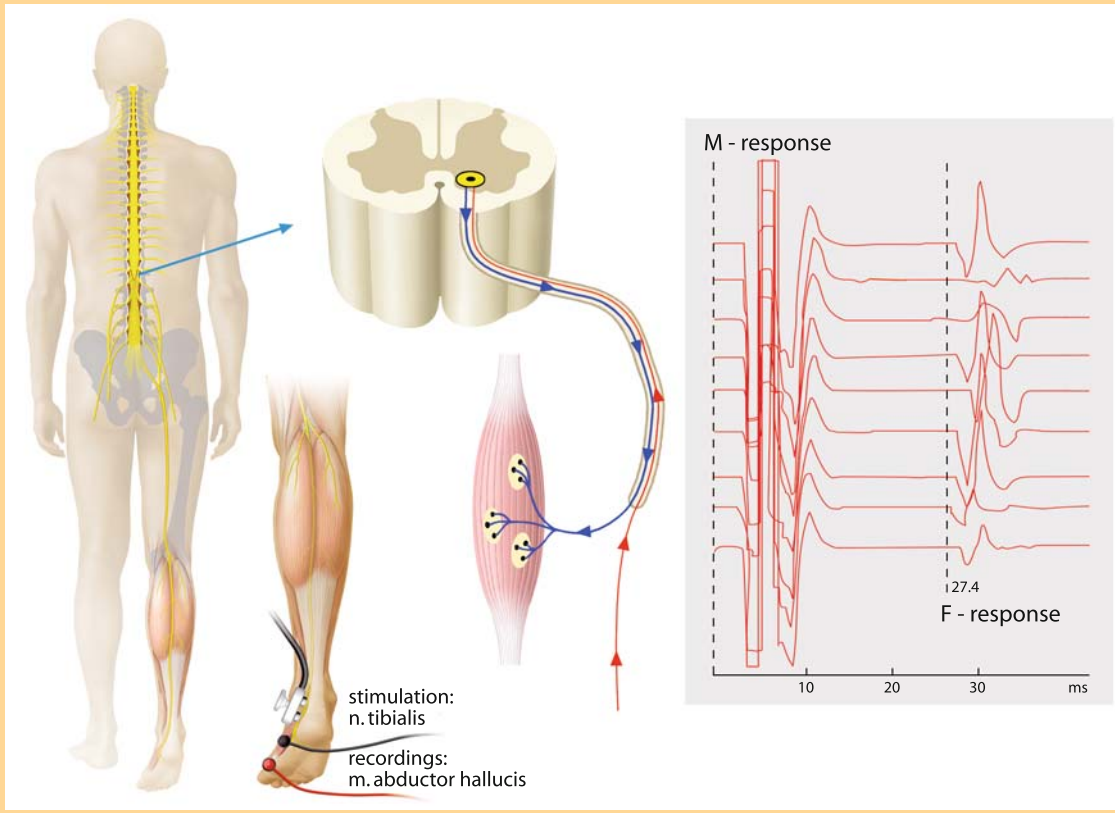

Figure 4. F-wave

The F-wave is elicited by antidromic excitation of motor axons and reflexion of this excitation at the motoneuron. The M-response is elicited by direct orthodromic excitation of the motor axon.

H-Reflex

The H-reflex recording is an electrophysiological investigation comparable to the tendon-tap reflexes. This **segmental reflex** is activated by an afferent sensory stimulus (electrical stimulation of the tibial nerve) and a monosynaptic transmission to the corresponding efferent motoneuron (Fig. 5) [6, 7].

Technique

By submaximal electrical stimulation of a nerve, sensory afferents induce a monosynaptically transmitted excitation of the corresponding alpha-motoneuron and an indirect motor response can be recorded by surface electrodes. The patient should be in a relaxed position without activation of the muscle.

Indications

The H-reflex provides information about sensorimotor interaction

The excitability and calculation of the tibial nerve H-reflex latency is a sensitive measure in **neuropathy** and for the assessment of disturbance within the **L5 – S1 nerve roots**. The H-reflex is less affected by spinal shock (it is reestablished within 24 h after SCI) than clinical reflexes and the F-wave.

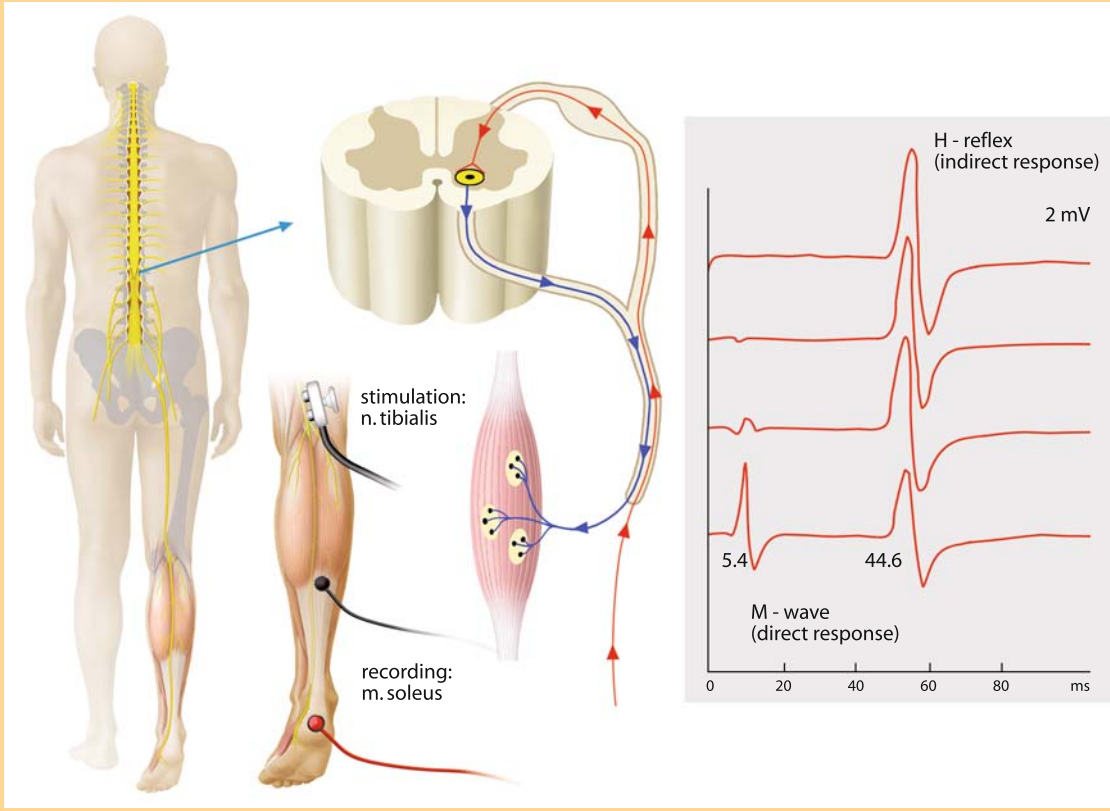

Figure 5. H-reflex

The H-reflex is elicited by excitation of low-threshold Ia-afferent nerve fibers which then excite the motoneuron monosynaptically (indirect response). The M-response is elicited by direct orthodromic excitation of the motor axon when using stronger stimulation intensity (indirect response).

Limitations

The H-reflex can only be recorded from n. tibialis

The H-reflex recording per se is not able to distinguish between sensory or motor nerve damage as the response is dependent on the whole reflex arc. It has to be acknowledged that the reflex response can be modulated by several conditioning maneuvers (**Jendrassik maneuver**) that are able to influence spinal excitability. Clinically reliable H-reflex recordings are **only achievable from the tibial nerves**.

Somatosensory Evoked Potentials

Somatosensory evoked potentials (SSEPs) enable the assessment of sensory nerve function across very long pathways through the body. By stimulation of distant body parts (distal peripheral nerves or dermatomes), nerve impulses are transmitted through parts of the peripheral and central nervous system and responses can be recorded at the cortical level. The additional recording of responses at different sites of the pathways (at the proximal segments of the peripheral nerve or the plexus, and even at different levels of the spinal cord) can be performed to localize the area or segment of the nerve affection. SSEPs do not represent one single type of sensory fiber but are most closely **related to vibration and proprioception**. These sensory qualities are propagated by the dorsal column within the spinal cord.

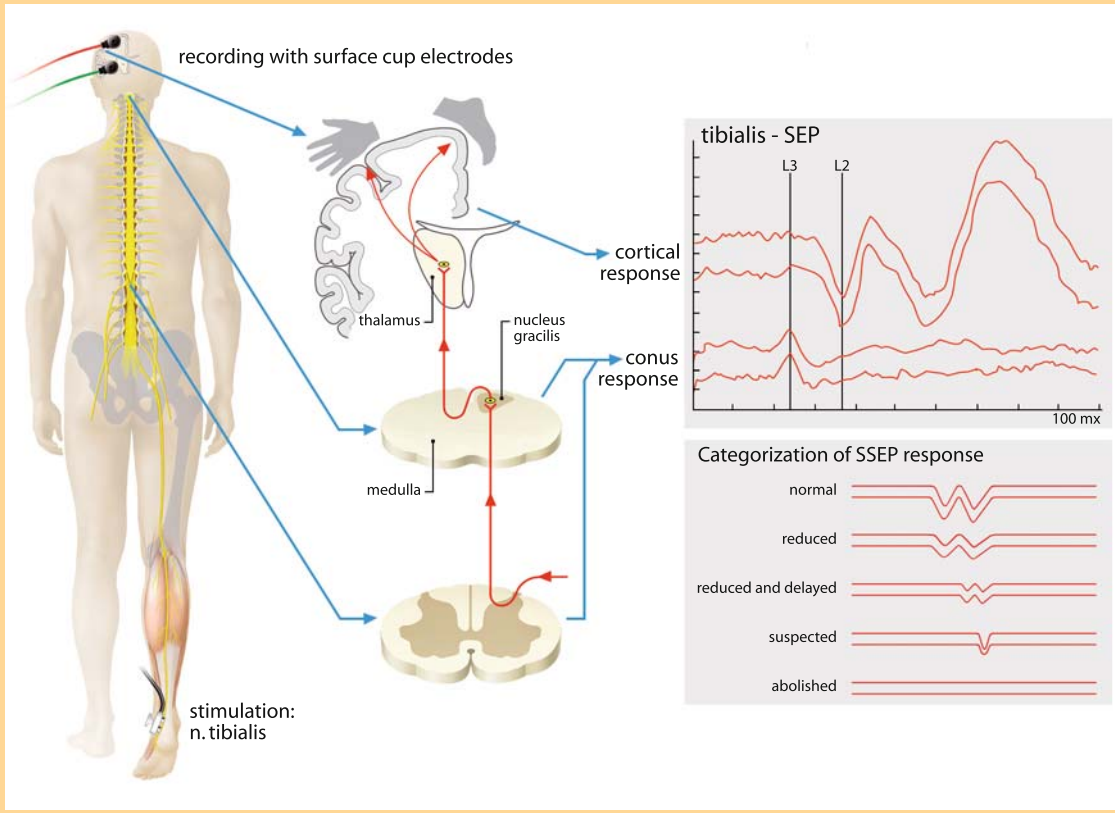

Figure 6. Somatosensory evoked potentials

SSEPs are elicted by peripheral stimulation of afferent nerves (e.g. n. tibialis, n. ulnaris) and recorded as stimulus-synchronized averaged brain activity.

Technique

SSEPs (Fig. 6) are cortical responses to repetitive electrical stimulations of peripheral nerves that can be recorded without the necessary cooperation of the patient (emergency, intraoperative) and can provide a survey of the sensory pathway from very distal to the cortical level [36, 37]. The recordings can be performed using **surface electrodes**, the electrical stimulations are below the level of painful sensation and the responses represent averages of 100 and more stimulations.

Indications

SSEPs assess damage of the dorsal column

Superior to clinical sensory testing, SSEPs provide objective measures (latencies and amplitudes) of dorsal column function and complement the subjective responses of patients to sensory testing. Especially in patients who are unable to cooperate sufficiently with difficult sensory tests or in whom due to a language barrier reliable clinical testing is not possible, SSEPs complement the clinical examination. **Repeated measures are valuable** for describing even minor changes within the sensory nerve fibers. In spinal disorders with nerve compression (spinal tumor or stenosis), even in clinically unsuspicious patients SSEPs can yield pathological findings. The responses are **only minimally influenced by medication.**

Limitations

SSEPs do not allow one to differentiate whether touch or pinprick sensation is affected

SSEP recordings are not sensitive enough to assess specific sensory deficits. They do not explicitly prove whether touch or pinprick sensation is affected, although the excitability of an SSEP response in a patient reporting complete sensory loss is proof that some sensory function is preserved. SSEP recordings do not relate specifically to pain syndromes, which are one of the leading clinical syndromes in spinal disorders.

Motor Evoked Potentials (Transcranial Magnetic Stimulation)

Motor evoked potentials (MEPs) comparable to SSEPs are able to assess the whole motor pathways from the cortical level down to the distal muscle and therefore are affected in **lesions of the peripheral** (peripheral nerve, plexus) and **central** (spinal, cortical) **nervous system.**

Technique

In awake subjects, **transcranial magnetic stimulation** (TMS) enables non-painful excitation of cortical motoneurons to induce MEPs transmitted by the corticospinal tract of the spinal cord and obtained from several muscles by surface electrodes (Fig. 7) [15, 18]. Patients are required to cooperate with the examina-

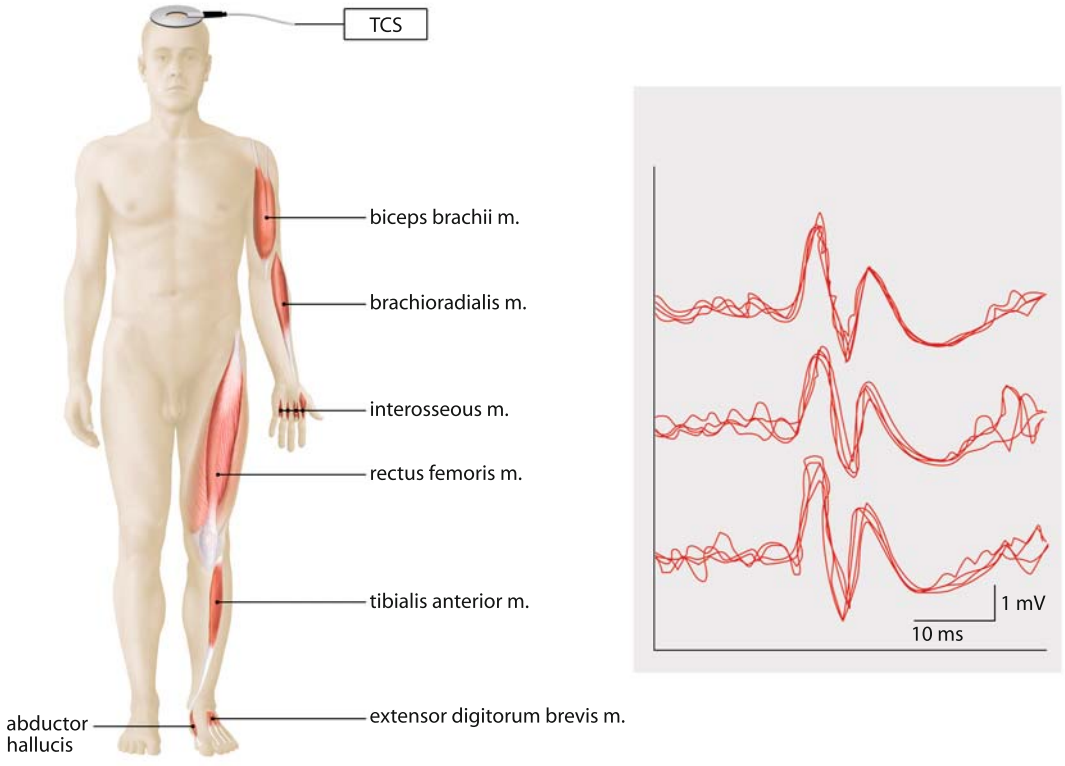

Figure 7. Motor evoked potentials

Transcranial magnetic stimulation at the skull level leads to excitation of motor cortical neurons which is conveyed to the spinal motoneurons. The excitation is recorded at the level of target muscles.

tion while they are asked to perform a small preactivation of the target muscle. Using the latter procedure, responses can be retrieved with a lower stimulation threshold and reliable latencies can be calculated to demonstrate delayed responses.

Indications

In addition to clinical motor testing (according to MRC grades), latencies and amplitudes can be obtained for an objective quantification of the conduction velocity and amount of response. MEP recordings are the method of choice for demonstrating subclinical affections of the corticospinal motor tracts that are less evident from clinical testing. The application of combined MEPs and motor NCS can be performed to distinguish between spinal and peripheral affection of the motor nerve fibers.

MEPs are the method of choice for assessing lesions of the corticospinal tract

Limitations

The results obtained are not directly related to the clinical motor strength, and MEP responses show a high variability of amplitude. **Patients need to cooperate** with the testing. In patients suffering from epilepsy or having intracranial ferromagnetic devices, TMS should be performed only with strict indications.

MEP responses are largely variable

Intraoperative Neuromonitoring

Intraoperative neuromonitoring is used for **real-time surveillance of nerve function** during spine surgery. Especially postsurgical neurological complications such as paralysis are mainly due to an impaired vascular supply of the spinal cord that cannot be controlled by the spine surgeon. Therefore, continuous monitoring of sensory and motor nerve function ensures that the surgical manipulations (suture of vessels or vascular compression due to stretching/correction of the spine) do not compromise the mandatory blood supply for the maintenance of nerve function. Especially in corrections of spinal deformities and during operations on spinal tumors, intraoperative neuromonitoring is able to improve surgical outcome.

Technique

In anesthetized patients, SSEPs and MEPs can be recorded to monitor spinal cord function during spine surgery [5, 21, 31]. Mainly needle electrodes (at the cortical level and muscles) are applied to ensure low impedance and reliable fixation during surgery. During anesthesia MEPs are routinely evoked by transcranial electrical (high voltage) stimulation with single or short train stimuli. While **SSEPs** are **averaged responses**, **MEPs** are retrieved as **single recordings**.

Indications

In spinal deformity surgery and in tumor surgery of the spine, intraoperative neuromonitoring of the spinal cord is a recommended procedure to provide a high level of safety for the patient and to give some guiding information to the surgeon. In spinal cord injury the relevance of neuromonitoring has not been established.

Neuromonitoring is indicated in surgery with potential spinal cord compromise

Limitations

The performance of intraoperative neuromonitoring requires a commitment of time (preparation of the setting) along with special equipment and trained staff. It has been shown that surgical teams using neuromonitoring have reduced the rate of neurological complications by more than 50 % [32]. However, even with spinal neuromonitoring some neurological complications can occur.

Role of Neurophysiology in Specific Disorders

Given the complexity of neuronal functions within and close to the spine (spinal cord, radical nerve fibers, plexus, peripheral nerves), there is no single electrophysiological measurement capable of being applied for testing, and combined measures need to be used. The required combination should be determined by a neurophysiologist, and the spine specialist should know the potential strengths and weaknesses of the different neurophysiological assessments.

Spinal Cord Injury

In traumatic disorders of the spine, neurological deficits are primarily examined according to the ASIA protocol, which allows for standardized assessment of sensorimotor deficit by describing the level and completeness of the SCI [17]. In patients not able to cooperate with a full clinical assessment, neurophysiological recordings can overcome this limitation and provide additional quantitative measures about spinal cord function.

Strengths

Neurophysiological studies allow neuronal damage to be objectified

Complementary to the clinical examination, **neurophysiological recordings**:

- objectify the neuronal damage (mainly independently of patient contribution) [11, 16, 27]
- describe the extent of spinal cord dysfunction in a superior manner to neuroimaging
- improve diagnosis and prognosis for treatment and rehabilitation [12]
- monitor the input of clinical treatment to the neural structures [13]

Weaknesses

The performance of neurophysiological recordings requires time and therefore needs to be carefully integrated into the clinical diagnosis and therapeutic procedures. There is also the need for specialized staff and equipment.

Cervical/Lumbar Radiculopathy

Neurophysiological studies allow radiculopathy to be differentiated from peripheral neuropathy

Radiculopathy due to disc protrusion is the most frequent spinal disorder and can be clinically diagnosed in cases with typical presentation without any additional neurophysiological recordings. However, in less typical cases or in the presence of additional accompanying neurological and medical disorders, EMG recordings are the method of choice for objectifying a radiculopathy of the motor nerve fibers.

Strengths

EMG recordings can be applied at all levels of radiculopathy. Using the needle EMG examination, the corresponding radicular muscles can be investigated:

- to objectify a motor radiculopathy
- to examine distal (extremities) or proximal (paraspinal) EMGs
- to exclude neuropathies that can mimic comparable pain syndromes (plexopathy)
- to reveal signs of reinnervation

Weaknesses

The following shortcomings of EMG recordings have to be acknowledged:

Neurophysiological studies are not applicable in anticoagulated patients

- EMG is not capable of documenting a pure sensory radiculopathy
- A normal EMG does not exclude a nerve compromise (i.e., severe pain in a radiculopathy) that has not yet induced motor nerve damage
- EMG is not applicable in anticoagulated patients

Cervical Myelopathy

Cervical myelopathy mainly is combined nerve damage within the spinal cord including: (1) affection of longitudinal pathways (dorsal column and corticospinal motor tract), and (2) segmental damage of the gray matter (alpha-motoneuron lesion). Predominantly patients complain about numbness of fingers, hands and feet, as well as unspecific difficulties in walking. These complaints can be easily misinterpreted as a neuropathic disorder.

Strengths

Combined neurophysiological recordings provide the opportunity to objectify and quantify a neuronal compromise at the cervical level and:

Neurophysiological studies allow myelopathy and neuropathy to be differentiated

- distinguish between focal demyelination of longitudinal pathways (MEP, SSEP) and gray matter damage (CMAP, EMG) [30, 33]
- confirm that a stenotic area with or without an intramedullary signal change can be related to the presented neurological deficit
- exclude that in mainly elderly people neuropathies become misdiagnosed

Weaknesses

Comparable to the poor correlation of radiological findings (extent and type of spinal canal stenosis) to clinical complaints:

- electrophysiological findings do not show a strong correlation with the extent of clinical complaints
- the specificity of neurophysiological recordings is reduced in combined spinal and peripheral nerve disorders

Lumbar Spinal Canal Stenosis

In typical clinical cases, the diagnosis of a neurogenic claudication is based on a combined clinical and radiological (CT, MRI) examination. With the increase in the elderly population and due to the improved techniques for identifying lumbar spinal canal stenosis, the extent of surgery performed due to neurogenic claudication has dramatically increased in the last 20 years.

Strengths

The combination of radiological, clinical and neurophysiological testing is improving diagnostic sensitivity and specificity. In atypical presentation of the disorder or in patients with other accompanying diseases:

- the affection of nerve function at the stenotic area can be disclosed and quantified [2, 4]
- neuropathies can be excluded that can induce similar pain syndromes (numbness of feet due to peripheral neuropathy) [1, 26]

Weaknesses

Comparable to cervical stenosis there is only a low correlation of the radiological findings (extent and type of spinal canal stenosis) to the clinical complaints

- electrophysiological findings are not correlated to the extent of clinical complaints
- in combined spinal and peripheral nerve disorders the specificity of the neurophysiological recordings is reduced

Neurophysiology in Differential Diagnosis

Not only in the population of elderly patients do several differential diagnoses have to be considered but especially when the complaints are demonstrated in an atypical presentation.

Peripheral Nerve Lesion Versus Radiculopathy

Neurophysiological studies allow radiculopathy to be differentiated from peripheral neuropathy

Damage to the nerve roots presents in a radicular distribution (see Chapters 8, 11) of sensory (dermatome) and motor (myotome) deficits, and electrophysiological measurements are able to distinguish a peripheral nerve affection from a radiculopathy. A **peripheral nerve lesion**, like the compression of the peroneal nerve close to the fibula head, induces pathological findings in NCS (conduction failure with reduced or even abolished CMAP) and pathological EMG findings in the distal muscles innervated by the peroneal nerve; while a complete motor L5 radiculopathy shows no NCS pathology but produces pathological EMG findings (signs of denervation) in both the distal (anterior tibial muscle) and the proximal (gluteus medius, paravertebral muscles) L5 innervated muscles.

Neuropathy Versus Spinal Canal Stenosis

Neurophysiological studies allow the exclusion of additional peripheral neuropathy

A polyneuropathy can mimic complaints similar to spinal canal stenosis (both lumbar and cervical) with numbness and some weakness mainly in the lower limbs. Also numbness of the fingers can be due to PNP, cervical myelopathy or carpal tunnel syndrome. Atypically presented complaints should indicate that combined SSEP and NCS recordings be performed, which are able to distinguish between these disorders. In spinal canal stenosis the peripheral nerve conduction velocity of the related nerves remains normal while the SSEP recordings become delayed due to a slowing within the spinal cord.

Neuropathy

Four major forms of neuropathy can be distinguished:

- sensorimotor neuropathy
- autonomic neuropathy
- mononeuropathy
- polyneuropathy

The most common form is diabetic peripheral neuropathy, which mainly affects the feet and legs. Neuropathic pain is common in cancer as a direct result of the cancer in peripheral nerves (e.g., compression by a tumor), as a side effect of many chemotherapy drugs, and renal disorders. Neuropathy often results in numbness, and abnormal sensations called dysesthesia and allodynia that occur either spontaneously or in reaction to external stimuli. Neuropathic pain is usually perceived as a steady burning and/or "pins and needles" and/or "electric shock" sensations.

Nerve entrapment syndromes are mononeuropathies which usually affect middle-aged and elderly patients. In patients suffering from atypical pain syndromes of the upper limbs, carpal tunnel syndrome (CTS) should be excluded. A thoracic outlet syndrome (TOS) and peripheral nerve compression at the elbow or the loge de Guyon can confuse the clinical diagnosis. While typical representations of these entrapment syndromes do not cause any particular clinical problems in diagnosis, atypical cases can be challenging. Nerve conduction studies are the method of choice for objectifying a nerve entrapment and are able to identify the localization of nerve compression.

Myopathy and Myotonic Disorders

In patients with walking difficulties and pain and fatigue after walking short distances, muscle disorders also have to be considered. Myopathies are neuromuscular disorders in which the primary symptom is muscle weakness due to dysfunction of muscle fibers but frequently present symptoms of muscle cramps, stiffness, and spasm. **Congenital myopathies** (mitochondrial myopathies, myoglobinurias) and muscular dystrophies (progressive weakness in voluntary muscles, sometimes evident at birth) are distinguished from **acquired myopathies** (dermatomyositis, myositis ossificans, polymyositis, inclusion body myositis). Neuromyotonias are characterized by alternating episodes of twitching and stiffness, while the stiff-man syndrome presents episodes of rigidity and reflex spasms that can be life threatening. EMG recordings are most sensitive for identifying **myopathic disorders** and are complemented by blood and biopsy work-ups for the specification of the disorder.

Neurophysiological studies are sensitive in diagnosing myopathic disorders

Hereditary and Neurodegenerative Disease

Neurogenic spine deformities are frequently seen in juvenile **neuromuscular disorders** (hereditary sensorimotor neuropathies, e.g., Charcot-Marie-Tooth neuropathy, spinal muscle atrophy, hereditary myopathies), and electrodiagnostic assessments are mandatory when the underlying clinical disorder has not yet been identified. In adults, spinal deformities can develop due to **neurodegenerative diseases** [rarely in amyotrophic lateral sclerosis (ALS), atypical Parkinson's syndrome with trunk instability], and it is mandatory to define the pathology as this should have an impact on the surgical approach. In these disorders combined electrophysiological recordings are applied to assess alpha-motoneuron or peripheral nerve affections.

Neurophysiological studies are helpful in diagnosing neurodegenerative disorders

Recapitulation

Neurophysiological modalities. The techniques and standards of clinical neurophysiological methods provide the capability to assess different components of the **peripheral and central nervous systems**. Besides the well-known EMG, several recordings are available that address very specific questions. Therefore, it is important to consider that **combined electrodiagnostic recordings** have to be applied to evaluate the different neuronal structures and functions. As spinal disorders are actually on the borderline between central (spinal) and peripheral (radicular, conus cauda) neuronal elements, the neurophysiological assessments need to cover these areas. Neurophysiological assessments only **complement the clinical neurological examination** and are intended to provide information that is not or is less precisely retrievable by clinical testing. These assessments in general do **not aim to evaluate complex body functions**, like walking and hand function, but to **objectify the function of neuronal subcomponents** (conduction velocity of nerve fibers) that contributes to the major function, as well as to improve the somatotopic localization of nerve damage.

Specific spinal disorders. The **neurophysiological investigations** should be **specifically targeted** to the assumed or evident spine disorders to identify and quantify the neuronal damage. In disorders that compromise the spinal cord or radicular nerves but have not yet induced structural damage, the neurophysiological recordings will not indicate any suspected disorder although the patients can be suffering from severe pain. Vice versa, in patients with only minor clinical complaints the neurophysiological recordings can reveal already advanced neural damage. Therefore, the main goal for neurophysiological recordings is to **objectify** whether a **radiologically exposed pathological finding** is related to assumed neuronal damage or to prove the presence of a neuronal compromise although the radiological findings are unsuspicious. In patients suffering from complex and/or multiple disorders the neurophysiological recordings can give confidence about the relevance of a pathological finding.

Neurophysiology for differential diagnosis. The different neurophysiological recordings allow for the diagnosis of a huge variety of neuronal diseases that have to be considered in spinal disorders. As recording the evoked potentials (SSEPs, MEPs) allows for the assessment of spinal cord function, EMG and nerve conduction studies focus on the peripheral nervous system and distinguish between the affection of motor and sensory fibers. These techniques enable the localization of injury and the distinction to be made between primary demyelination and axonal damage. The recordings can be utilized for follow-up recordings to monitor both the progression and the recovery from an injury/disorder.

Key Articles

Merton PA, Morton MH (1980) Stimulation of the cerebral cortex in the intact human subject. Nature 285:227
Landmark paper introducing transcranial magnetic stimulation for the assessment of motor pathways of the central nervous system in the awake human subject.

Forbes HJ, Allan PW, Waller CS, Jones SJ, Edgar MA, Webb PJ, Ransford AO (1991) Spinal cord monitoring in scoliosis surgery. Experience in 1168 cases. J Bone Joint Surg (Br) 73B:487 – 91
First proof of the significance of intraoperative neuromonitoring in scoliosis surgery to reduce postoperative neurological deficits.

Owen JH, Sponseller PD, Szymanski J, Hurdle M (1995) Efficacy of multimodality spinal cord monitoring during surgery for neuromuscular scoliosis. Spine 20:1480 – 88
This study demonstrated the improvement of neuromonitoring by the application of combined recordings.

de Noordhout AM, Rapisarda G, Bogacz D, Gerard P, De Pasqua V, Pennisi G, Delawaide PJ (1999) Corticomotoneuronal synaptic connections in normal man: an electrophysiological study. Brain 122:1327 – 1340
This study showed that direct cortico-motoneuronal connections can be assessed by motor evoked potentials.

Jones KE, Lyons M, Bawa P, Lemon RN (1994) Recruitment order of motoneurons during functional tasks. Exp Brain Res 100(3):503 – 508
This paper showed the ability to assess different types of motoneurons in humans by the performance of specific motor tasks.

Yamada T (2000) Neuroanatomic substrates of lower extremity somatosensory evoked potentials. J Clin Neurophysiol 17(3):269 – 79
This paper summarizes the technical issues and the clinical indication of tibial SSEPs, as well as the pitfalls that have to be considered for the application in diagnostics of neurological and spine disorders.

Angel RW, Hofmann WW (1963) The H reflex in normal, spastic, and rigid subjects. Arch Neurol 9:591 – 6
Landmark paper introducing the H-reflex for clinical diagnostics.

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