Götz Penkert · Josef Böhm
Thomas Schelle

Focal Peripheral Neuropathies

Imaging, Neurological, and Neurosurgical Approaches

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Götz Penkert
Neurosurgical Department
Friederiken-Hospital
Hannover
Germany

Thomas Schelle
Neurological Department
Municipal Hospital Dessau
Dessau-Rosslau
Germany

Josef Böhm
Neurological Department
Freiberg Hospital
Freiberg
Germany

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Preface

Management of peripheral nerve irritation and compression syndromes needs interdisciplinary concepts to find the exact diagnosis and to choose the most suitable surgical procedure. During the last few years, imaging techniques have revolutionized our diagnostic capabilities. Of course, clinical and neurological evaluation remains principal in regard to diagnostic efforts. Meanwhile, however, neurological disciplines have increasingly applied high resolution ultrasound techniques to peripheral nerve structures and lesions. Therefore, it is one of the purposes of this book to focus on these new developments and to assess their special value when applied to each individual focal neuropathy.

Microsurgical techniques could decisively improve results of repair of traumatized peripheral nerves. Although the introduction of microsurgery started 50 years ago, it has taken several decades for all surgical disciplines to accept its value. In contrast to that, if microsurgery was merely understood in terms of intraneural surgery, its utilization on nerve entrapment syndromes would invariably remain an exception.

Nevertheless, younger surgeons who start dealing with peripheral nerve lesions need to first understand which degree of intraneural lesion and which kind of reactive tissue fibrosis can occur due to nerve compression or even damage. Their next step will then be to make an assessment of the nerve lesion

when confronted with a real case. At the same time, they must learn to handle a nerve structure as gently as possible in order to ensure reduction of neural damage and avoidance of new extra- or intraneural scarring. We apologize if we repeat knowledge and facts which have already been reported prior to this work.

A further purpose of this book is to help younger physicians focus increasingly on important differential-diagnostic questions in order to avoid unnecessary or even damaging surgery. There exists a surprisingly large field of degenerative, genetic, or inflammatory mono- or poly-neuropathies. Likely depending on individual lack of experience, the surgeon might easily suppose a false diagnosis, and consequently begin surgery which is really not indicated. We are optimistic that the new imaging techniques will help us more and more to discover all these focal neuropathies which should never be surgically treated.

We have to realize new tendencies in peripheral nerve surgery, especially 'key hole' approaches. More patients wish to get surgery resulting in a skin scar as small as possible. It will not be the purpose of this book to favor these special techniques. We expect them to be far more expensive than normal open surgical methods and we are aware that expensive and specialized tools are not available everywhere. Although criticism from specialized centers will possibly arise, everyone needs to realize that severe nerve irritation syndromes occur everywhere and normally do not require specialized treatment centers.

The latter argument gives us reason to somewhat emphasize the capacities of neurosonography imaging techniques in our presentation and – likely a subject of further criticism – to slightly neglect the equally important extensive development of MR imaging of nerves, which, until now, has been reserved for specialized centers.

Hannover, Germany Götz Penkert

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First of all, my greatest gratitude and respect goes to my colleagues, Dr. Josef Böhm, Freiberg, and Dr. Thomas Schelle, Dessau, who agreed to contribute to this work with their huge clinical experience in the field of clinical and technical diagnostic procedures of focal peripheral nerve lesions due to inflammatory, genetic, or degenerative origin, or due to mechanical irritation and compression. During the making of this book, the learning process about the immense progress in neurological diagnostic abilities has provoked quite an awakening within myself as a neurosurgeon. The reliability of an increasingly prompt response to differential-diagnostic questions has also been astonishing. Nevertheless, additional acknowledgment has to go to Dr. Henrich Kele, Hamburg, who has pioneered the development of nerve ultrasound sonography in our region. Peripheral neurology has unfortunately been neglected too much in many neurological departments, so that one needs to realize that the mentioned pioneers really gained special merit in practicing new diagnostic approaches to focal neuropathies. I am sure that their experience will immensely contribute to the success of this book. It will help younger physicians to improve the process of diagnostic assessment and to distinguish between cases needing medicamentous treatment combined with careful observation and cases needing releasing surgery.

My gratitude has to be extended to Springer and its staff, who expressed enough confidence to publish our work. Of course, without their help this book would not have been completed.

Special thanks go to my wife, Ortrud, who once more spent many hours writing and correcting the text.

Hannover, Germany GötzPenkert

Abbreviations

AB	Adductor brevis muscle
ABP	Abductor pollicis brevis muscle
ADC	Apparent diffusion coefficient
ADM	Abductor digiti minimi muscle
AIN	Anterior interosseus nerve
AL	Adductor longus mucle
ALS	Amyotrophic lateral sclerosis
AM	Adductor magnus muscle
ASIS	Anterior superior iliac spine
CEUS	Contrast enhanced ultrasound
CIDP	Chronic inflammatory demyelinating neuropathy
CMAP	Compound muscle action potential
CMT	Charcot-Marie-Tooth Disease
CRPS	Complex regional pain syndrome
CSA	Cross sectional area
CSF	Cerebrospinal fluid
CSS	Churg-Strauss syndrome
CT	Computed tomography
CTS	Carpal tunnel syndrome
DADS	Acquired demyelinating symmetrical neuropathy
DML	Distal motor latency
DTI	Diffusion tensor imaging
DWI	Diffusion-weighted MR imaging
EDB	Extensor digitorum brevis muscle
EDX	Electrodiagnostic examination

EIP	Extensor indicis proprius muscle
EMG	Electromyography
EO	External oblique muscle
FA	Fractional anisotropy
FCU	Flexor carpi ulnaris muscle
FDP 2	Flexor digitorum profundus muscle of the second digit
FOV	Field of view
FPL	Flexor pollicis longus muscle
GCS	Guyon's canal syndrome
HMSN	Hereditary motor-sensory neuropathy
HRUS	High-resolution ultrasound
IO	Internal oblique muscle
LFCN	Lateral femoral cutaneous nerve
MA	Magic angle artefact
MADSAM	Multifocal acquired demyelinating sensory and motor neuropathy
MCN	Musculocutaneous nerve
MIP	Maximum intensity projections
MMN	Multifocal motor neuropathies
MNCV	Motor nerve conduction velocities
MPR	Multiplanar reformats
MRI	Magnetic resonance imaging
MRN	Magnetic resonance neurography
MT	Magnetisation transfer imaging
MUAP	Motor unit action potential
NCS	Nerve conduction studies
PAN	Polyarteritis nodosa
PD(W)	Proton density weighted
PEC	Pectineus muscle
PIN	Posterior interosseus nerve
PMP	Peripheral myelin protein
PNS	Peripheral nerve stimulation
PQ	Pronator quadratus muscle

SNAP	Sensory nerve action potential
SNCV	Sensory nerve conduction velocity
SPACE	Single slab three-dimensional sequence
SPAIR	Spectral adiabatic inversion recovery
SPIR	Spectral presaturation with inversion recovery
sPNS	Subcutaneous peripheral nerve stimulation
SRN	Superficial radial nerve
STIR	Short-tau-inversion-recovery
TA	Transverse abdominal muscle
THI	Tissue harmonic imaging
TMS	Transcutaneous magnetic stimulation
TOS	Thoracic outlet syndrome
TR	Repetition time
TSE	Turbo spin echo
TTLD	Median-thenar to ulnar-thenar latency difference
TTS	Tarsal tunnel syndrome
UNE	Ulnar neuropathy at the elbow
WG	Wegener's granulomatosis

Contents

Chapter 1
Introduction

Götz Penkert

Revolutionary novelties were discovered during the 1960s when microsurgery was introduced in the large field of nerve traumatology and nerve repair. Inducing significantly improved results, this development is largely due to Hanno Millesi. We will continuously remain in his debt for his pioneering achievements. Focal neuropathies have, however, been neglected because of their relatively lesser importance and, in our opinion, because we did not expect surgical novelties in this field.

People today, in the so-called civilized countries, require "perfect body feeling". They will not accept any slight sensory impairment or pain, even if only occasional or perhaps disappearing during daily activity. It becomes more and more of a challenge to solve their troubles and to remove their symptoms. It is interesting that they will not tolerate reduced symptoms but demand complete release. Dealing with focal neuropathies therefore needs brain storming preoperatively, not afterwards.

Improved diagnosis standards are required as well as perfect results after surgery, when indicated. A final diagnosis not allowing surgical treatment is disappointing but will be accepted. However, whenever surgery is carried out, it represents both a

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challenge and a hazard. Pitfalls quickly lead to litigation in our experience.

Therefore, we should try to define today's diagnostic standards in the field of focal neuropathies on the one hand, but on the other make evident that controversies concerning therapy do persist.

Chapter 2
Anatomy of the Peripheral Nerve System

Götz Penkert

2.1 Macroscopic Anatomy of the Peripheral Nerve System and Variations

Each physician who starts to deal with peripheral nerves – and particularly focal peripheral nerve pathologies – needs to be able to visualize in detail the macroscopic anatomical image of the surgical region; he or she has to be motivated to look up any remaining special questions in the anatomical atlas. We therefore avoid repetition of the complete and normal anatomy and subsequently restrict ourselves to the unexpected but important variations of nerve distribution in the human extremities. Special and clinically important anatomical details will of course be pointed out in Chaps. 8 and 9 when surgical aspects of focal nerve entrapments will be described separately.

The phylogenetic and ontogenetic development of humans explains how greatly nerve ramifications can vary; we must be aware of these variations, especially during surgical exploration, and diagnostic estimation and evaluation of imaging needs to be based on reliable knowledge covering their possible occurrence.

Rare cases exist with a brachial plexus consisting of only two trunks at supraclavicular level: the root C5 and C6 unite to the

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superior trunk, for instance, and roots C7, C8, and T1 unite to form a unique second trunk, the inferior trunk. A medial trunk is absent in these cases [1]. More frequently, C5, C6, and C7 unite to form one superior trunk, whereas C8 and T1 form the inferior trunk. Knowledge of these variations might be rather important in thoracic outlet surgery which will be described in detail in Sect. 8.2.1.

Likewise, variations of the ramification of the lateral cord into the musculocutaneous and median nerves are rare. This ramification may be displaced towards distal, so that the musculocutaneous nerve can arise from the median nerve at mid-upper arm level [2].

More frequently you will find ramification variants between the median and ulnar nerves at lower arm level: the well-known Martin-Gruber anastomosis consists of motor fibers which normally run downwards in the median nerve but leave this nerve at mid-forearm level and find a connection to the ulnar nerve. Sometimes these fibers use the anterior interosseous nerve as connecting structure. At a far distal level, these motor fibers have to rejoin nerve fibers which supply the thenar muscle. If this anastomosis arises from the deep branch of the ulnar nerve within the palm, it is referred to as Riche-Cannieu [3]. Knowledge of these anatomical variations becomes very important when a physician is confronted with a median nerve compression or pain syndrome presenting with sensory deficits but totally missing motor deficits. In particular, neurophysiological evaluations may occasionally be characterized by such special features. These cases and their evaluation need a correct assessment.

In the lower limbs, anatomical variations of nerve ramifications are less important. Comparable to the median and ulnar nerves of the forearm, the medial and lateral branches of the tibial nerve may vary concerning their level of ramification. As they run through a common canal (“tarsal tunnel”), both are involved when, for instance, they are compressed so that nerve variations become insignificant. The same holds true with the common

peroneal nerve which may be entrapped beside the head of the fibula. Varying levels of ramification of the sciatic into the tibial and peroneal nerves are nonrelevant for diagnostic or surgical procedures.

Small variations of the lateral femoral cutaneous nerve course have to be taken into account: the nerve may run through a slit between parts of the inguinal ligament but may also proceed across the bone of the iliac crest [4]. In the latter case, the small nerve is quite likely to become damaged during any kind of surgery within this region, such as bone removal for fusion surgery and endoscopic laparotomy, and also during nerve decompression surgery itself.

2.2 Microscopic Anatomy of the Peripheral Nerve System

Release of entrapped focal nerve segments rarely requires intraneural microsurgical techniques as will be demonstrated later in Sect. 7.1. The reason for this is that intraneural surgery always induces secondary tissue reactions. As a consequence, fascicle structures within the nerve trunk lose their ability to slide against each other. Pain symptoms would worsen. The goal of all kinds of entrapment surgery is to ensure pain relief for the patient instead. Therefore, focal entrapment surgery must usually remain restricted to the paraneural space.

In contrast to this surgical rule, the physician who carries out a diagnosis and who verifies it by means of imaging techniques should be aware of the special microscopic intraneural nerve pattern: changing diameter aspects regularly occur within a peripheral nerve course at its different levels. Nowadays, modern imaging is able to demonstrate this intraneural architecture – even signs of nerve scarring, inflammation, edema, or segmental compression. It can distinguish involved nerve

sectors from uninvolved ones. Because of these very revolutionary developments, we must clarify and perhaps repeat microanatomical facts during the following.

The axon membrane is surrounded by Schwann cells and their myelin sheath. These two components form the nerve fiber. Each nerve fiber is enclosed in the endoneurium; a bundle of these nerve fibers and their individual endoneurial sheath is again surrounded by a sheath of connective tissue, the perineurium. Furthermore, a bundle of these structures can be surrounded by connective tissue as a sheath and form a structure which is designated as a fascicle. Depending on the different levels, nerve architecture may be visibly characterized by an arrangement of three, four, five, or even more fascicles, the so-called group arrangement. The fascicles are separated from other fascicles by loose connective tissue with small vessels in between. The epineurium then surrounds the whole trunk [5]. The different fascicles within the epineurium have sliding capacities amongst each other, a fact that bears particular importance in the neighborhood of joints where inflecting and extending movements have to be tolerated for a complete life time. It is important not to neglect a loose connective tissue layer that surrounds the epineurium – the paraneurium – mentioned above as a place that demands restriction of entrapment surgery. It guarantees sliding capacities of the entire nerve trunk against the surroundings, even against bony structures, such as the back of the humerus, within the elbow sulcus, or beneath the tibiofibular joint (Figs. 2.1 and 2.2) [6].

The intraneural fascicle pattern regularly changes when we follow the nerve trunk into the periphery: Beside the spine, nerve roots and trunks consist of one or two thick fascicles. These are subdivided into special sectors by means of small membranes. More and more toward the periphery, the nerve architecture changes into the above-mentioned group arrangement of several fascicles. The subdivision to group arrangement

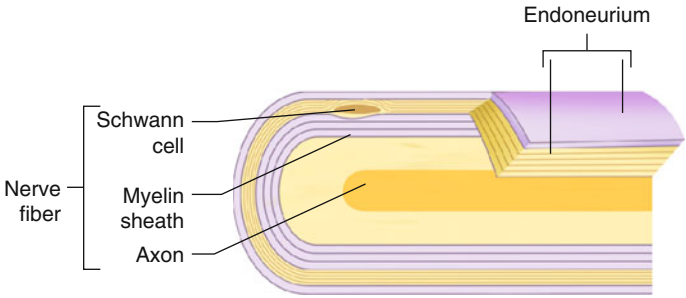

Fig. 2.1 Components of a nerve fiber surrounded by endoneurium

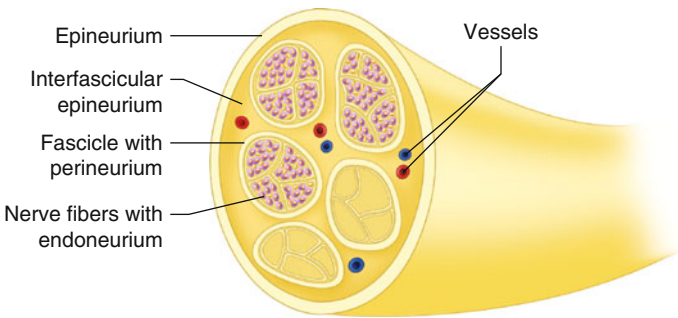

Fig. 2.2 Nerve trunk with five fascicles in group arrangement

achieves more of a multi-fascicular pattern at the periphery. In summary, the nerve starts with mono-fascicular architecture, it then subdivides into a group arrangement, and at the end it often demonstrates a multi-fascicular arrangement. Still at this peripheral level, the different fascicle groups also carry individual functions, whereas the special group arrangement is not equivalent to a special functional arrangement at a more proximal

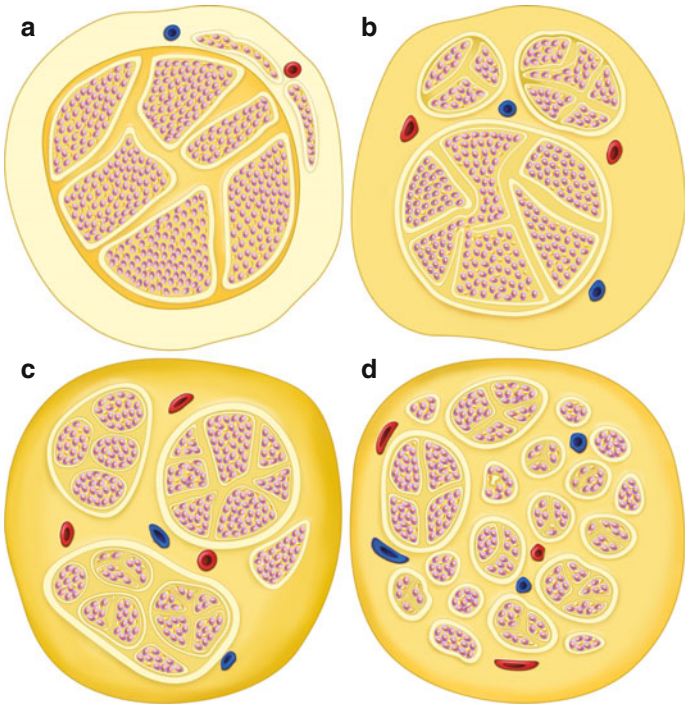

Fig. 2.3 Variation of the nerve pattern from proximal to peripheral levels: (**a**) monofascicular structure, (**b**) oligofascicular pattern, (**c**) group arrangement, (**d**) multifascicular structure – group arrangement lost

level. We can therefore understand why extremity nerves affected at high levels produce diffuse symptoms, whereas nerve lesions at the peripheral level can present with circumscriptive deficits. This fact also explains why neurophysiological evaluation is more difficult and less significant the more central the nerve lesion is located (Fig. 2.3) [7].

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Chapter 3
Pathophysiology of Nerve Lesions

Götz Penkert

3.1 Degrees of Nerve Lesion

The following chapter mainly considers trauma-related peripheral nerve lesions. It is of low importance in explaining nerve irritation or entrapment; as already mentioned, it should not influence the surgeon too much. However, we are quite frequently confronted with patient histories in which slightly damaging forces have influenced vulnerable nerve structures more as expected. We are consequently asked to find the origin and reason for this, in addition to offering explanations and a solution. The fact is that neurophysiology, which evaluates pathological nerve action velocity, needs a nerve damage as prerequisite. In Chaps. 5 and 6, we will go into detail and demonstrate how special kinds of slight focal nerve damage can still be related to far previously developed concepts mentioned below. Thus, the diagnostic procedure, which measures electric excitability and images the nerve pattern, has to refer to these potentially substantial nerve alterations.

In 1943, Seddon established the well-known concept of three nerve lesion degrees [1]:

Grade I: Neurapraxia/focal damage of the myelin sheath
Grade II: Axonotmesis/interruption of axon continuity

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Grade III: Neurotmesis/additional interruption of nerve connective tissue continuity

Theoretically, neurapraxia offers excellent chances of regeneration; a relatively quick reformation of the myelin sheath can be expected within a few weeks following the relief from compression. In the case of axonotmesis, the regeneration will take much longer; this works via axonal regrowth into the periphery and depends on the level of lesion and sprouting distance, assuming that all damaging forces are removed. Neurotmesis, of course, remains without any chance of recovery.

In 1951, Sunderland distinguished five degrees of nerve lesion [2]:

Grade I: As the myelin sheath is the most sensitive structure of the nerve fiber, he argued that focal demyelization occurs first/identical to Seddon's idea of neurapraxia.

Grade II: As result of higher compressing forces, he deduced, the axons undergo Wallerian degeneration and lose their continuity. The process of re-sprouting takes place within preserved endoneurial surroundings so that each fiber regains its former target – nearby a restitutio ad integrum can be expected.

Grade III: Additional endoneurial structure damage occurs in this grade, so that mis-sprouting starts when axons re-grow. This situation definitively leads to a functional deficit that the patient later has to tolerate.

Grade IV: Still higher damage to connective tissue structures significantly increases the amount of mis-sprouting on the one hand, or it may even result in blocking the axon sprouts completely at the site of lesion on the other. Despite preservation of nerve continuity, a complete loss of all regeneration capacities then happens, a damage, which subsequently leads to a permanent loss for the patient.

Grade V: Identical to neurotmesis/outward visible interruption of nerve continuity.

Due to Sunderland's sub-estimation of Seddon's idea of axonotmesis, we are enabled to offer a more reasonable explanation as to why spontaneous recovery cannot always be expected despite of the achievement of a careful nerve release by the surgeon. Secondly, we will better understand imaging results in the future.

3.2 Types of Secondary Scar Tissue Reaction

In 1992, Millesi established – in addition to Sunderland – a classification of reactive nerve fibrosis types, which is related to severely traumatized nerves [3]. However, advanced compression syndromes, for instance long lasting ulnar nerve neuropathies at elbow level, frequently do not recover or even lead to severe pain. Patients now require an explanation. The reaction of the connective tissue surrounding nerve fiber, fascicle, and fascicle groups is designated in these cases as fibrosis. It may not only result from sudden nerve trauma but also from chronic nerve compression, particularly in sensitive and exposed locations such as beside the elbow. Surgeons have already frequently exposed such fibrotic nerve segments during relief. They designated them as pseudoneuroma, – 'pseudo' because axon continuity is internally preserved. Its fibrosis is, firstly, irreversible and, secondly, it destroys the important sliding capacities of fascicles and fascicle groups against each other. It seems to be reasonable that, following such tissue reactions, axonal function can subsequently deteriorate further, in which case nerve decompression may take place too late [4].

Today's nerve imaging is able to evaluate the amount of intraneural fibrosis and thus estimate recovery chances. As we

try to demonstrate later, such intraneural images can influence the surgeon in the future, in particular concerning cases of chronic ulnar nerve and tarsal tunnel compression. In 1992, Millesi distinguished between a reactive fibrosis of the

- epineurium (Type A),
- interfascicular epineurium and perineurium (Type B), or
- endoneurium (Type C).

Type C lesions have the worst prognosis. The fact that ultrasound imaging and multi-resonance imaging are nowadays confirming the correctness of his classification is of interest.

The myelin sheath and axons principally need release of any compressing forces. In continuity, traumatized nerves then need different steps of micro-surgical neurolysis. These techniques will be discussed later in Sect. 7.1 where we mainly focus on traumatized nerves [5]. In contrast to trauma treatment, micro-surgery – when applied to focal nerve entrapments – can nevertheless be dangerous, although a nerve pseudo-neuroma can stimulate the surgeon to try his best underneath the microscope. Possible reasons and explanations are as follows:

1. Micro-surgical fascicle separation destroys individual fiber connections between fascicles or fascicle groups (it damages the plexiforme nerve structure according to Sunderland). Innumerable painful mini-neuromas between the fascicles will occur.
2. Micro-surgery leaves behind connective tissue reactions – the above-mentioned fibrosis to some extent – depending on the surgical trauma. After nerve injury, neurolysis aims at reducing reactive scarring to a steady state of lesser and final fibrosis to offer garantee of functional improvement. Instead, after simple focal nerve release a fibrosis should not occur at all; it is our experience that such fibrosis can be a source of increasing pain.

Thus, the following unsolved questions concerning pre- and postoperative pain remain [6]:

What is pain?
Which structure within the nerve generates pain?
Why do some entrapments result in severe pain while others do not?
Why does nerve trauma frequently generate less disturbing pain?
Why does surgery of traumatized nerves rarely increase the pain level?
Why is entrapment surgery comparatively much more sensitive in terms of postoperative pain?

We must finally state that the three classifications of nerve lesions do not provide sufficient explanation of these sensitive and complex pain phenomena related to focal nerve damage (see also Sect. 4.2).

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Chapter 4
Clinical Presentation of Focal Nerve Lesions

Götz Penkert

4.1 Diagnosis, Assessment, and Completion

Each focal neuropathy first needs a careful exploration of the patient's history and evaluation of his clinical symptoms. Technical investigations remain as an additional option, though being of increasing importance, as mentioned previously. This statement is necessary insofar as today's students and young physicians in neurologic departments tend to overrate modern imaging techniques as the primary diagnostic tool in every case. This holds true with regard to central nervous diseases as much as peripheral nerve lesions.

1. A list of questions to be asked in any case. First, there is a need to explore the patient's history in detail: the kind and time of onset of symptoms, sudden or gradual onset, with or without a recallable incident, with or without previous inflammatory disease, progress and velocity of progress, which symptom occurred at first, which came in later, which increased; also questions about pain, time of onset, eventual disappearance and sudden motor deficits, quality, intensity, and radiation of pain, as well as dependence on extremity or

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hand-movement should be included; in this context, questions about habitual positioning of extremities during day or night may be useful in the search for information on pressure mechanisms.

2. Clinical examination. It should not be too time consuming to find the involved sensory area of the extremity: is it a dermatoma, autonomous nerve distribution field, or something in between – a question that allows one to distinguish between more centrally or more peripherally located lesions; the investigation of numb areas always needs to be compared with the contralateral extremity.

   Next, a question about motor deficits will arise – are they referable to a root, nerve trunk, or a nerve branch; in the latter case an exact knowledge of its function and the site of its branch-off from the main trunk is required. Findings, in which a restriction of motor deficits on one single nerve distribution is lacking, are of special interest. This would indicate a lesion at a higher level or, quite another problem, an immunological-inflammatory disease. All the described differential-diagnostic disorders will then be discussed in Chaps. 5, 6, and 10.

   Therefore, the existence of degenerative peripheral nerve diseases in a patient's previous history is never to be neglected because it can simulate, conceal, or overlap a compression neuropathy, and it will also negatively influence the prognosis of nerve recovery.

3. Further clinical examination has to aim at a clear designation of which nerve is actually involved. Palpation and percussion of the presumably affected nerve trunk will help us: at different points of the suspected nerve, percussion will frequently trigger a typical electric current-like pain that the patient experiences in the sensory area associated with the nerve's distribution field. The point where this triggered pain reaches a maximum can lead us to the location of the focal nerve involvement. Our opinion of this TINEL sign is that several

nerve axons, after interruption of continuity, develop this irritability at their ends when re-sprouting begins. This represents a slight phantom pain elicited by axons in discontinuity. Its existence does not indicate the quantity of affected nerve fibers or the prognosis; but it does indicate axon involvement and its location.

Because of their outstanding importance, particularly concerning focal neuropathies including their assessment and therapy, we are going to dedicate associated pain types to the next chapter. In Chaps. 5 and 6, the large field of technical tools in diagnostics will follow – including neurography, electro-myography, and nerve ultra-sound sonography –which is essential as an additional guide towards a final diagnosis.

4.2 Pain Related Focal Neuropathies

Focal neuropathies associated with more or less severe pain need additional questioning in order to estimate the pain quality. When dealing with patients who suffer from nerve-related pain over months or even years, the physician will recognize their difficulties in describing their pain in their own words. Usually they start with how irritated they feel, how many daily activities are disturbed, how unbearable it feels to touch the painful skin area, or how pain-inducing a change of temperature is. Several patients say that they can demonstrate an especially hypo- or hypersensitive tissue area which they then suppose to be connected with the source of their pain. It is therefore of great importance to put carefully directed questions to make a correct pain assessment.

Entrapment syndromes of limb nerves generate a more or less bearable pain. It remains unexplained that the pain often does not remain restricted to the autonomous distribution area of the affected nerve; rather it appears diffuse and it usually disappears after nerve decompression. The carpal tunnel syndrome

could be the best example, widely known, and associated with diffuse pain and immediate pain disappearance after surgical relief. This type of nerve trunk-related pain does not fit in with our nerve lesion classifications of the preceding chapter on the one hand, but also not with the concept of so-called nociceptive generated pain on the other. We only know that an intraneural and substantial nerve lesion predominantly does not exist in these cases. In addition, as mentioned before, we have no reliable knowledge about the substances or the structures which generate this type of pain. We only know that surgery has to be as gentle as possible to guarantee avoiding any substantial damage. If possible, the surgeon has to preserve paraneural and intraneural sliding capacities.

Quite another type of pain will be generated if a substantial intraneural focal nerve lesion has been left behind. The location that activates these types of pain is not at all times the site of the damaged nerve segment. Four different types of pain may then appear [1]:

1. Neuroma pain, a normally non-existent pain which however occurs when the involved focal nerve segment is triggered by palpation or percussion. This maneuver thereby elicits an electric current-like pain which the patient localizes in the distribution area of the affected nerve.
2. Hyperpathia, a normally non-existent pain which is, however, of unbearable intensity when the skin in the distribution area of the affected nerve is repeatedly slightly stimulated.
3. Allodynia, a pain which is permanently present as a burning sensation; innocuous stimulation can also increase the pain level.
4. Causalgia, a permanent existing burning no longer restricted to the autonomous distribution area of the affected nerve, but overlapping into neighboring skin areas. This pain type is termed CRPS II (complex regional pain syndrome, sympathetic dystrophy) when sympathetic deregulations also occur [2].

Pain types 2–4 are designated as “neuropathic pain.” Neuroma pain is generated in the periphery, appearing about 4–6 weeks

after a focal nerve injury, and achieving a steady state weeks later. In contrast to that, neuropathic pain is finally generated in the central nervous system, and it has a rather sudden onset, appears from one to a few days after the nerve injury, and achieves a steady state immediately. Onset, time course, and intensity of neuropathic pain differ greatly from neuroma pain.

It is therefore reasonable to evaluate all the details of the patient's anamnesis to obtain the correct impression of the individual type of pain. Difficulties can arise at certain locations when a causalgia is presented with an enlarged over-sensitive skin area. The experience is, for instance, that it is frequently barely possible to determine which of the inguinal nerves is really affected after hernia surgery.

According to animal models and clinical studies, the following pathophysiological mechanisms presumably contribute to the central-nervously generated pain types:

1. Continuous abnormal excitation of afferent fibers due to nerve fiber compression [3]
2. Impaired intraneural microcirculation [4]
3. Abnormal excitatory coupling between sympathetic fibers and afferent nociceptive fibers, particularly within the spinal ganglion [5]
4. Disturbed axonal substance transport in both directions, resulting in a loss of control of the biochemical activity in the nerve cell body [6]
5. Transsynaptic electrophysiological activities from afferent neurons to higher-order neurons in the spinal cord and brain [7]

In previous decades, nerve surgeons were convinced that either microneurolysis or resection of the damaged nerve segment and repair by autologous grafts could solve intractable nerve pain. They believed that re-sprouting of sensory axons with achievement of their former targets could re-stabilize former nociceptive functions. The results were partially disappointing [8]. It is not the purpose of this book to go into further detail,

but physicians who mainly deal with nerve-related pain today recognize these mechanisms proximal to the focal nerve damage as responsible for pain-persistence following any kind of microsurgery. In the case of a neuropathic pain type, they even warn of the risk of increased pain intensity from surgery to surgery [9]. It is therefore a great responsibility to assess sufficiently early which pain type is presented and to exclude peripheral surgery at the site of the focus when a neuropathic pain has occurred. Pain that is generated in the central nervous system cannot be removed by peripherically applied ablative methods. There is meanwhile a consensus between pain surgeons that only augmentative methods offer any help here [10]: the peripheral nerve stimulation technique (PNS) by electrodes and stimulator (synonym: neuromodulation) according to the 'gate control theory' by Melzack and Wall [11].

Quite another type, localization, and time course characterize the immunologically induced plexus neuritis (Parsonage – Turner Syndrome). Patients describe an brutal pain with sudden onset, localizing it mostly in the shoulder girdle and rarely in the upper or forearm region. They report persistence of the pain for about 2 or 3 days, then a quick disappearance and, at the same time, a motor deficit affecting the same extremity. We will describe diagnostic and clinical details in Chaps. 6 and 10. However, we should repeat once more how important pain anamnesis is and how the correlation between occurrence of pain, numbness, and paresis can guide one to a reliable differential diagnosis.

4.3 Conclusions

Painful focal neuropathies of inflammatory – autoimmunological, degenerative, or ischemic – origin do not need surgery but partially immune-suppressive or immune-stimulating medication. Painful focal neuropathies of mechanically irritating mechanisms need decompressing surgery. Focal neuropathies

with circumscribed nerve damage as the origin of pain need first a careful pain assessment and then surgical treatment adapted to the particular pain type. In the case of pain due to a pseudoneuroma, microneurolysis might be considered (see Sect. 7.1) instead of, as in the case of pain due to a neuroma in continuity, resection and nerve repair. Something like "peeling off" the neuroma as often written in medical reports is nonsense. In the case of one of the neuropathic pain types, peripheral nerve stimulation (PNS) is today the method of choice. Careful pain exploration and the ability to make adequate intraoperative decisions are indispensable.

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Chapter 5
Electrodiagnostic Testing of the Peripheral Nerves

Thomas Schelle

5.1 Introduction

Electrodiagnostic examination (EDX) consists of two major parts: nerve conduction studies (NCS), including long latency reflex testing (F-waves), and needle electromyography (EMG). In addition, somatosensory evoked potentials and magnetic stimulation motor evoked potentials are needed sometimes, but the two latter techniques are outside the scope of this chapter. NCS and EMG are usually performed together. In contrast to nerve imaging such as magnetic resonance imaging (MRI) and high-resolution ultrasound (HRUS) (see Chap. 6), EDX provides functional information about the electrical properties of the peripheral nerves. EDX should only be carried out by specially trained staff, such as by a certified physician, or under his or her direct supervision [1].

What does EDX do? It extends the clinical examination. It can differentiate between disorders of the peripheral nerves, the neuromuscular junction, and the muscle itself. In a peripheral nerve disease in particular, EDX provides useful data regarding the localization of focal involvement (e.g., anterior horn cell,

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nerve root, nerve plexus, peripheral nerve), the type (e.g., demyelinating or axonal), the severity (incomplete versus complete), the duration (subacute or chronic), and the prognosis (reinnervation or no reinnervation) [2, 3].

What is EDX not able to do? It cannot replace a careful anamnesis survey and clinical examination by a neurologist experienced in the field of peripheral nerve disorders [4, 5]. Furthermore, EDX is unable to depict the underlying pathological features of a peripheral nerve disease. For instance, a carpal tunnel syndrome is usually caused by compression of the median nerve owing to narrowness within the carpal tunnel, but it may also occur because of compression by a mass lesion such as a ganglion cyst, an intraneural hematoma, or a peripheral nerve tumor. In all these cases, EDX provides the same result. Only imaging techniques (see Chap. 6) are able to demonstrate the underlying pathological features in those cases [6].

In the next sections, we will focus on the basics of EDX in disorders of the peripheral nerve only. The scope of this book does not include diseases of the muscles and the neuromuscular junction. For a more detailed description, please refer to specialist literature [2, 3].

5.2 Types of Nerve Fibers Assessable with EDX

According to the classification of Erlanger and Gasser, there are three different types of nerve fibers: large myelinated A-alpha fibers, small myelinated A-delta fibers, and unmyelinated C fibers [7]. A very important fact is that conventional EDX can only assess the largest, fast-conducting, and well-myelinated A-alpha fibers of the motor, sensory, and "mixed" peripheral nerves, including efferent motor fibers and afferent sensory fibers mediating the sense of touch, position, and vibration.

Slow conducting A-delta and C fibers mediating pain, cold, and warm sensations are only measureable with special techniques, such as heart rate variability test or sympathetic skin response test, which are discussed elsewhere [2, 3]. This fact explains why some painful conditions, such as small fiber neuropathy, cannot be examined using conventional EDX [4, 8].

5.3 Motor Nerve Conduction Studies

5.3.1 Technique and Parameters

In a motor NCS, the peripheral nerve is stimulated with a transcutaneous depolarizing square wave electrical pulse (stimulation electrode) of very short duration. The stimulus generates a muscle contraction in the target muscle, and its intensity must be supramaximal in order to excite all A-alpha fibers. Furthermore, a ground electrode is also necessary, providing a zero-voltage reference point. With two self-adhesive surface recording electrodes that are placed on the muscle belly (pick-up electrode) and on the tendon of the target muscle (reference electrode), the compound muscle action potential (CMAP) arising from the activated muscle fibers is recorded. The stimulation can be carried out proximally to the recording electrodes at different sites of the nerve course (e.g., wrist, forearm, elbow, upper-arm, and Erb's point) [9].

In the case of deeply situated peripheral nerves, such as the lumbar plexus, stimulation is also possible with needle electrodes or, alternatively, by means of transcutaneous magnetic stimulation (TMS). This also applies to deeper lying muscles, where the CMAP can also be recorded with needle electrodes. However, these special techniques are discussed elsewhere [2, 3].

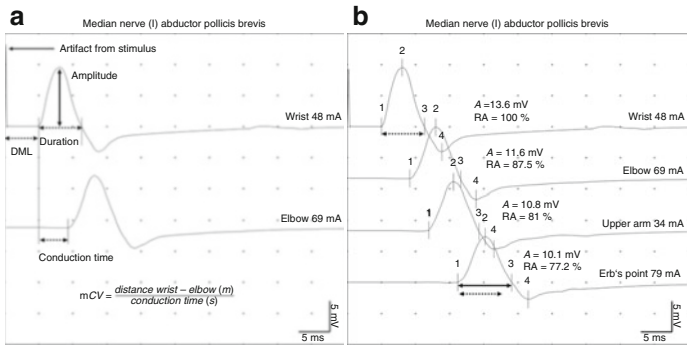


Fig. 5.1 (**a**) Important parameters derived from motor nerve conduction studies and calculation of the motor nerve conduction velocity. (**b**) Physiological "temporal dispersion." Note the amplitude decay (RA, relative amplitude) and the prolonged duration of the compound muscle action potential (CMAP) recorded after proximal stimulation compared with the CMAP obtained after distal stimulation

Neurophysiological convention specifies that negative voltage results in an upward deflection of the CMAP. Taking this fact into account, the routine technique described above allows a couple of parameters derived from the CMAP, that are used in the diagnosis of peripheral nerve diseases to be measured and calculated (Fig. 5.1a) [9–12]:

Distal motor latency (*DML*) is the time between stimulus onset and onset of the negative peak of the CMAP. It reflects the time required for conduction through the distal axons (fastest A-alpha fibers), the neuromuscular transmission time, and the time needed to generate a muscle action potential. It is measured in milliseconds.

CMAP Amplitude is measured from the baseline to the negative peak of the CMAP. It is determined by the number of activated muscle fibers. It is measured in millivolts.

Duration is defined as the time from the onset of the CMAP to the return to baseline at the end of the CMAP. It reflects the range of conduction velocities within the nerve. It is measured in milliseconds.

Conduction time is measured in milliseconds on the basis of the difference between the motor latencies after proximal and distal stimulation.

Motor nerve conduction velocity is calculated for every nerve segment after distal and proximal stimulation by determining the quotient of the distance between the two stimulation points over the conduction time. It is measured in meters per second. The minimum distance between the two stimulation points should be at least 10 cm to avoid inaccuracy [9–12].

Note that the larger the distance between the stimulation and recording electrodes, the longer the duration of the CMAP owing to an increased range of conduction velocities. This phenomenon is additionally characterized by an increasing decay of the amplitude of the CMAP, also called *temporal dispersion* (Fig. 5.1b) [13, 14].

Age-matched normal values for NCS, including motor nerve conduction velocity, distal motor latency, and the CMAP amplitude of every single peripheral nerve are either derived from studies in the laboratories or obtained from related literature [2, 3, 9, 15].

There are many physiological factors affecting nerve conduction. Only a few are addressed here. Generally, nerves of the upper extremity conduct faster than those of the lower extremity. Moreover, the nerve conduction velocity in proximal nerve segments is faster than in the distal parts of a peripheral nerve [16]. The most important factors influencing NCS in the clinical routine are age and skin temperature. In infants and patients of advanced age, nerve conduction velocities are diminished. Furthermore, a decrease in skin temperature also decreases the conduction velocities [17–19]. Therefore, standardization and

documentation of the skin temperature during electrodiagnostic testing is mandatory. In the case of low skin/room temperatures, a hot water bath or ceiling-mounted radiant heaters can be effective in raising the skin temperature of the limbs.

5.3.2 Demyelinating Disorders

Demyelinating disorders of the peripheral nerve result in impairment of conduction owing to the focal or generalized pathological features of the myelin sheath [11]. As a consequence, EDX demonstrates a slowing of the motor nerve conduction velocities and/or a prolongation of distal motor latencies. Depending on the type of nerve involvement, different patterns of motor nerve conduction velocity deterioration can be observed [2, 3, 9–12, 15]:

- Focal: affecting only one nerve segment (e.g., in the early stages of entrapment in ulnar neuropathy at the elbow). An example is shown in Fig. 5.3a.
- Multifocal: affecting certain nerve segments in one or more different peripheral nerves (e.g., multifocal motor neuropathy, acute and chronic inflammatory demyelinating neuropathy).
- Generalized: affecting all nerve segments in different nerves (e.g., hereditary sensorimotor neuropathy type 1).

If only the distal nerve segment is affected, prolongation of the distal motor latency will result, whereas nerve conduction velocity and CMAP amplitude are normal. This applies in particular to the early stages of carpal tunnel syndrome, tarsal tunnel syndrome, and Guyon's canal syndrome. In contrast to focal entrapments, a generalized demyelinating neuropathy leads to prolongation of the distal motor latencies in addition to a slowing of the nerve conduction velocities (Fig. 5.2). Distal motor latency is used for evaluation because

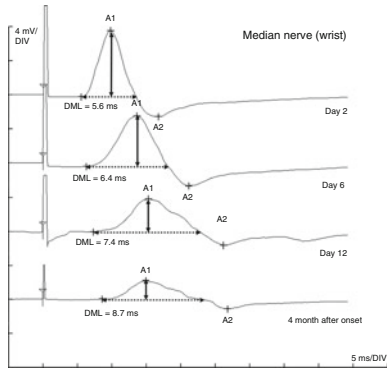


Fig. 5.2 Distal CMAP and distal motor latency (DML) of the median nerve in a patient with acute inflammatory demyelinating neuropathy 2, 6, 12 days, and 4 months after onset of the clinical symptoms. Note the prolonged DML due to the distal nerve segment being affected and pathological "temporal dispersion"

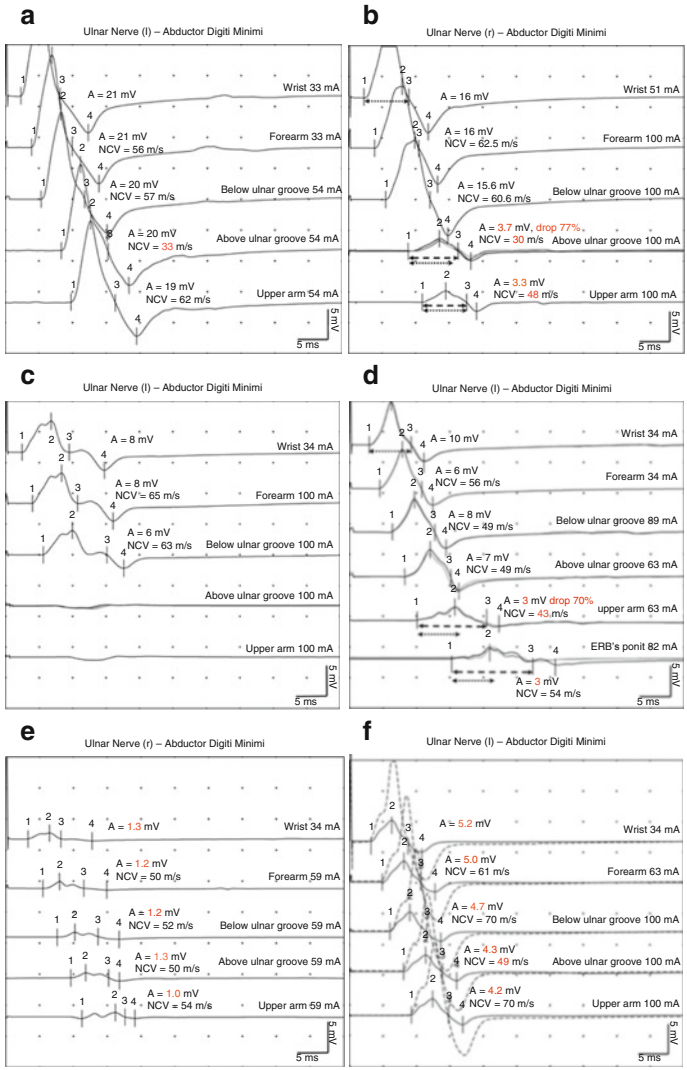
the nerve conduction velocity of distal nerve segments cannot be calculated. DML can therefore be explained by the fact that the exact distance between the stimulation electrode and the endplate region of the supplied muscle is unknown. Moreover, distal motor latency contains the indeterminable time needed for neuromuscular transmission and the generation of muscle contraction (CMAP) [2, 3, 9–12, 15].

Apart from prolonged distal motor latencies and lowered conduction velocities, there are two other conditions that typically occur in demyelinating nerve disorders, referred to as "abnormal temporal dispersion" and "conduction block" [13, 14]. As mentioned above, in a healthy peripheral nerve the duration of the CMAP after proximal stimulation normally increases and the CMAP amplitude decreases compared with the CMAP recorded after distal stimulation. This feature is termed "temporal dispersion." In the case of a demyelinating disorder, temporal dispersion is further increased because of the lowered conduction velocity

and increasing heterogeneity of the motor nerve conduction velocities of different A-alpha fibers generating the CMAP. Consequently, besides the prolonged duration of CMAP and the decay of its amplitude, it appears to be split at and above the site of the lesion, whereas the distal CMAP remains normal (Fig. 5.3d) [9–12]. In scientific studies, different consensus criteria regarding the "abnormal temporal dispersion" have been published. According to the American Academy of Neurology, temporal dispersion is abnormal "if the CMAP obtained after proximal stimulation shows more than a 20 % decay of the amplitude and more than a 15 % increase of the duration compared with the CMAP obtained after distal stimulation" [13]. In contrast, the criteria of the American Association of Neuromuscular and Electrodiagnostic Medicine assume an abnormal temporal dispersion "if the duration of the proximal CMAP is prolonged by more than 30 % compared with the duration of the distal CMAP." It is assumed that segmental demyelination over longer distances is responsible for this phenomenon [14].

On the other hand, a short segmental demyelination is attributed to a "conduction block." If the conduction block is complete, a normal CMAP can be recorded after distal stimulation, whereas stimulation proximal to the lesion will generate no CMAP (Fig. 5.3c). An incomplete conduction block is assumed if the CMAP amplitude (or area under the curve) after proximal

Fig. 5.3 Different types of demyelinating lesions on the example of ulnar neuropathy at the elbow obtained with fractionated motor nerve conduction studies. (**a**) Segmental slowing of the motor nerve conduction velocity. (**b**) Incomplete conduction block. (**c**) Complete conduction block. (**d**) Abnormal "temporal dispersion." For details please refer to the text. (**e**) Pure axonal loss. The site of the lesion is not detectable with use of a motor nerve conduction study. (**f**) Both axonal loss and focal demyelination. *Dotted line*: CMAP of the ulnar nerve of the healthy side


a
Ulnar Nerve (l) – Abductor Digiti Minimi
A = 21 mV
Wrist 33 mA
A = 21 mV
NCV = 56 m/s
Forearm 33 mA
A = 20 mV
NCV = 57 m/s
Below ulnar groove 54 mA
A = 20 mV
NCV = 33 m/s
Above ulnar groove 54 mA
A = 19 mV
NCV = 62 m/s
Upper arm 54 mA
5 mV
5 ms
b
Ulnar Nerve (r) – Abductor Digiti Minimi
A = 16 mV
Wrist 51 mA
A = 16 mV
NCV = 62.5 m/s
Forearm 100 mA
A = 15.6 mV
NCV = 60.6 m/s
Below ulnar groove 100 mA
A = 3.7 mV, drop 77%
NCV = 30 m/s
Above ulnar groove 100 mA
A = 3.3 mV
NCV = 48 m/s
Upper arm 100 mA
5 mV
5 ms
c
Ulnar Nerve (l) – Abductor Digiti Minimi
A = 8 mV
Wrist 34 mA
A = 8 mV
NCV = 65 m/s
Forearm 100 mA
A = 6 mV
NCV = 63 m/s
Below ulnar groove 100 mA
Above ulnar groove 100 mA
Upper arm 100 mA
5 mV
5 ms
d
Ulnar Nerve (l) – Abductor Digiti Minimi
A = 10 mV
Wrist 34 mA
A = 6 mV
NCV = 56 m/s
Forearm 34 mA
A = 8 mV
NCV = 49 m/s
Below ulnar groove 89 mA
A = 7 mV
NCV = 49 m/s
Above ulnar groove 63 mA
A = 3 mV drop 70%
NCV = 43 m/s
upper arm 63 mA
ERB's ponit 82 mA
A = 3 mV
NCV = 54 m/s
5 mV
5 ms
e
Ulnar Nerve (r) – Abductor Digiti Minimi
A = 1.3 mV
Wrist 34 mA
A = 1.2 mV
NCV = 50 m/s
Forearm 59 mA
A = 1.2 mV
NCV = 52 m/s
Below ulnar groove 59 mA
A = 1.3 mV
NCV = 50 m/s
Above ulnar groove 59 mA
A = 1.0 mV
NCV = 54 m/s
Upper arm 59 mA
5 mV
5 ms
f
Ulnar Nerve (l) – Abductor Digiti Minimi
A = 5.2 mV
Wrist 34 mA
A = 5.0 mV
NCV = 61 m/s
Forearm 63 mA
A = 4.7 mV
NCV = 70 m/s
Below ulnar groove 100 mA
A = 4.3 mV
NCV = 49 m/s
Above ulnar groove 100 mA
A = 4.2 mV
NCV = 70 m/s
Upper arm 100 mA
5 mV
5 ms

stimulation is reduced only when with the CMAP after distal stimulation [9]. Depending on the affected nerve, there are again different criteria defining an incomplete conduction block. According to the American Association of Neuromuscular and Electrodiagnostic Medicine, a "drop of more than 50 % in proximal CMAP compared with CMAP obtained after distal stimulation in the median and ulnar nerves and more than 60 % in the tibial and peroneal nerves is considered as pathological, while CMAP duration increases by no more than 30 %" [14]. In contrast, the criteria of the American Academy of Neurology require a "20 % drop of the (proximal) CMAP area or amplitude compared after distal and proximal stimulation" [13].

Unfortunately, the diagnosis process for demyelinating disorders of the peripheral nerves can become very challenging. Primary axonal loss is sometimes accompanied by secondary demyelination (Fig. 5.3f). This explains why slowed conduction velocities and prolonged distal motor latencies can occur in an axonal disorder [12]. Moreover, during the first few days after severe axonal nerve damage this condition will present a complete or incomplete conduction block because Wallerian degeneration has nor been completed yet and the segment distal to the damage is still conducting. This phenomenon has frequently been referred to as a "pseudo-conduction-block" [20]. Therefore, different criteria have been suggested, considering motor conduction velocities, distal motor latencies, and F-wave latencies. For instance, Van den Bergh and Pieret assume inter alia a motor conduction velocity slowing of <70 % of the lower limit of normal and a prolonged distal motor latency of >150 % of the upper limit of normal to be characteristic of a demyelinating condition [21].

5.3.3 Axonal Disorders

Axonal loss leads to a reduction of the activated muscle fibers and is thus paramount to a reduction of CMAP amplitude. In a fractionated motor nerve conduction study this applies to all

generated CMAPS – distally and proximally. As a consequence, and in contrast to focal demyelinating disorders, a localization of the lesion with motor NCS is impossible (Fig. 5.3e) [10–12]. This important lack of diagnostic power can be solved by needle electromyography and nerve imaging (see Sect. 5.6 and Chap. 6) [6, 22]. Furthermore, at an early stage (days 0–14) after onset of an axonal injury, this presentation can be misdiagnosed as a conduction block (see Sect. 5.3.2) [20]. If axonal loss also includes the fastest conducting A-alpha fibers, motor nerve conduction velocities may also be reduced. However, the extent of conduction slowing is less than following nerve fiber demyelination, and it usually ranges above 70 % of the lower limits of normal [2, 3, 21].

5.4 Sensory Nerve Conduction Studies

5.4.1 Technique and Parameters

As in a motor NCS, the peripheral nerve is stimulated with a transcutaneous depolarizing square wave electrical pulse (stimulation electrode) of a very short duration. However, a lower stimulation intensity is required than in a motor NCS. In contrast to motor NCS, a self-adhesive recording electrode can be placed either distally (antidromic recording) or proximally (orthodromic recording) to the stimulus site because the evoked electrical activity propagates in both directions. Furthermore, a ground electrode is also necessary [9]. In clinical practice, antidromic recordings are preferred, mainly because the amplitude of the sensory nerve action potential (SNAP) is larger than that obtained by orthodromic recordings [10–12]. Sensory nerve conduction studies can also be performed with needle electrodes; however, these special techniques are discussed elsewhere [2, 3]. Compared with CMAPs, SNAPs are smaller and measured in microvolts. Therefore, an averaging of several

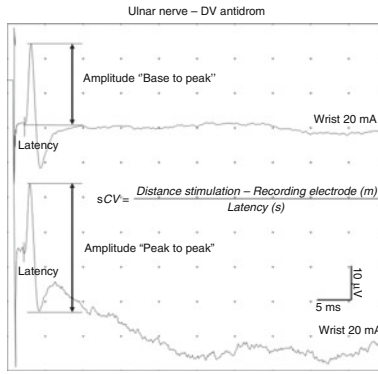


Fig. 5.4 Important parameters derived from sensory nerve conduction studies and calculation of the sensory nerve conduction velocity

SNAPs is required to improve the signal-to-noise ratio (SNR). Most important parameters derived from sensory NCS are as follows (Fig. 5.4) [10–12]:

SNAP amplitude reflects the number of the largest activated sensory nerve fibers (A-alpha). It is measured (base to peak or peak to peak) in microvolts.

SNAP conduction velocity is easily calculated using only a single site measurement because the distance between the recording and stimulating electrodes and the sensory latency is directly measurable. It is measured in meters per second.

As mentioned above, only the largest and fastest conducting sensory A-alpha fibers, which functionally supply senses of touch, position, and vibration senses, can be assessed using sensory NCS. Usually, small fiber neuropathies affecting C-fibers mediating pain do not show any abnormality in conventional sensory NCS [4, 8]. Quantitative sensory testing and autonomic testing or skin biopsy are necessary to provide the

correct diagnosis in this situation. These special techniques are described elsewhere [2, 3].

5.4.2 Demyelination Disorders

As in motor NCS, the primary feature of demyelination is a reduction of sensory nerve conduction velocity [10–12]. Additionally, the duration of SNAP increases owing to an enlarged range of nerve conduction velocity. It leads to "temporal dispersion" and "phase cancellation" (Fig. 5.5). Consequently, the SNAP amplitude will also drop in a demyelinating disorder [10–12, 23].

5.4.3 Axonal Disorders

Axonal disorders will primarily reduce the amplitude of the SNAP generated after electrical stimulation, whereas its duration is normal. As in motor NCS, further axonal loss of the fastest conducting fibers will also lead to a modest slowing of the sensory nerve conduction velocities (Fig. 5.5) [10–12, 21, 23].

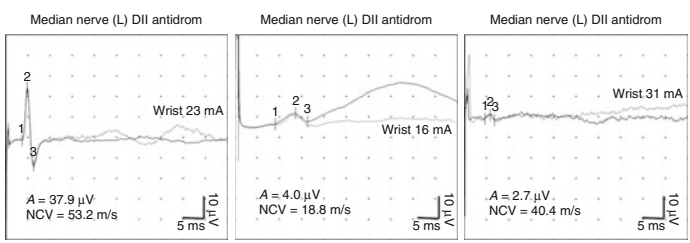


Fig. 5.5 Sensory nerve conduction studies of the median nerve. *Left*: physiological. *Middle*: demyelinating disorder with slowing of motor nerve conduction velocity and "temporal dispersion." *Right*: pure axonal loss

Sensory NCS are useful to determine whether the site of the nerve lesion is pre- or postganglionic. In the case of a preganglionic lesion the connection between the cell bodies of the dorsal root ganglion and the peripheral nerve remains intact. Consequently, a normal SNAP and conduction velocity can be obtained by sensory NCS, despite anesthesia in the dermatome supplied by that root. However, an absent or pathologically changed SNAP indicates a postganglionic lesion either of the nerve plexus or of a single peripheral nerve [24]. Apart from this electrophysiological dogma, in some recent studies it has been shown that – in a small percentage of subjects (2.4–12.1 %) with preganglionic lesions due to a L5 or S1 radiculopathy – the SNAP amplitude was abnormal. This exception may be due to dorsal root ganglion compression proximal to the spinal foramen or intra-spinally, and should be always kept in mind [25, 26].

5.5 Long Latency Reflexes (F-Waves)

5.5.1 Technique and Parameters

F-waves are a type of late motor response. Their name is derived from the foot, where they were first described. With a reversed stimulation electrode, F-waves can be recorded in the same setting as stated in Sect. 5.3.1 concerning motor nerve conduction studies [9]. If a nerve is electrically stimulated above its final segment the action potential propagates in both directions.

The orthodromic propagated action potential generates CMAP, also known as an M-response. On the other hand, the antidromic propagated action potential reaches the anterior horn cell body and depolarizes the axon hillock. As a result, a

small portion of the alpha-motor neurons backfire and a second orthodromic action potential reaches the muscle. This causes a late muscle depolarization that involves only a small portion of the muscle fibers, referred to as an F-wave [27–31]. Recording F-waves will therefore allow the electrodiagnostic testing of long and proximal nerve segments (e.g., the lumbosacral plexus or nerve roots) which are inaccessible to conventional electrical nerve stimulation [29]. In contrast to the M-response, F-waves usually vary in latency and shape because different populations of motor neurons backfire with each stimulus. Therefore, normally, "10–20 consecutive stimulations will be needed and the most reliable measure of the F-wave is its minimum latency of 10–20 firings" [29, 31]. It strongly depends on the body height, which should be taken into account. Moreover, the "F-wave persistence" is commonly used and represents the number of F-waves obtained per number of stimulations [27–31]. Regarding other parameters (*dispersion*, *amplitude-ratios*) derived from F-wave studies please refer to specialist literature [29].

5.5.2 Clinical Application

In generalized demyelinating nerve disorders (e.g., chronic and acute inflammatory demyelinating neuropathy) the proximal nerve segments or nerve roots are often affected by focal demyelination. This fact leads to decreased F-wave persistence or even to their absence (Fig. 5.6). Furthermore, the minimum F-latency is prolonged [32, 33]. F-waves may also be found to be pathological in other proximal axonal or demyelinating peripheral nerve disorders. Discussion of the value of F-wave studies in detecting radiculopathies has, however, been controversial [29]. Early studies reported lower sensitivity (50–80 %),

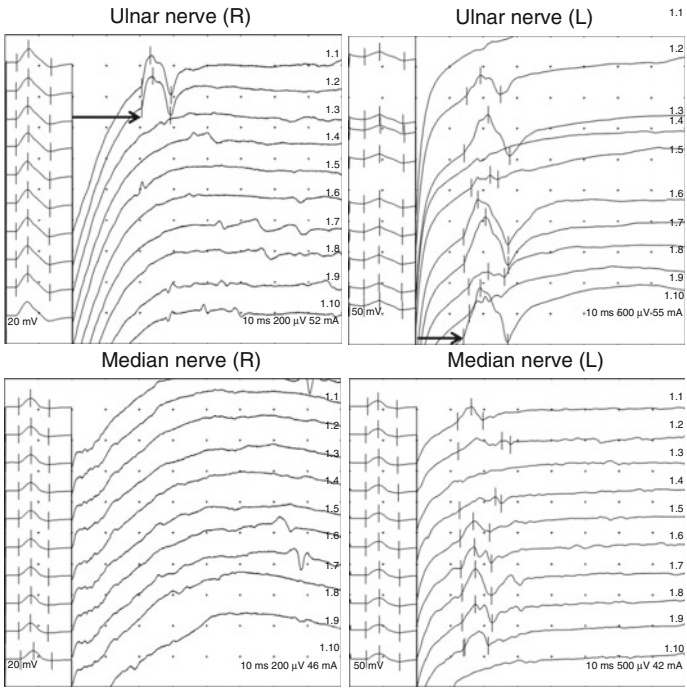


Fig. 5.6 F-waves in a patient with a lesion of the lower brachial plexus at the right side. Note the prolonged F-latency (*arrow*) and the decreased persistence in the right ulnar nerve and the absent F-waves in the right median nerve

whereas recent studies in patients with L5/S1 radiculopathies, considering not only latencies but also additional parameters such as persistence, dispersion, and amplitude ratios, reported sensitivities (90 % for L5 and 80 % for S1 radiculopathy), comparable to needle EMG [29, 34, 35]. Unfortunately, few data exist to evaluate the diagnostic value of F-waves in cervical radiculopathies. A recent study reported low sensitivity of only 55 % [36].

5.6 Needle Electromyography

5.6.1 Technique and Parameters in Healthy Subjects

Needle electromyography is commonly used to assess axonal nerve lesions, thereby delivering complimentary information to nerve conduction studies. In pure demyelinating disorders, however, no or only a few additional findings can be obtained by myography [22, 37–39]. In this section, we focus on the basic application of needle electromyography in disorders of the peripheral nerve. For special techniques such as single fiber electromyography, macro-electromyography, turns–amplitude analyses, application on myopathies, and disorders of the neuromuscular junction, please refer to specialist literature [2, 3].

A concentric needle electrode is inserted into the target muscle and the electrical activity of its muscle fibers is recorded during rest and periods of voluntary muscle contraction with minimal and maximal force. As in nerve conduction studies, a self-adhesive ground electrode is placed somewhere on the skin to provide a zero voltage reference point [9].

5.6.1.1 Normal Findings at Rest

The needle insertion in the muscle belly leads to a mechanical irritation of muscle fibers close to the tip of the needle electrode. As a result every forward movement of the needle produces bursts of electrical activity that vary in shape. This activity is also referred to as "*insertional activity*, never lasting longer than 300 ms" (Fig. 5.7a) [9, 22]. Accidentally hitting the endplate area of the target muscle, which is rather unpleasant for the patient, can irritate terminal axons, and results in two other types of spontaneous activity known as "endplate spikes" and

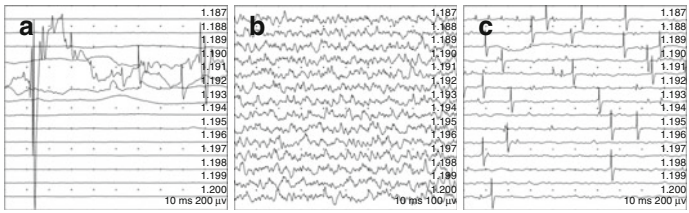


Fig. 5.7 Normal activity during needle electromyography at rest. (**a**) Insertional activity followed by electrical silence. (**b**, **c**) Endplate noise and endplate spikes (note the initial upward deflection and irregularity)

"endplate noise," also referred to as "endplate potentials" (Fig. 5.7b, c). Endplate spikes are biphasic potentials characterized by a negative (upward) onset and an irregular discharge frequency, whereas endplate noise is monophasic and of lower amplitude, but also shows a negative onset and an irregular discharge frequency [40].

5.6.1.2 Normal Findings During Voluntary Muscle

Contraction of Minimal Force

Voluntary muscle contraction leads to activation of several "motor units". A motor unit is represented by a single α-motor neuron in the spinal cord and all the individual muscle fibers it innervates [41–43]. As the extent of voluntary muscle contraction increases, more and more motor units are recruited. The potentials generated by a motor unit (motor unit action potential, MUAP) can be recorded using modern EDX equipment. However, with conventional needle electrodes it is impossible to assess the whole territory of a motor unit. Only the muscle fibers close to its tip (located within the recording radius of the needle electrode) generate the MUAP. Special techniques ("Macro-EMG") will allow the entire motor unit to be recorded [2, 3, 22].

Typically, motor units do not fire rhythmically, which means with the same discharge frequency, in contrast to pathological spontaneous activity (see Sect. 5.6.2.1). In healthy muscles these discharges of a single motor unit never exceed 20–50 Hz [42, 43]. The most important parameters of a MUAP are (Fig. 5.9a):

Amplitude is related to the size and density of the muscle fibers within the recording radius of the needle electrode [44–46].

Duration is determined by the number and size of muscle fibers of a particular motor unit close to the recording electrode. Furthermore, the spatial dispersion of the terminal axons and differing conduction velocities within the terminal axons can influence it [46].

Phases determine the number of baseline crossings of the MUAP. Up to four phases are considered normal. MUAPs with more than four phases are called "polyphasic." In healthy individuals polyphasic MUAPs should never exceed 10 % of all recorded and analyzed MUAPs of an individual muscle. In the case of asynchronous firing muscle fibers within the entire motor unit, which occurs in both neuropathic and myopathic disorders, the number of polyphasic MUAPs is increased. This also holds true for an increased number of *turns* – defined by changed direction of the MUAP without crossing the baseline [22, 46, 47].

Stability: individual MUAPs of healthy individuals do not change their morphology or shape. They are stable. Unstable MUAPs indicate diseases of the neuromuscular junction (e.g., myasthenia gravis), or they occur secondary to neuropathic or myopathic disorders [22].

Factors that influence the MUAP amplitude and duration are the type of muscle and the age. The latter may be explained by

an increasing loss of anterior horn cells in individuals of advanced age. This leads to an incorporation of denervated muscle fibers into surviving motor units, resulting in an increase in the amplitude of the MUAP [22]. Normal values for MUAP amplitude and duration for individual muscles are either derived from studies in the laboratories or obtained from related literature [2, 3, 9].

5.6.1.3 Normal Findings During Voluntary Muscle Contraction of Maximal Force

As explained above, as the force of the muscle contraction increases, more and more motor units, and finally the largest ones, are recruited. Greater voluntary activity will also increase their discharge frequency. In a healthy individual this process proceeds gently and continuously. Accordingly, a dense "interference pattern" appears and the individual MUAPs can no longer be differentiated from each other [9, 22].

5.6.2 Needle Electromyography in Focal and Generalized Neuropathies

5.6.2.1 Findings at Rest

Except for insertional activity and endplate potentials (see Sect. 5.6.1.1), no other type of electrical activity should be observed in healthy muscle tissue. This means that in the absence of a forward movement of the needle electrode the normal muscle is electrically silent in its resting state. Pathologically altered insertional activity is characterized by a "prolonged insertional activity" lasting longer than 300 ms after the forward movement of the needle and/or "pathological spontaneous activity." On the other hand, a diminished or absent insertional

activity occurs in muscles affected by an acute compartment syndrome, whereas additional increased needle insertion resistance is typical of a late stage of axonal loss where muscle parenchyma is replaced with connective tissue owing to absent reinnervation [9].

In the case of axonotmesis, the affected muscle fibers mainly show two patterns of pathological spontaneous activity: "fibrillations" and "positive sharp waves." Both appear approximately 10–14 days after onset of an axonal loss because the Wallerian degeneration needs to be completed first. In contrast to MUAPs, they always fire with the same discharge frequency. Compared with normal endplate potentials, biphasic fibrillations (Fig. 5.8a) and monophasic positive sharp waves (Fig. 5.8b) are characterized by an initial positive (downward) deflection. They result in the separation of muscle fibers from the degenerated axons. The extent of denervation is measurable semi-quantitatively by the frequency of occurrence of spontaneous activity within different muscle areas. Additionally, the distribution of the denervation pattern within different individual muscles allows localization of the underlying pathology as well as differentiation between focal and generalized axonal disorders. Either with successfully completed incorporation of muscle fibers in surviving motor units or with their death because of failed reinnervation, fibrillations and positive sharp waves will disappear again [22, 48–50]. "Complex repetitive discharges" are complex waveforms consisting of fibrillations and positive sharp waves. If they are observed in a neuropathy, they indicate a chronic denervation process where denervation is followed by reinnervation and subsequent denervation. They are characterized by an abrupt beginning and end (Fig. 5.8c) [22, 51, 52]. "Fasciculations" are spontaneous and not voluntarily activated MUAPs (Fig. 5.8d). They can be observed in healthy subjects (benign fasciculations). If they occur along with fibrillations, positive sharp waves or abnormally changed MUAPs, they are considered to be pathological and indicate a neuropathic disorder. Anterior

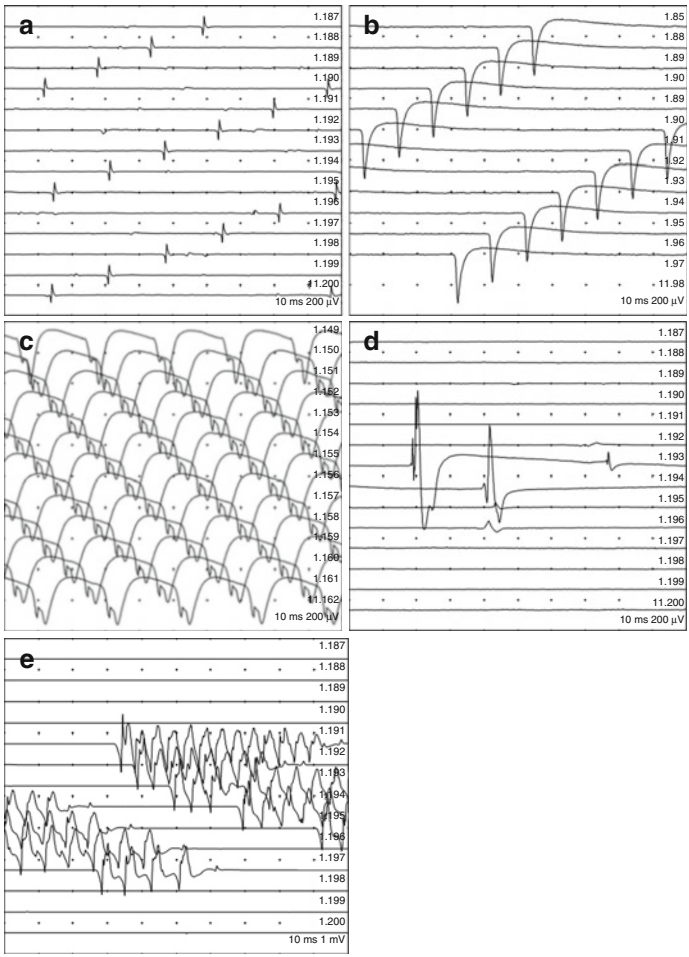


Fig. 5.8 Different abnormal findings (spontaneous activity) during needle electromyography at rest in neuropathies. (**a**) Fibrillations. (**b**) Positive sharp waves (note the initial downward deflection and the regular discharging frequency). (**c**) Complex repetitive discharges. (**d**) Fasciculations with the same appearance as motor unit action potentials (MUAPs). (**e**) Myokymic discharges

horn cell diseases also show the above-mentioned combination of different types of pathological spontaneous activity. Groups of two, three or more fasciculations, also known as "doublets, triplets and multiplets," have the same clinical significance [53]. "Myokymic discharges" are characterized by repetitive bursts of MUAPs (Fig. 5.8e) and occur along with radiculopathies and radiation neuropathies [54].

5.6.2.2 Findings During Voluntary Muscle Contraction of Minimal Force

At an acute stage of complete axonal loss, there is no voluntary muscle contraction and no individual MUAPs can be recorded. If the axonal damage is incomplete the remaining motor units generate MUAPs with normal duration, amplitude, and number of phases at this time. However, a decreased recruitment pattern in weak muscles – owing to the loss of motor units – and an increased discharge frequency of the remaining motor units can indicate an early incomplete lesion. In contrast, this finding does not occur when there is insufficient cooperation from the patient during examination nor in the case of central paralysis where the second neuron is unaffected. If muscles clinically appear completely paralyzed, e.g., in the case of a neuropathy, needle electromyography can demonstrate that the function of some individual motor units is preserved [9, 55].

Based on the intactness of anterior horn cells, nerve connective tissue, and the continuity of the epi- and perineurium respectively, and not on a disrupted or inadequately surgically repaired nerve, reinnervation of muscle fibers starts along the original nerve fiber tracts with a growth rate of about 1–2 mm/day [56]. A long distance between the injured nerve segment and its target muscle only leads to incomplete reinnervation because a restructuring of connective tissue first has to take place, or, as mentioned in Sect. 5.3.2, resulting fibroses may additionally hinder the sprouting process of axons. In the case

of a preceding complete denervation, the ingrowing axons initially reach only a small number of surviving muscle fibers; thus, the corresponding MUAPs appear with a short duration, low amplitude, reduced recruitment, and increased number of phases owing to the "spatial dispersion" of endplates, as usually seen in myopathies (Fig. 5.9b) [22]. With reinnervation

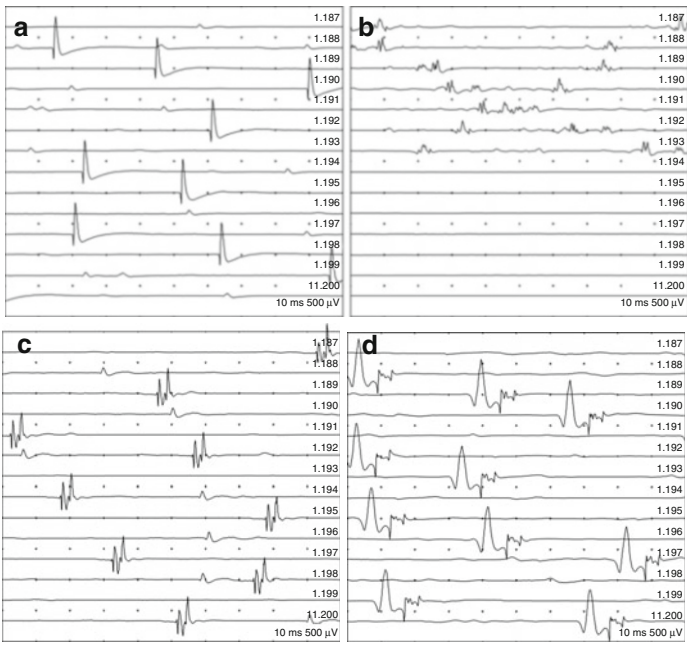


Fig. 5.9 Electromyographic findings during voluntary muscle contraction at minimal force in neuropathies. (**a**) Normal MUAPs. (**b**) Early reinnervation. MUAPs appear small and polyphasic as in a myopathy. (**c**, **d**) Reinnervation with polyphasic MUAPs and late components. Note the prolonged duration and increased amplitude. (**e**) Late stages of reinnervation. Finally, the polyphasic components will disappear, although amplitude increases further

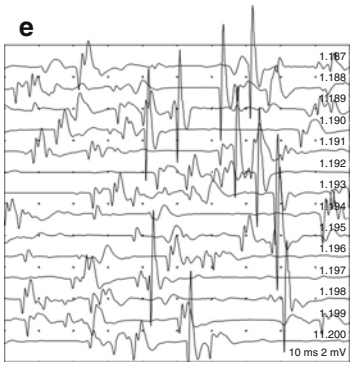


Fig. 5.9 (continued)

proceeding in the absence of fibrosis or inadequate nerve reconstruction, the territory of the motor unit increases in size because more and more muscle fibers become incorporated. The same process occurs in chronic partial denervation, where surviving motor units incorporate – by means of collateral axonal sprouting – muscle fibers from dead ones. Therefore, the number of muscle fibers supplied by such a "neuropathic" motor unit can be higher than in normal individuals [57]. Thus, the resulting "neuropathic" MUAPs appear with a longer duration together with increased amplitude (Fig. 5.9c). Because of slowed conduction along the newly-formed terminal axons and increased "spatial dispersion" of the endplates, these MUAPs also have more phases – and sometimes satellite potentials (Fig. 5.9d). In the late stages, polyphasic changes of the MUAP will disappear again. In addition to that, increasing conduction velocities and lowered "spatial dispersion" also occur; however, the amplitude remains larger than it usually appears in healthy subjects [22]. An example is shown in Fig. 5.9e.

5.6.2.3 Findings During Voluntary Muscle Contraction of Maximal Force

Because of the smaller total number of motor units, the density of the interference pattern is thinned out. In severe cases, individual MUAPs will then be visible. If reinnervation takes place the remaining motor units increase their territory by incorporating denervated muscle fibers. Accordingly, the amplitude of interference pattern is also increased [22, 37–39].

5.7 Limitations and Pitfalls

Both nerve conduction studies and needle electromyography may be unpleasant for some patients. While nerve conduction studies with surface electrodes are non-invasive, repeated needle electrode insertion occasionally leads to complications such as bleeding, infection or nerve injury in individuals undergoing electromyography [58]. The literature states that needle electromyography is relatively contra-indicated in patients treated with antiplatelet agents and anticoagulants [9]. However, in recent studies there has been no statistically significant difference between muscle hematoma occurrence following needle electromyography in patients undergoing therapy with warfarin or antiplatelet agents and in nontreated individuals. Only a small amount of subclinical muscle hematoma was observed in the group receiving blood-thinning medication [59, 60]. Furthermore, nerve conduction studies may theoretically impair the function of implanted biomedical devices such as pacemakers or cardiac defibrillators. In some recent studies, though, it was demonstrated that, in nerve conduction studies at routine sites, no electrical impulses could be detected by the sensing systems of these devices [61, 62]. It has therefore been suggested that the current guidelines restricting nerve conduction

studies in such patients should be subject to revision [62]. Even repetitive nerve stimulation is safe in patients with implanted cardiac defibrillators, and it appears safe in patients with implanted pacemakers using bipolar sensing. Caution is only recommended in pacemaker patients with unipolar sensing systems [63].

The correctness of results provided by EDX substantially depends on the experience of the investigator. Many pitfalls can arise such as false evaluation due to anomalous innervation, improper machine settings, and insufficient patient preparation (skin temperature). For a detailed overview please refer to specialist literature [64].

5.8 Conclusions

Electrodiagnostic examination provides clinically important information on both focal and generalized disorders of the peripheral nervous system. On the other hand, it cannot be overemphasized that EDX is only an extension of our clinical examination and anamnesis survey and, although we never neglect it, it will not replace either the clinical examination or an anamnesis survey. Nerve conduction studies facilitate differentiation between axonal and demyelinating disorders, and between a focal and generalized affection of the peripheral nerve. With fractionated motor nerve conduction studies or inching techniques, a demyelinating process can be located: first, by means of focal conduction velocity slowing or prolongation of distal motor latency; second, by evaluating a conduction block; and third, by signs of abnormal temporal dispersion. Moreover, sensory nerve conduction studies allow preganglionic lesions to be distinguished from postganglionic lesions in the majority of cases. Long latency reflexes (F-waves) provide a way of examining the proximal parts of the peripheral nervous system

(plexus and root segments) that are otherwise inaccessible to conventional electrical nerve stimulation. In all types of axonal loss (acute, subacute, chronic), needle electromyography, along with nerve conduction studies, provides information on their extent, localization, and acuity. Additionally, needle electromyography provides a highly reliable assessment of an important question – whether reinnervation takes place or not – which will help to determine the prognosis of a focal peripheral nerve disorder.

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Chapter 6
Imaging of the Peripheral Nerves

Thomas Schelle

6.1 High Resolution Ultrasound (HRUS)

6.1.1 Introduction

In 1988, Fornage was the first to introduce the sonography of peripheral nerves [1]. However, with the previously available ultrasound equipment, only the larger pathological alterations such as mass lesions could be detected. In the meantime, all major manufacturers have improved the resolution and quality of their ultrasound images dramatically by means of broad band linear array transducers up to 18 MHz as well as by the introduction of new image processing technologies such as compound imaging and tissue harmonic imaging. Consequently, today, modern techniques facilitate the identification of a number of disorders of the peripheral nerve, even detecting alterations of a single fascicle and its perineurium [2]. At the moment, however, it remains unclear whether additional new techniques like ultrasound contrast agents or elastometric measurements will generate still more information in order to extend further the diagnostic power of this

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method. Nevertheless, in a recent study it has been demonstrated that contrast enhanced ultrasound (CEUS) seems to be able to detect several perfusion patterns in soft-tissue masses and thus at least to predict malignancy in peripheral nerve sheath tumors [3].

6.1.2 Minimum Technical Requirements

An ultrasound device with a linear broad band transducer up to at least 10 MHz is the minimum technical requirement. With this equipment, an axial resolution of 0.2 mm and a lateral resolution of 0.6 mm can be achieved in the ultrasound image. Ideal are multi-frequency transducers with a wide frequency range from 6 to 18 MHz. Furthermore, the device must have the capability to record various images under different sub-apertures in real time simultaneously. Afterwards, these single frames are mathematically compounded, and, from that, the final image emerges (Compound Imaging). The result is, first, significant noise reduction, and, second, better representation of the tissue boundaries. In addition, the use of tissue harmonic imaging (THI) can improve contrast and lateral resolution, especially in deep situated tissue structures [4]. Moreover, a highly qualitative colour-Doppler function is required to assess the amount of nerve vascularisation. Of course, the examiner should have a good knowledge of the musculoskeletal cross sectional anatomy he intends to display [5].

6.1.3 Examination Technique

Peripheral nerves can be quickly identified by means of anatomical landmarks. Initially, the investigator has to locate anatomical structures such as bones, muscles or blood vessels which lie

close to the affected nerve. Once found, from this point, the nerve can simply be traced upwards and downwards in cross sections [5]. For details concerning special nerve entrapments please refer to the ultrasound-imaging sections in Chaps. 8 and 9. Nerves and tendons are sonographically characterized by their anisotropy. For that reason, the optimal echogenicity of a nerve tissue can only be obtained by a strict perpendicular insonation. In order to improve further the image quality of diagnostic ultrasound, particularly in regions with an uneven body surface (e.g. the elbow), the use of aqueous coupling devices is recommended. As peripheral nerves often change their depth in their extremity course, a continuous adjustment of the electronic focus as well as of the transmission frequency of the probe is very important. With increasing transmission frequency (and resolution) the penetration depth decreases, and vice versa. Therefore, for example, the intrapelvic section of the sciatic nerve cannot be examined with high resolution sonography. Magnetic resonance tomography here proves to be superior and is thus an alternative solution. Pathological alterations are always documented in two planes (longitudinally as well as in cross section). Sometimes it may be preferable to record short video sequences. Special questions even require passive movement of the limb to be examined (e.g. snapping triceps syndrome) [5].

6.1.4 Ultrasound Anatomy of the Normal Peripheral Nerve

The cross sectional appearance of the normal peripheral nerve resembles a honeycomb (Fig. 6.1a). Axons and the surrounding endoneurium are arranged in bundles. Some of them form a single fascicle. High resolution ultrasound can resolve and depict single fascicles; however, single axons or the endoneurium are not visible [6]. Depending on the transmission frequency and

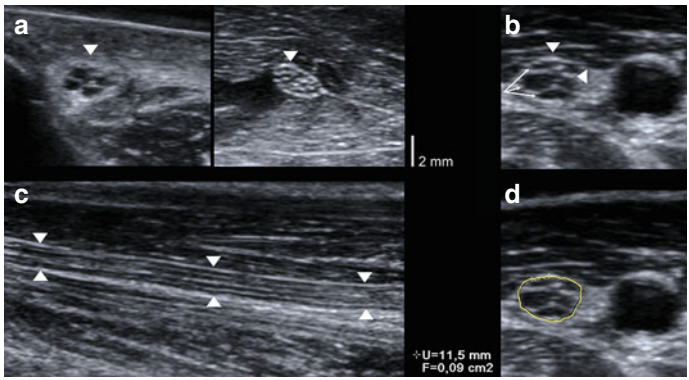

Fig. 6.1 Sonographic findings in the normal peripheral nerve. (**a**) Honeycomb-like appearance in cross sections. (**b**) Hypoechoic fascicles (*arrowheads*), hyperechoic peri- and epineurium (*arrows*). (**c**) Cable-like appearance in longitudinal sections. *Arrowheads*: peripheral nerve. (**d**) Correct measurement of the cross sectional area between hypoechoic fascicles and hyperechoic epineurium

resolution of the probe, the number of fascicles in the ultrasound image can differ from that determined histologically [7]. In cross sections single fascicles are sonographically characterized by a hypoechoic, tubular appearance. They are embedded in the perineurium and surrounded by the epineurium (or outer layers of perineurium). In contrast to fascicles, epineurium and perineurium show a hyperechoic appearance [6]. Figure 6.1c illustrates these findings. Frequently, it can be difficult to distinguish the very thin epineurium or outer perineurium from the surrounding tissue. In such situations it may help to move the probe upwards or downwards into a different position. In longitudinal sections, the peripheral nerve shows a cable-like course, in contrast to the more fibrillar echotexture of the tendons (Fig. 6.1b). In addition, tendons can be easily distinguished from the nerves sonographically because they always end in a muscle proximally [5, 7].

The most important quantitative parameter in the examination of the peripheral nerve is the measurement of its cross sectional area (CSA) [5]. Sometimes ratios between CSA obtained at different locations within one periphereal nerve or different peripheral nerves are calculated [8]. The measurement should be performed between the border of the hypoechoic fascicles and the hyperechoic epineurium (Fig. 6.1d). If no reference values are available, comparison with the CSA of the healthy side is suggested [5]. In most of the recent studies nerve CSA shows a positive correlation with age and is generally larger in males than in females [9–13]. Moreover is has been published that the nerve CSA was significantly larger in Dutch subjects when compared with Indan subjects. However, further studies are needed to determine the geographical dependency of the CSA. In the meantime it is therefore suggested that laboratories should obtain their own normal values [14]. Please refer to Boehm et al. 2014 and the imaging sections in Chaps. 8 and 9 to get an overview about published normal values of CSA [9].

As described in Sect. 2.2, the nerve starts at the root level with a mono-fascicular architecture, it then subdivides into a fascicular group arrangement and at the end it often demonstrates a multi-fascicular arrangement in the periphery. In parallel to this changing group arrangement, the amount of connective tissue between fascicles (inter fascicular epi- and perineurium) increases. This explains why cervical roots often demonstrate only one individual fascicle in sonography; meanwhile, the number of fascicles, e.g. in the median nerve, increases from proximal to distal. An example is shown in Fig. 6.2.

With the aid of colour flow Doppler functions of the current ultrasound equipment, the device is now able to identify the normal epineural vessels more frequently. However, excessive vascularisation ("hypervascularisation") is considered as non-physiological by some authors [15–17]. In a few conditions (e.g. entrapment syndromes) attempts have recently been made to quantify these changes [16, 17]. Due to lack of adequate studies

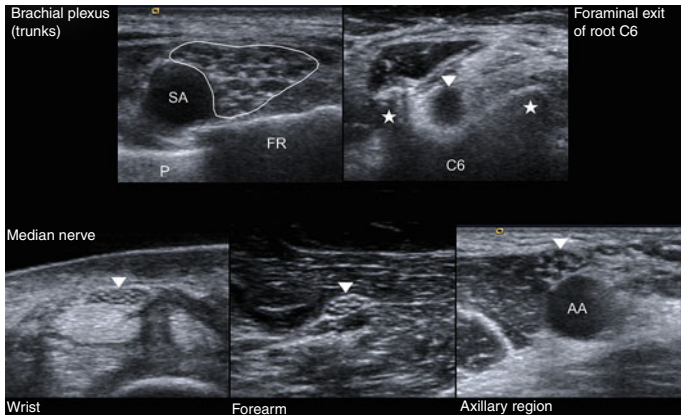

Fig. 6.2 Different sonographical appearance of the peripheral nerve depending on the anatomical localisation. *Upper part*, *left*: *SA* subclavian artery, *P* pleura, *FR* first rib. *Upper part*, *right*: *arrowhead* – root C6, *asterisks* – tubercula of the transverse processus of C6. *Lower part*: *arrowheads* – median nerve at different anatomical positions

and diagnostic criteria, we recommend to simply compare and record the vascularisation of the affected and the unaffected side qualitatively (Fig. 6.3). However, it has at least been shown that an increased intraneural vascularisation is associated with axonal loss in ulnar neuropathy cases at the elbow [17].

Another novel approach to detect abnormalities is assessing the echogenicity of nerve cross sections quantitatively [18, 19]. Thereby, quantification of nerve density – defined as the ratio between hypoechoic and hyperechoic areas/pixels of peripheral nerves in the ultrasound image – is capable of discriminating between normal and pathological nerves in carpal tunnel syndromes, respectively, between mild and severe CTS cases [19]. However, until now it remains unclear whether this method will be accepted in the clinical routine.

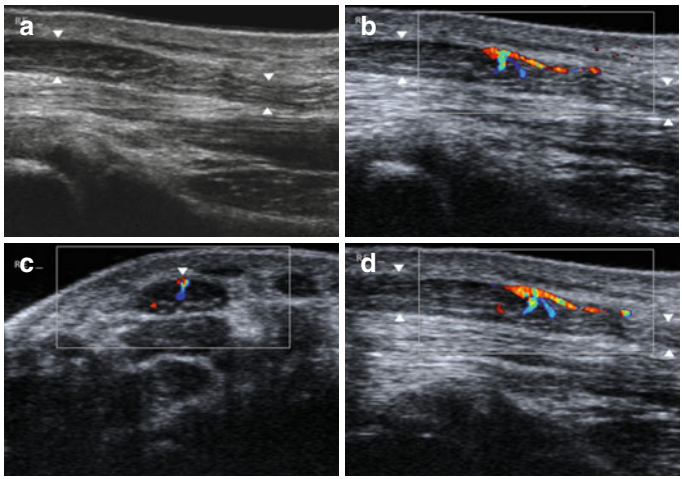

Fig. 6.3 Pathological vascularisation of the epineural vessels affecting the median nerve (*arrowheads*) proximal to entrapment (carpal tunnel syndrome). (**a**) Grayscale image. (**b**–**d**) Color-flow Doppler images showing the abnormal vascularisation in cross section (**c**) and longitudinal sections (**b**, **d**)

6.1.5 Application to Entrapment Neuropathies

If a peripheral nerve is externally compressed or entrapped by ligaments, bone fragments, callus or other pathological conditions, it always responds with the same morphological alteration. At the site of compression, an abrupt flattening occurs with a decrease of the cross sectional area and a decreased nerve diameter in longitudinal sections (Fig. 6.4b). Proximally and sometimes distally to the site of compression, a segmental swelling of the nerve can be observed and, consequently, the cross sectional area and/or nerve diameter increases within that nerve segment. Furthermore, in such cross sections, a typical hypoechoic swelling of the fascicles appears.

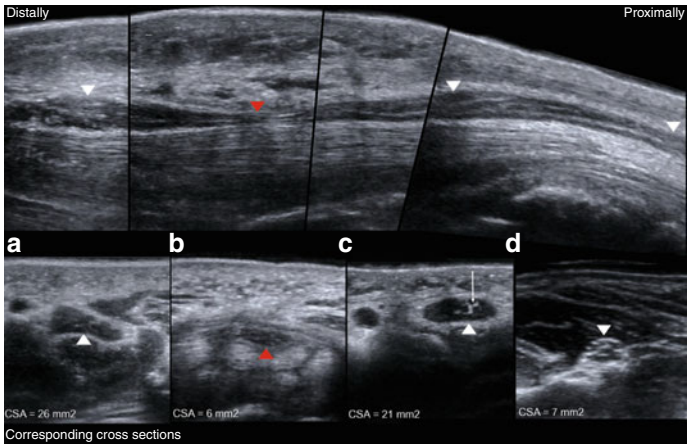

Fig. 6.4 Typical sonographical findings in an entrapment neuropathy. *Upper part*: longitudinal reconstruction of the median nerve at the wrist (carpal tunnel). *Lower part*: corresponding cross sections. *White arrowheads*: median nerve distal and proximal to the site of compression. *Red arrowheads*: median nerve at the site of compression. (**d**) Unaffected median nerve at the forearm with normal cross sectional area and honeycomb-like echotexture. (**c**) Proximal nerve swelling with hypoechoic fascicular effacement, oedema and hyperechoic epineural thickening as well as increased cross sectional area. *Small arrow*: suspected perineural fibrosis. (**b**) At the site of compression abrupt flattening of the median nerve (*red arrowhead*) with decreased cross sectional area. (**a**) Distal nerve swelling with similar changes as described in (**b**)

Later on, the fascicles are effaced due to the increasing oedema and the honeycomb-like echotexture is replaced by a homogeneous hypoechoic one [20–23]. This phenomenon is sometimes falsely referred to as "pseudoneuroma". If a distal nerve-swelling is also present, e.g. in advanced stages of carpal tunnel syndrome, the nerve demonstrates an hour-glass-like appearance (Fig. 6.4a, c). Actually, the cause of increased size and loss of echogenicity of compressed nerves is not well understood. Some authors discuss an increased vascularity of the nerve proximal to the site of entrapment [22]. As mentioned above, this has been

demonstrated using colour flow Doppler [15–17] (Fig. 6.3). After treatment of carpal tunnel syndrome with steroid injections, nerve swelling and vascularity as well as echogenicity decreased significantly [23]. Sometimes a blurred demarcation of the outer epineurium of the enlarged nerve segments can be observed, probably caused by an epineuritis, which may turn into a fibrosis of the epineurium later on. This reaction will be recognisable by a thickening of the hyperechoic epineurium on ultrasound [21] (Fig. 6.4c). According to our own experience, these typical signs may also be seen in conditions after nerve surgery, and, of course, following nerve injuries. Furthermore, it was reported that a thickened outer epineurium is a sonographic sign suggesting snapping ulnar nerve syndrome [24]. As a consequence of passive displacement of peripheral nerves in late stages of entrapment, the proximally thickened nerve segment may slowly develop signs of a reactive perineural fibrosis, a real pseudoneuroma. We have observed hyperechogenic spots within proximally enlarged hypoechoic nerve segments in patients with severe carpal tunnel syndrome. They may probably reflect the perineural fibrosis but a replacement of nerve tissue by fat after axonal loss is also conceivable (Fig. 6.4c). Interestingly, also in these late stages, proximal cut-off values of median nerve cross sections remain enlarged, and thus do not reflect signs of secondary nerve atrophy in the majority of cases [25].

In addition to these qualitative alterations characterizing nerve entrapment, quantitative measurements of different cross sectional areas have been introduced as the most commonly used parameter. One possible opportunity is related to the use of cut-off values at the site of the proximal nerve swelling. On the other hand, calculation of a ratio between the cross sectional area at the site of proximal nerve swelling and the cross sectional area which we obtain more proximally, where the nerve is in a healthy condition, is quite a reliable practice. These ratios are referred to as “wrist to forearm ratio” in carpal tunnel syndrome and “cubital to humeral ratio” in ulnar neuropathy at the elbow [26–30]. However, flattening ratios of nerve diameter,

amount of bowing of the flexor retinaculum, as well as characteristics of nerve mobility and echogenicity have been used infrequently [31].

Former systematic meta-analyses of clinical trials with measurements of the cross sectional area in carpal tunnel syndrome have evaluated inferiority compared with electrodiagnostic testing [32–34]. In contrast, the American Association of Neuromuscular and Electrodiagnostic Medicine recently published a systematic evidence-based review regarding the potential value of high resolution ultrasound as a "first line tool" in the diagnosis of carpal tunnel syndrome. Considering a total of 45 studies concerning sensitivity and specificity of HRUS compared to electrodiagnostic testing (4 of them had evidence-class I), HRUS was assigned recommendation level A. Moreover, HRUS for detection of structural and anatomical abnormalities in carpal tunnel syndrome was assigned recommendation level B after considering a total of 23 studies [35]. For more details (cut-off values or ratios in median nerve) please refer to Chap. 8. Unfortunately, the results derived from these studies concerning the correlation of cross sectional area measures with electrodiagnostic severity in carpal tunnel syndrome remain contradictory. Some investigators have found a strong correlation [36, 37], whereas others did not [38–40]. Concerning the ulnar neuropathy at the elbow, systematic evidence-based recommendations or systematic meta-analyses are currently missing. However, the most recent report still stated that CSA and swelling ratio indeed seem to have comparable diagnostic values [30, 41].

In summary, sensitivity and specificity obtained by high resolution ultrasound and electrodiagnostic testing in nerve entrapment do not differ significantly. However, only classical nerve conduction studies allow to asses the severity of a nerve entrapment, which means differentiating between demyelinising and axonal lesion. Furthermore, until now there has been a lack of evidence-based recommendations concerning ulnar neuropathy at the elbow [35, 39, 42].

In some individual cases (patients who refuse electrodiagnostic testing, children, patients with different overlapping conditions such as nerve entrapment and any kind of a generalized neuropathy, and patients in advanced stage of entrapment with axonal loss in whom the results of electrodiagnostic testing are ambiguous), HRUS may be used solely for assessment of nerve entrapment, taking into account the qualitative and quantitative changes mentioned above [5, 43, 44]. The remaining majority of patients should continue undergoing nerve conduction studies as the first-line test. Nevertheless, HRUS can provide additional information that may influence the diagnostic and therapeutic approach.

The median nerve in particular can present a couple of anatomical variations (high division, accessory branches, persistent median artery, etc.) in approximately 33 % of the symptomatic individuals [2, 26, 45]. High division of the median nerve is probably not a risk factor in developing a carpal tunnel syndrome and was observed in 14.4 % of healthy subjects and in 18.5 % of symptomatic patients [45]. Even small nerve branches such as the palmar cutaneous branch of the median nerve and its variations may be depicted [46]. Some might hold an increased risk of injury, particularly when modern endoscopic carpal tunnel release is applied. HRUS can detect dangerous anomalies before intervention [47, 48]. An example is shown in Fig. 6.5a.

Additionally, there are a lot of space-occupying lesions in the carpal tunnel that cause nerve compression (extraneural or intraneural ganglion cysts, which are described in Sect. 13.3.8, tumors of the peripheral nerve sheath, bone fragments, thrombosis of median artery, tendinitis, accessory muscles, etc.). Some require a different therapeutic approach (Fig. 6.5b, c). In these cases too, HRUS provides reliable information before surgery [2, 26, 49, 50].

Ulnar neuropathy at the elbow is sometimes characterized by two or even more compression mechanisms (ancoaneus epitrochlearis muscle, Struther's ligament, subluxation or luxation

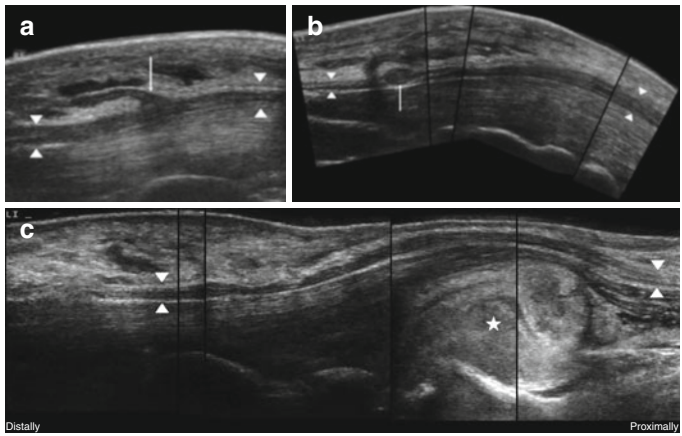

Fig. 6.5 HRUS, anatomical variations and secondary causes of nerve compression in the median nerve (*arrowheads*) at the level of carpal tunnel. (**a**) Transligamentous running branch supplying the thenar-muscles (*arrow*). (**b**) Small schwannoma (*arrow*) within the carpal tunnel. (**c**) Acute bleeding (*asterisk*) in a patient undergoing anticoagulant therapy

tendency, etc.). In a recent study, subluxation occurred in 14 % and luxation in 6.7 %, whereas in healthy controls only 5.7 % had either subluxation or luxation of the ulnar nerve [51]. According to another report, the values for subluxation and luxation were similar. A prevalence of accessory muscle was found to be 8.8 % among patients with ulnar neuropathy. Furthermore, secondary causes of compression (osteophytes, ganglionic cyst, osseous fragment and other post-traumatic lesions) have been observed in 12.1 % of cases [52]. The detection of post-traumatic lesions with multiple and/or atypical sites of entrapment as well as cubitus varus deformity is particularly important because they have an influence on the surgical approach. Therefore, HRUS provides a fast and cost-effective

way to perform the required surgery [53, 54]. For images and further details please refer to Chap. 8 (ulnar nerve).

If surgical intervention fails, e.g. after carpal tunnel release, high resolution HRUS will evaluate exactly the pitfalls (remaining stenosis caused by incomplete flexor retinaculum release or scar tissue, major nerve injury or even misleading preoperative diagnosis) [55–57]. However, a remaining reduction of median nerve cross sectional area after carpal tunnel release is at present still of limited value in the assessment of poor outcomes [58, 59]. An example is shown in Chap. 8 (median nerve).

Finally, ultrasound guided injection of corticosteroids in patients with ulnar neuropathy at the elbow as well as sonographically guided percutaneous needle release of the carpal tunnel may represent an alternative to the standard surgical approach in future [60, 61].

Unfortunately, there are only case reports or small case series regarding rare entrapment neuropathies such as tarsal tunnel syndrome, Guyon's canal syndrome, etc. Because of the lack of systematic studies, only normal values and some cut-off values but no CSA-ratios are available at the present. Diagnosis should be based on the qualitative characteristics of nerve entrapment mentioned above and on the published normal values of nerve cross sectional areas. Thus, nerve conduction studies in conjunction with needle electromyography still remain the first line tool within the diagnostic pathway. Similar to the more common entrapment neuropathies, HRUS can again demonstrate special features such as ganglion cysts in the tarsal tunnel or in the Guyon's canal as well as fibrous ligaments of the long peroneal muscle in case of peroneal nerve entrapment [21, 62]. Please refer to Chaps. 8 and 9 for additional details.

In summary, the diagnostic value of high resolution ultrasound in nerve entrapment mainly consists, first, of detection of anatomical anomalies and, second, of the evaluation of special pitfalls.

6.1.6 *Application to Focal Traumatic Nerve Lesions*

As previously mentioned, there are different degrees of severity of a nerve injury. For a detailed description please refer to Sects. 3.1 and 3.2. The Sunderland classification is based on the morphological changes of the injured nerve, and it is therefore the most appropriate to describe the corresponding sonomorphological alterations of a nerve injury [63]. As mentioned above, high resolution ultrasound can depict single fascicles, the perineurium and the epineurium of a peripheral nerve; however, it cannot resolve a single axon, its myelin-sheath and the corresponding endoneurium. Hence, a sonographical differentiation between grade I, II and III injuries is not possible. This restriction has less significance because, in practice, we need to distinguish between the minor lesions capable of spontaneous recovery, where surgical intervention is hardly needed, and the major lesions, where surgical treatment is almost always required. In the high resolution ultrasound related literature, the term "minor lesion" is applied to grade I–III lesions in accordance with the Sunderland stratification, whereas the term "major lesion" corresponds to a grade IV or V injury [64–66].

Minor lesions create no or only subtle changes on ultrasound. According to our experience the normal echotexture (honeycomb) below, above and at the site of injury is preserved. In the case of demyelination (grade I), either no alteration or a focal hypoechoic edema of fascicles and a consecutive segmental enlargement similar to entrapment neuropathies may be observed. If an axonal loss occurred with preserved perineurium (grades II and III, loss of axon continuity without or with injury to the endoneurium) the entire nerve segment, especially distal to the injury, will show hypoechoic enlarged fascicles and nerve swelling (Fig. 6.6a). However, mostly these subtle alterations are only recognizable if compared with the unaffected side [65].

In accordance with the latest studies performed by MR neurography in rats with traction injuries, the nerve enlargement and oedema will probably reflect the Wallerian degeneration; it may partly disappear when nerve regeneration takes place [66]. In grades II to III, continuity of the epineurium is always preserved. A pseudoneuroma due to intraneural connective tissue fibrosis may occur weeks later, but not a real neuroma formation [67].

However, major lesions always create distinct sonographic changes regardless of the time elapsed after injury. Grade IV lesions with preserved epineurium continuity are characterized by a partial or complete loss of the normal echotexture (honeycomb-like) at the site of the injury; it reflects the advanced destruction of fascicles and perineurium. In early stages, intraneural bleeding can sometimes be observed. Further characteristics – quite similar to grade II to III lesions and due to Wallerian degeneration – are mainly to be found in the nerve segment distally to the lesion. An additionally disrupted continuity of the epineurium with retracted nerve stumps indicates a grade V lesion (Fig. 6.6b, c). This kind of damage is easy to evaluate [64–66, 68, 69]. In the case of a later stage of a major lesion without any previous surgical treatment, HRUS can demonstrate a neuroma in continuity (in a grade IV lesion) or even a stump neuroma (then indicating a grade V lesion). A neuroma consists of nerve fibers regenerating from proximal and intraneural scar tissue which stops the sprouting process. Due to disrupted continuity of the peri- and/or endoneurium, and to severe reactive fibrosis, the newly sprouting nerve fibers are unable to find their way down to the target and thus will form a neuroma [70]. Neuromas sonographically appear as a globose, ovoid, fusiform and sometimes irregular, rather hypoechoic mass and have well-defined margins. The fascicles of the proximal nerve segment often enter the neuroma in a chaotic manner and disappear within it. Sometimes neuromas demonstrate an increased vascularisation. As mentioned, they are located either within the continuity of a nerve trunk or at the end of the proximal nerve

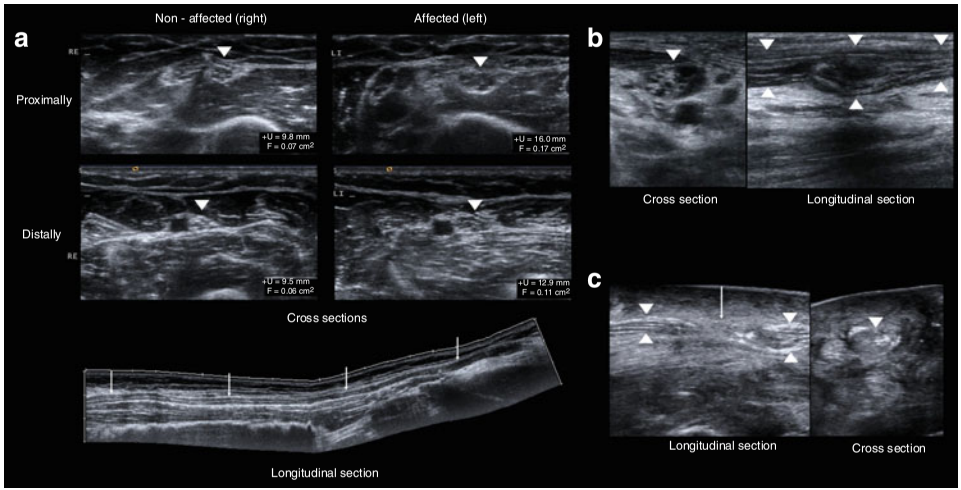

Fig. 6.6 Minor and major lesions following a nerve injury in HRUS. (**a**) Minor lesion (II/III according to SUNDERLAND) presenting an oedema of the fascicles and an enlargement of the entire nerve segment especially distal to the lesion after traction injury of ulnar nerve and compared with the unaffected side. The normal honeycomb-like appearance of the nerve in cross sections is preserved. *Arrowheads* – ulnar nerve. *Arrows* – decreasing oedema from proximal to distal. (**b**) Type IV lesion with a partly destroyed normal echotexture of the nerve (*arrowheads*) in cross sections and the cable-like appearance in longitudinal sections at the site of the injury; however, the continuity of the epineurium is preserved. (**c**) Type V lesion with interrupted continuity (*arrow*) of the peripheral nerve (*arrowheads*), complete destruction of the normal echotexture in cross sections at the site of the injury and retracted nerve stumps

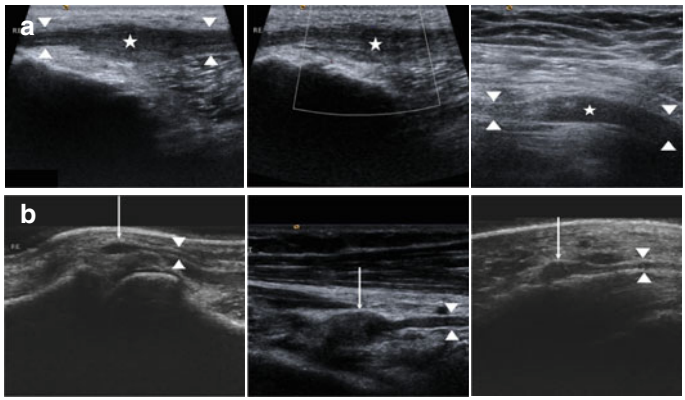

Fig. 6.7 Neuroma formation in HRUS. (**a**) Grade IV lesion with neuroma in continuity (*asterisk*) of the common peroneal nerve (*left*) and the femoral nerve (*right*). *Arrowheads*: nerve in continuity. Note the absent vascularisation. (**b**) Several grade V lesions with stump neuroma arising from the proximal nerve stump (*arrow*)

stump [64–66, 68, 69]. Examples are given in Fig. 6.7a, b. The differentiation with respect to smaller tumors of the peripheral nerve sheath can be challenging. This applies especially to a neuroma in continuity. Here, a detailed anamnesis survey must be used. If the history of a suitable injury is reported, it becomes more likely that the mass will not be a peripheral nerve sheath tumor.

As a complementary method, and particularly if deterioration or loss of nerve function is trauma related, high resolution sonography provides several important advantages in comparison to mere electrodiagnostic testing. First, the therapeutic relevant major lesions can be detected very early with an accuracy of 93 %, whereas electrodiagnostic testing alone provides an accurate statement whether reinnervation takes place (minor lesion) or fails (major lesion) still rather late [68]. By means of electrodiagnosis only, such an measurement is still obvious after a period of 2 or 3 months. However, patients with

major lesions will surely benefit from earlier surgical treatment because the progressing atrophy of the target organs ("time is muscle") can be stopped earlier [65, 66]. Second, in contrast to electrodiagnostic testing, high resolution sonography allows the exact localisation of the involved nerve segment [71]. This fact is especially important if traction injuries have occurred which might have affected the nerve at various points. For instance, electromyography studies cannot distinguish between a single and multiple serial lesions. Regardless, the extent of the electromyographic examination that is sometimes unpleasant to patients can thereby often be significantly reduced. Moreover, it is important that exact lesion localization helps to minimize surgical time and trauma.

Furthermore, HRUS can provide extremely helpful information on detecting the cause of postoperatively continuous and painful nerve irritation (e.g. scar tissue, bone fragments, callus, or pseudarthrosis in the neighborhood), and provides detailed answers to questions regarding the failure of electrodiagnostic testing. Finally, after nerve repair, mostly by means of nerve grafting, HRUS can depict the site of the fascicle coaptation. Surgical suture material appears as a hyperechoic nodule. At the site of the nerve adaptation, the development of a neuroma can re-occur and disturb axon sprouting. Rapid growth of these "suture neuromas" will result in an unfavorable outcome regarding functional nerve recovery, so the surgeon has to consider re-suturing [64–66, 68, 69, 71]. Figure 6.8a–e illustrates some of the above-mentioned conditions.

In several surgical institutions the high resolution ultrasound is also applied intra-operatively. The short distance between probe and nerve allows examination with the highest possible resolution. Thereby, important details like the rate of epineural or perineural fibrosis can be assessed which acts as an aid to differentiate the type of surgery required, microsurgical neurolysis or segment resection and grafting, or even a combination of both methods applied on separate nerve sectors [5, 72].

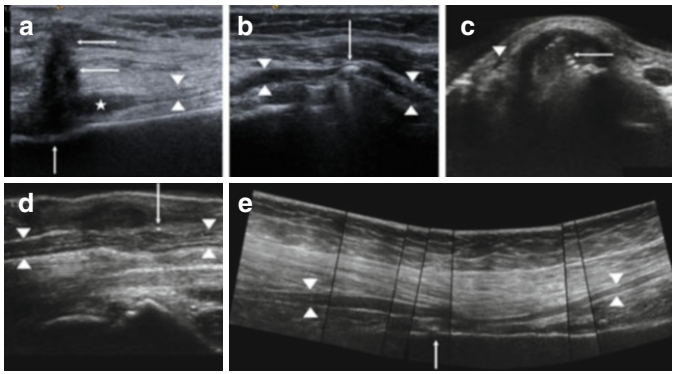

Fig. 6.8 Continuous post-traumatic nerve irritations in HRUS. (**a**) Scarring (*arrow*) in front of a stump neuroma (*asterisk*) of the superficial peroneal nerve (*arrowheads*) after chainsaw injury. (**b**) Cerclage (*arrow*) compressing the common radial nerve (*arrowheads*) at the upper arm. (**c**) Ostheosynthesis (*arrow*) affecting the tendons and the superficial radial nerve (*arrowhead*). (**d**) Artefact by sutures (*arrow*) after surgery of the median nerve (*arrowheads*). (**e**) Sciatic nerve (*arrowheads*) is compressed due to cerclage and reactive scarring (*arrow*)

6.1.7 Application to Ganglion Cysts, Nerve-Sheath-Tumors, and Other Intraneural Space-Occupying Lesions

As mentioned in Sect. 6.1.5, a peripheral nerve can be externally compressed or entrapped by a couple of soft tissue space-occupying lesions. With regard to surgical aspects of nerve tumors or tumor-like conditions, please refer to Chap. 13. Nevertheless, in order to offer complete continuity in our presentation of the principles of sonographic imaging and, furthermore, in order to give a comparative survey of imaging of all kinds of focal neuropathies (entrapment, traumatic lesions in continuity of different degrees, pseudoneuroma,

neuroma, neoplasms, inflammation pathologies), we continue with tumors. Surgical aspects will be presented in Chaps. 11 and 13.

According to our experience, when we apply high resolution sonography to special focal neuropathies which are tumor related, extraneurally located ganglion cysts are the most common space-occupying lesions we observe. From this point of view we want to describe briefly their typical sonographical findings here, although they are more often located outside the peripheral nerve. Ganglion cysts can arise from a joint or from a tendon sheath [73]. The appearance of ganglion cysts ranges from either simple to complex. A simple ganglion cyst is monolocular and characterized by an anechoic internal echotexture with a thin and well-defined margin, an absent vascularisation in colour Doppler, and a variable post-acoustic enhancement. Complex ganglion cysts may show septations, internal hyperechoic reflections as well as an irregular shape. They are usually larger than simple ganglion cysts. Echotexture may be hypoechoic, but sometimes with a partially solid component. Some show a colour Doppler flow, and nearly all have a post-acoustic enhancement [74–76]. Occasionally the communication with the joint or tendon is visible [74]. Since ganglion cysts have infrequent vascularisation, they may be differentiated by this fact from other mass lesions [74–76]. An overview of typical sonographical findings is presented in Fig. 6.9.

Among the different types of benign soft tissue tumors only 10 % originate from the peripheral nerve structure (nerve trunks, plexus or roots). They are also referred to as benign tumors of the peripheral nerve sheath. The most common are the schwannoma (synonym: neurinoma) and the neurofibroma [77]. A solid schwannoma usually affects a single fascicle only, whereas a solid neurofibroma arises from a fascicle group, a fact that helps to explain why schwannomas are easier to treat surgically as described in Chap. 12. Both mass lesions are characterized by a slow growth over years or decades. Therefore, neurological

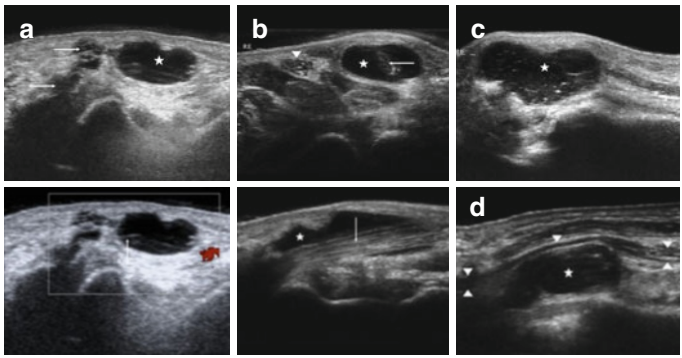

Fig. 6.9 Typical sonographical representation of ganglion cysts. (**a**) Ganglion cyst (*asterisk*) arising from the basic joint of the thumb (*small arrows*). Note the post acoustic enhancement. No Vascularization was not detected with the aid of colour Doppler. (**b**) Ganglion cyst (*asterisk*) arising from the tendon of the flexor carpi radialis muscle (*arrow*), *arrowhead*: median nerve. (**c**) Complex ganglion cysts with septations as well as internal reflections (*asterisk*). (**d**) Ganglion cyst (*asterisk*) compressing the ulnar nerve (*arrowheads*) at Guyon's canal

abnormalities appear very late. As an early symptom, a positive Hoffmann-Tinel sign and irradiating pain provoked during pressure at the site of the space-occupying lesion is noted. In neurofibromatosis type I multiple solid neurofibromas within a peripheral nerve may be observed. On the other hand, the rarely occurring plexiform neurofibroma is a special form of multiple manifestations of neurofibromas within one nerve trunk. Due to its net-like growth, gradually all fascicles are involved. Subsequently, early development of a sensorimotor failure followed by axonal loss as well as other severe neurological symptoms, particularly pain, is common in plexiform manifestations [78, 79]. Moreover, in patients with neurofibromatosis type 1, the risk of malignant transformation is enlarged [80]. The rare perineurioma shows a similar type of growth within a peripheral nerve [78, 79].

The typical songraphical presentation of solid schwannomas or neurofibromas is characterized by a peripheral nerve that centrically or eccentrically enters and exits a rather homogeneous hypoechoic ovoid or fusiform mass which has well defined margins and a post acoustic enhancement (Fig. 6.10a). In larger solitary neurofibromas and schwannomas, good vascularization may be observed with the aid of color-flow Doppler. Timely progressed schwannomas in particular form additional regressive signs such as hypoechoic cysts, calcifications, and larger hyperechoic areas [81, 82]. This is demonstrated in Fig. 6.10b–d. However, in a recent report it has been shown that a differentiation between schwannoma and

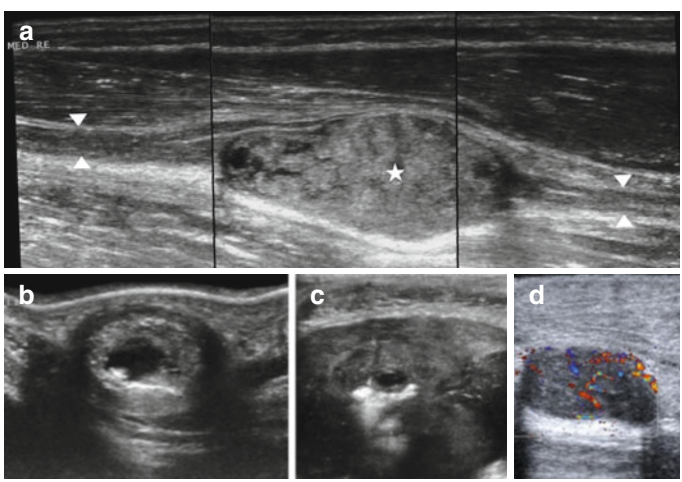

Fig. 6.10 Typical sonographical representation of a benign tumor of the peripheral nerve sheath. (**a**) Median nerve (*arrowheads*) enters and exits a schwannoma (*asterisk*) at the level of the forearm. Note the post acoustic enhancement. (**b**, **c**) Regressive changes in late shwannomas like cysts and areas of hyperechogenicity. (**d**) Well vascularized tumor of the peripheral nerve sheath

neurofibroma using HRUS is impossible. A more eccentrical growth may suggest a schwannoma [83].

Quite another severe challenge arises when differentiation between a plexiform growing tumor, a tumor-like lesion (see Sects. 13.3.3 and 13.3.4, and Chap. 14) or a focal manifestation of an immune-neuropathy (see Chap. 10) is sonographically required [84] (Fig. 6.11a). According to our experience, all of these completely different focal lesions can show a focal fusiform enlargement of the nerve and a hypertrophic-hypoechoic remodelling of its fascicles. For further detailed information on the various inflammatory nerve diseases that we have to take into

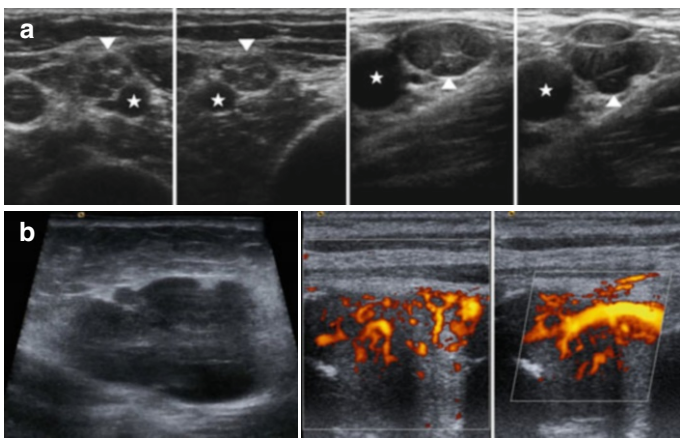

Fig. 6.11 Challenging differentiation between plexiform growing tumors of the peripheral nerve sheath (perineurioma, neurofibroma) and other neuropathies in HRUS. Malign tumors of the peripheral nerve sheath. (**a**) *Left* – hypoechoic swelling of the fascicles of the median nerve (*arrowheads*) in a case of Parsonage Turner syndrome at the upper arm. *Asterisk* shows the brachial artery. *Right* – perineurioma of the ulnar nerve at the axilla (*arrowhead*) shows a similar appearance. *Asterisk*: axillary artery. (**b**) Sonographical representation of a malign tumor of the peripheral nerve sheath. Note the anarchic vascular pattern, the irregular margins and the infiltrative growth

account when different treatment modalities are up for decision, refer again to Chap. 10. In the case of a plexiform neurofibroma, a history of a neurocutaneous syndrome (neurofibromatosis type I) is often but not always present, whereas in an inflammatory neuropathy, the fact that different nerves are clinically, subclinically (in EDX!), and sonographically affected can help. Finally, the more significant contrast agent uptake of a tumor in MR (magnetic resonance) neurography may support the differential diagnosis. However, sometimes a biopsy of nerve fascicle leads to a definitive diagnosis [84, 85].

Furthermore we may be very rarely confronted with a perineurioma which shows similar sonographical appearance as plexiform neurofibroma. On the other hand the very rare granular cell tumor presents similar to a late case of schwannoma with internal calcifications but irregular borders [82]. However, these are rarities.

Malignant tumors of the peripheral nerve sheath are equally rare. They are characterized by fast growth within some weeks or months and an early incidence of neurological abnormalities [78, 79]. Sonographically, an infiltrative growth, an irregular tumor surface, and an inhomogeneous echotexture are observed. The criteria mentioned above yield a sensitivity of 82%, but the specificity is only 38 % in HRUS. If the mass demonstrates good vascularisation and further characteristics such as anarchic vascular patterns, vessel trifurcations, vessel occlusions and stenosis, these findings can help to improve the sensitivity and specificity [86] (Fig. 6.11b). It has recently been made evident that contrast enhanced ultrasound (CEUS) enables differentiation between benign and malign soft tissue masses. Therefore, the type of perfusion pattern after administration of ultrasound contrast agent was used. The combination of three features (first, perfusion pattern of the mass either inhomogeneous or perfusion at the margins only, second, size >3.3 cm, and third, location below the superficial fascia) yields a sensitivity of 89 % and a specificity of 85 % as well as a positive predictive value concerning a malignancy of 86 % [3].

Among intraneural space-occupying lesions which do not originate from nerve tissue itself are rarities such as lipomas, fibrolipomas, haemangiomas [87–90] and tumor like lesions such as amyolidoma, lymphoma, sarcoidosis-lymphoma syndrome, leprosy or extramedullary affection of the peripheral nerve due to plasmacytoma. We will briefly discuss some of these in Chap. 14.

These lesions also include the uncommon solitary or multiple intraneural ganglion cysts. The peroneal nerve near the tibio-fibular joint is commonly affected, but several different locations in the neighbourhood of joints come into consideration such as, the tibial nerve in the tarsal tunnel. Due to degenerative and destructive alterations of joints, e.g. of the tibio-fibular or ankle joints, a dissection of the epineurium of the nerve branch supplying them occurs. As a consequence, a retrograde filling of the nerve with synovia is assumed [91]. Commonly, the patients suffer pain. Fluctuating weakness is more frequent [92]. Multicystic intraneural manifestations are then frequently to be detected sonographically, and they are characterized by a hypoechoic and string of pearls-like enlargement of the entire affected nerve segment. The fascicles and the perineurium can be clearly identified as “floating” or “compressed” in the synovial fluid, and 18 % of the non-traumatic lesions of the peroneal nerve are said to be caused by intraneural ganglion cysts [93]. Figure 6.12 illustrates these rare findings. Surgical treatment of these intraneural manifestations is quite different if compared to the extraneural ganglia described above (see Sect. 13.3 8) [91].

6.1.8 Application to Polyneuropathies

It has long been known that multifocal palpable nerve enlargements exist in some types of hereditary motor and sensory neuropathy. It is therefore not a surprise that these nerve

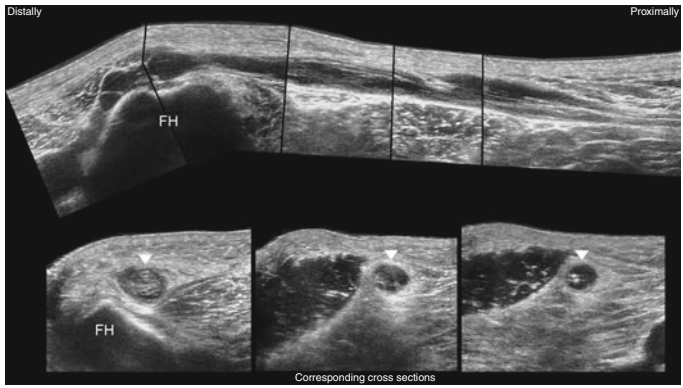

Fig. 6.12 Typical sonographical representation of multiple intraneural ganglion cysts of the common peroneal nerve (*arrowheads*). *FH* fibular head. The segmental filling of the nerve segment with synovia is clearly visualized

enlargements can be detected with high resolution ultrasound. This holds true especially for two hereditary neuropathy types, first, the hereditary motor and sensory neuropathy types Ia and Ib and, second, the hereditary neuropathy with liability to pressure palsies. In sonography we find generalized or multifocal nerve enlargements with a focal increase of the cross sectional area, hypoechoic fascicular swelling or even fascicular masking in different peripheral nerves [2, 94, 95] (Fig. 6.11a).

Furthermore, some immune neuropathies show similar alterations: among them are the acute inflammatory demyelinating neuropathy, the chronic inflammatory demyelinating neuropathy (CIDP) and its variants and the multifocal motor neuropathy (see also Sect. 10.5). Typically, not only peripheral nerve segments are involved, but additionally cervical roots and the supraclavicular brachial plexus (Fig. 6.13). If we want to find the differential diagnosis it is important that nerve enlargements are often

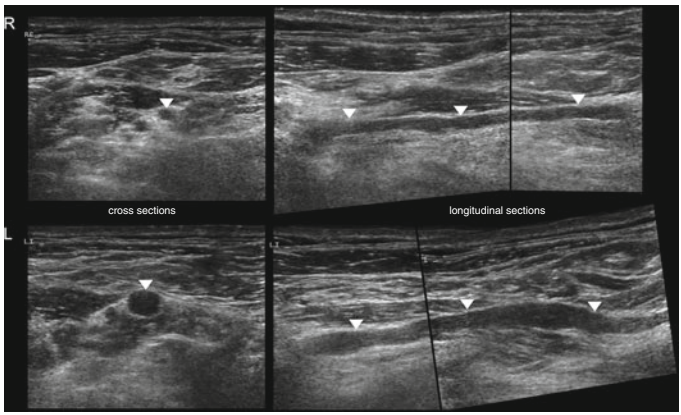

Fig. 6.13 HRUS, segmental enlargement of the root C5 (*arrowheads*) at the affected side (*left*) in comparison to the unaffected side (*right*) in a patient with multifocal motor neuropathy

observed far away from the site of the perhaps expected typical entrapment site, e.g. carpal tunnel, ulnar groove, supinator tunnel. It is interesting that a correlation exists between the sonographically diagnosed segmental nerve enlargements and the conduction blocks registered by electrodiagnostic testing [96–100].

In a recent report nerve enlargement was commonly diffuse and generally more than twice normal size in subjects with hereditary motor and sensory neuropathy type I (CMT-1) but not in immune neuropathies which showed no, mild or regional nerve enlargement [101]. Several abnormalities have been described regarding further types of polyneuropathies. However, no special characteristics were reported to allow a clear distinction between their different types by means of ultrasound imaging. However, there was evidence that sonographically detectable alterations can be observed before clinical and electrophysiological manifestations occur [100].

6.1.9 Application to Ultrasound Guided Nerve Blocks

In regional anaesthesia, ultrasound guided nerve blocks have been in use for a longer period [102]. It will have to be assessed whether this will further lower the rate of complications which is already very low. In 3% of cases, transient neurological abnormalities occur, and permanent nerve damage is rather rare [103]. In nerves with a high rate of anatomical variability, such as the lateral cutaneous femoral nerve, the success rate of the block can be dramatically increased with ultrasound guidance [104]. Furthermore, less time elapses before onset of the block and its effect lasts longer, the consumption of local anaesthetics is lowered, and finally, the risk of iatrogenic vascular puncture with a damaging hematoma is decreased [105, 106].

The ultrasound guided puncture is applicable in the conservative management of focal painful neuropathies such as meralgia paresthetica or Morton's metatarsalgia (see Sects. 9.2 and 9.11). Moreover, pain management with ultrasound guided phenol injections in stump neuromas were successfully carried out [107].

6.1.10 Secondary Alterations of the Muscle Following Nerve Damage

Axonal loss leads to a denervation of the muscle substance. In its early stage the reaction is characterized by an oedema of muscles fibers. Ultrasound images reveal them as hyperechoic (Fig. 6.14a). Furthermore, even muscle twitching (fasciculations) can be observed. If no regeneration follows nerve injury muscular atrophy starts. This process is characterized by remodelling of muscle tissue with connective and fatty tissue.

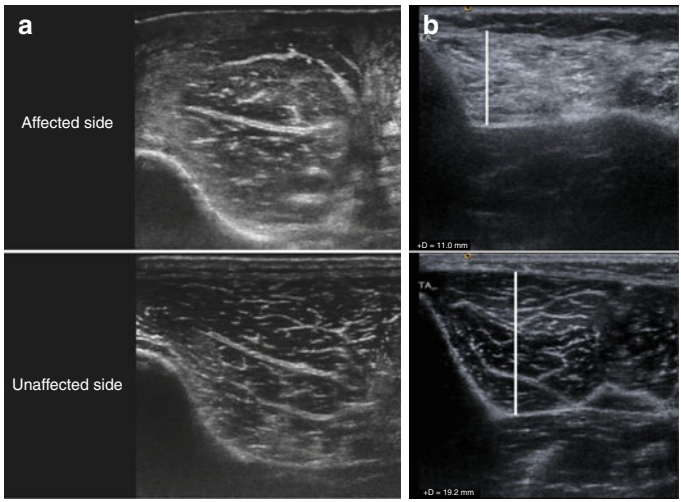

Fig. 6.14 HRUS, secondary alterations of the muscle following nerve damage. (**a**) Early stage with oedema. (**b**) Late stage after transformation by connective tissue and decreased muscle diameter (tibialis anterior muscle). Note the hyperechoic appearance in both cases

The hyperechoic appearance of the affected muscle remains, however, as the muscle diameter decreases [108]. Figure 6.14b illustrates an example.

6.1.11 Conclusions

High resolution ultrasound provides important additional information on the entire aspect of varying occurrences of focal neuropathies, e.g. nerve abnormalities, irritating structures, entrapment dependent on functional limb positions, different intraneural tissue reactions, extra- or intraneural space-occupying lesions, intramuscular reactions and differential-diagnostically

important inflammatory diseases. Continuous examination of the entire nerve is possible as well as its dynamic assessment during active and passive limb or muscle movement. There are no side effects and no contraindications. Children can be examined pain free. The method is increasingly more freely available and less cost- and time-consuming than MRI. Limitations only result from deeply situated nerves, obese patients and the phenomenon of acoustic shadowing: osteosynthesis material, hematoma and scar tissue formation can hide small nerve structures from detection. Furthermore, of course, it is not to be neglected that the method depends on the learning curve and the final experience of the examiner. In a very recent study the diagnostic values of MRI vs nerve ultrasound regarding the contribution to the differential diagnosis in mononeuropathies and brachial plexopathies were compared. As a result, ultrasound was valued as more sensitive than MRI (93 % vs 67 %); both techniques yielded an equivalent specificity (86 %) [109]. Additionally, HRUS could better identify multifocal lesions. Therefore authors suggested using HRUS as the preferred initial imaging modality for suspected peripheral nerve pathology [5].

6.2 Magnetic Resonance Imaging (MR Neurography)

6.2.1 Introduction and Technical Notes

While high resolution ultrasound techniques were increasingly applied in the clinical routine, magnetic resonance imaging techniques came to the fore for optimizing the evaluation of peripheral nerve disorders [110, 111]. With the new generation of high-field (3 T) MR scanners and appropriate multichannel

phased array surface coils, not only mass lesions of the peripheral nerve sheath but also entrapment neuropathies, peripheral nerve injuries and generalized neuropathies can be evaluated [112, 113]. However, 3 T MR is neither freely available nor cost effective at the moment.

In 1989 it was first described in an animal model that axonal damage of the tibial nerve in rats is followed by increasing signal intensity on T2-weighted axial MR images [114]. A few years later, along with common T1-weighted axial images, the introduction of new T2 imaging sequences – characterized by high fluid sensitivity, homogeneous fat saturation, a strong T2 weighting such as "spectral adiabatic inversion recovery (SPAIR)" and "short-tau-inversion-recovery (STIR)" – allowed a much more detailed estimation of pathologically altered nerve microstructures. This also applies to "proton density weighted (PD)", fat saturated images. Moreover, it has been shown that administration of MR contrast agent on fat saturated T1-weighted images (e.g. "T1 spectral presaturation with inversion recovery, SPIR") could generate additional information if mass lesions or inflammation were suspected. Therefore, a new term referred to as "MR neurography" has been introduced [112, 113, 115]. Beside the high resolution, two-dimensional (2D) sequences obtained from 3 T MR equipment mentioned above and three-dimensional anatomic nonselective (3D) sequences of peripheral nerve anatomy and pathology are currently state of the art in MR neurography protocols [116]. These 3D sequences provide an increased contrast resolution, improved fat suppression, and less image distortion [117, 118]. They are termed differently by the vendors [116]. Three-dimensional sequences allow multiplanar reformats (MPR), although their in-plane resolution is less than in high resolution 2D sequences [116]. A very recent advance was achieved by the introduction of "diffusion-weighted MR imaging (DWI)" that is useful to suppress structures with unwanted high signal intensity adjacent to nerves both on fat-saturated T2- and T1-weighted

images, such as veins [116, 119, 120]. Another novel technique in peripheral nerve imaging is "diffusion tensor imaging (DTI)" well known from brain and spinal cord studies. Beside the 3D nerve visualisation, DTI allows the calculation of "apparent diffusion coefficient (ADC)" and "fractional anisotropy (FA)". "ADC characterizes molecular diffusivity under motion restriction and FA represents the directional variability in the diffusion [116]". In animal model trials decreased FA values were more correlated to axonal loss than to demyelination and increasing FA values reflected nerve recovery [122]. Therefore, this novel technique may be applied to evaluate axonal sprouting in future. Another new diagnostic approach is to apply DTI on patients with carpal tunnel syndrome. Mainly they showed increased ADC values (diffusivity) and decreased fractional anisotropy (FA) of the median nerve compared to healthy individuals and successful carpal tunnel release decreased the ADC values but not FA [123]. Thereby, DTI may also play an important role evaluating failed carpal tunnel surgery in future. A further application of ADC values will be the differentiation between malign and benign peripheral nerve sheath tumors. Higher ADC values are seen with benign peripheral nerve sheath tumors, while low ADC values are more common in malignant space-occupying lesions [124]. Finally, further advances can be expected by means of new contrast agents [125], nerve segmentation, and magnetisation transfer (MT) imaging. For more details please refer to specialist literature [116].

6.2.2 Technical Requirements and Examination Sequences

As stated above, in addition to a current 3-Tesla high field MR scanner with appropriate multichannel phased array surface coils, special examination sequences are needed for the correct

Table 6.1 3 T MR extremity neurography examination protocol according to [126] with permission from SPRINGER

MR sequence	FOV (cm)	Slice thickness (mm)	TR/TE (ms)
Axial T1 TSE	15–20	3	550/7.9
Axial T2 SPAIR	15–20	3	2,840/67
Coronal PD	15–20	3	4,100/37
3D SPAIR SPACE	15–20	Isotropic (0.5–1 mm)	1,020/91
3D DW-PSIF	15–20	Isotropic (0.9 mm)	10/5

MR magnetic resonance, *FOV* field of view, *TR* repetition time, *TE* echo time, *TSE* turbo spin echo, *SPAIR* spectral adiabatic inversion recovery, *PD* proton density, *SPACE* single slab three-dimensional sequence with variable excitation pulse, *3D DW-PSIF* three-dimensional diffusion weighted reversed fast imaging with steady state precession (fat-suppressed sequence with a diffusion moment, b value of 80–90 s/mm^2)

representation of peripheral nerves in physiological and pathological conditions.

In our institution we use 2D high resolution axial images as follows: T1-weighted turbo spin echo sequences for the representation of the anatomy of the peripheral nerve, T2-weighted sequences with fat saturation (STIR, SPAIR) and proton density weighted sequences for the detection of abnormal nerve hyperintensity, and finally, fat saturated T1-weighted sequences (SPIR) if administration of contrast agent is required. Together with the special 3D imaging sequences mentioned above, "multiplanar isotropic reformats (MPR)" and "maximum intensity projections (MIP)" become available. Tables 6.1 and 6.2 demonstrate two examination protocols for brachial plexus and nerves of the extremities according to Chhabra et al. [126].

In cases where we do not have a clear picture of the localization of the pathological process, a two-step examination is recommended, primarily with a large "field of view (FOV)", whereas the target zone is examined afterwards with a small FOV [126]. We try first to localize the process exactly using

Table 6.2 3 T MR neurography examination protocol for the evaluation of the brachial plexus according to [126] with permission from SPRINGER

MR sequence	FOV (cm)	Slice thickness (mm)	TR/TE (ms)
T1 coronal both sides	30	4	881/11
3D STIR SPACE both sides	30	0.5–1	1,500/97
3D T2 SPACE	25	0.5–1	1,500/97
T1 sagittal affected side	22–24	3	804/8
STIR sagittal affected side	22–24	3	5,210/18
Axial T1 TSE	25–30	3	550/7.9

MR magnetic resonance, *FOV* field of view, *TR* repetition time, *TE* echo time, *STIR* short tau inversion recovery, *SPACE* single slab three-dimensional sequence with variable excitation pulse, *TSE* turbo spin echo

high resolution ultrasound if possible. After ultrasound guided labelling of the target process the FOV may be dimensioned as small as possible.

As stated above (high resolution ultrasound) the interpreter of 3 T MR images should have an excellent knowledge of the normal and pathological cross sectional anatomy of the peripheral nervous system and its variations.

In contrast to high resolution ultrasound, there are only a few systematic studies providing normal values of nerve diameter, cross sectional area, T2 hyperintensity, ADC and FA in the different peripheral nerves. In particular, if the abnormalities are subtle there is a need to compare findings of the affected side with the unaffected side in order to interpret the image correctly [126].

6.2.3 Normal and Pathologically Changed Peripheral Nerve on MRN

Similar to high resolution ultrasound, depiction of a single axon with its endoneurial sheath is not feasible with 3 T MRN; however, whole nerve fascicles, situated in the perineural sheath

and the surrounding fat planes, can be visualized in nerves approaching a diameter of 3 mm and above [112, 127].

Size, course, fascicular pattern, and perineural fat planes are best assessed on axial (2D) T1-weighted images. Moreover, T1-weighted images are appropriate to differentiate between peripheral nerves and vessels. Arteries are characterized by a special flow void signal, whereas veins are to be identified as T1 hyperintense due to the inflow phenomenon. The size of the peripheral nerve – without any focal or diffuse hypertrophy – is similar to the accompanying arteries. The normal fascicular pattern, similar to a honeycomb, is preserved without any enlargement, disruption, or masking of single or multiple nerve fascicles. The entire course of the nerve appears smooth without focal deviations or discontinuity and with clearly visible perineural fat planes [126]. On the other hand, focal or diffuse nerve enlargement, exceeding the size of adjacent arteries, enlargement, effacement, or disruption of the fascicles with loss of the normal honeycomb-like appearance in cross sections, nerve deviations, discontinuity, effaced perineural fat planes, epineurium thickening are useful signs suggesting neuropathy [126].

Normal peripheral nerves (and fascicles) remain isointense to the skeletal muscle on both T1 and T2 (SPAIR) weighted images, whereas they can appear with a mild hyperintensity on STIR images [126]. As mentioned above, comparison with the contralateral side or with neighboring nerves may help to identify them as a normal or variant [128]. Apart from this, a peripheral nerve which shows hyperintensity more than the skeletal muscle on T2-weighted images and which approaches or exceeds the signal intensity of the adjacent vessels is considered pathological. According to Chhabra et al. (2013), three stages of nerve hyperintensity have been established [112]":

- Mild hyperintensity, which slightly exceeds the signal of skeletal muscles, is nonspecific and therefore needs to be correlated with symptoms of neuropathy

- Moderate hyperintensity, which approaches the surrounding in plane signal of adjacent veins, pathological
- Severe hyperintensity, which is similar or exceeds the signal of adjacent vessels, pathological"

Normal nerves, except of dorsal root ganglia, which are outside the blood-nerve barrier, do not show any enhancement after administration of intravenous contrast agent (gadolinium) on fat saturated T1-weighted images (e.g. T1 SPIR). In contrast, impairment of the blood-nerve barrier caused by peripheral nerve sheath tumors usually leads to a contrast uptake on T1 SPIR [126]. In recent literature is stated that peripheral nerves affected by infection/inflammation also enhance contrast uptake [112]. However, especially in autoimmune neuropathies, there is a striking lack of systematic studies regarding the time course of contrast uptake with contradictory disclosures. According to our own experience, and in agreement with reports from others, we sometimes found post-contrast enhancement in immune neuropathies, and sometimes we did not [84, 129–131] (for details refer to Sects. 10.5.1 and 10.5.2). On the other hand there is stated that entrapped or injured nerves in a subacute stage show usually no contrast uptake [112, 126]. However we found contrast uptake in some post-traumatic neuroma.

Pitfalls can arise regarding the correct interpretation of the MRN images: The most important artefact which may mimic a mild T2 lesion according to Chhabra (2013) [112] is the "magic angle artefact (MA)" both on PD- and T2-weighted images. "It is confined to regions of tightly bound collagen at approximately 55° from the main magnetic field. Thus the peripheral nerve appears most hyperintense in the case of an angular deviation of 55° of its main longitudinal axis relative to the field direction of the main static magnetic field". This applies especially to the supraclavicular brachial plexus and the proximal common peroneal nerve [132]. However, it has been shown that by using 3 T MRI confusion with a moderate or severe T2 lesion is unlikely

[133]. Another pitfall may lead to the presence of small vessels (veins) outside and inside the perineurium, since they may have a similar T2 intensity to (moderate or severe) pathologically altered nerves. Their winded course as well as the entry into or exit out of the nerve allows the correct discrimination from a peripheral nerve lesion. In any doubt, administration of contrast agent on T1-weighted fat saturated images may help to differentiate due to the strong contrast enhancement of vessels [132].

6.2.4 Specific Conditions

Despite the increasing number of published articles about MRN at PUBMED there is still a striking lack of systematic prospective studies in this field of peripheral nerve imaging. A lot of findings are based on retrospective observations or case reports. Moreover, the diagnostic value of the recently described imaging techniques (DWI, DTI) remains unclear at the moment. Therefore, we intend to describe only practically relevant pathological conditions of the peripheral nerve with sufficient scientific evidence in the following section. Nevertheless, as stated above, new developments could further improve the diagnostic power of MRN.

6.2.5 Application to Entrapment Neuropathies

As in high resolution ultrasound, there are some basic commonalities in nerve entrapment neuropathies when examined by MRN. The nerve segment just proximal and at the site of the entrapment develops a focal increasing hyperintensity of nerve fascicles on T2-weighted images (Fig. 6.15). The single fascicles may be enlarged or effaced on T1-weighted images. These alterations additionally lead to an increased cross sectional area

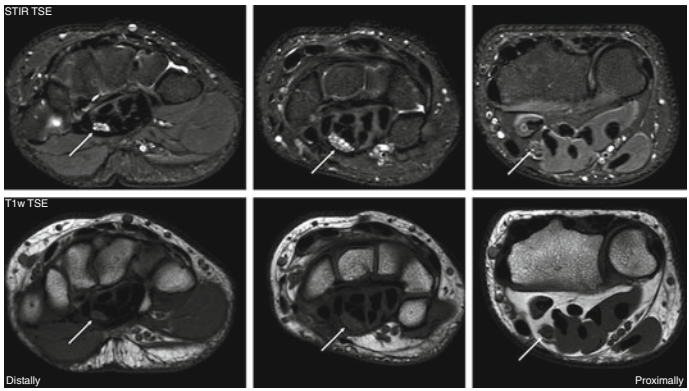

Fig. 6.15 MRI in carpal tunnel syndrome. *Small arrows*: median nerve. Note the hyperintensity at and just above the site of compression (*left* and *middle*) on STIR images. The images at the right side show the normal nerve at the forearm

of the nerve segment proximally to the site of the entrapment and to a flattening at the site of compression (Fig. 6.16) [134]. Focal nerve enlargement and T2 hyperintensity occurs due to various hypothesized mechanisms, such as impairment of axoplasmic flow, venous congestion with oedema or demyelination [110, 111]. In an advanced disease, a perineural fibrosis can be seen on T1-weighted images as a disruption in the normally smooth contour of the perineural fat plane [134]. According to our own experience, these late stages are often accompanied by axonal loss. Due to Wallerian degeneration, particularly, the distal nerve segment may become hyperintense over a long distance, in accordance with experimental studies in animals [67]. As mentioned above, administration of contrast agent is usually not necessary if an entrapment neuropathy is suspected.

Carpal tunnel syndrome: In contrast to ultrasound studies, there are only a few reports regarding normal values in CTS. The upper limit of normal median nerve CSA was 14.4 mm^2 in one study and the mean CSA of the median nerve was 11.8 mm^2

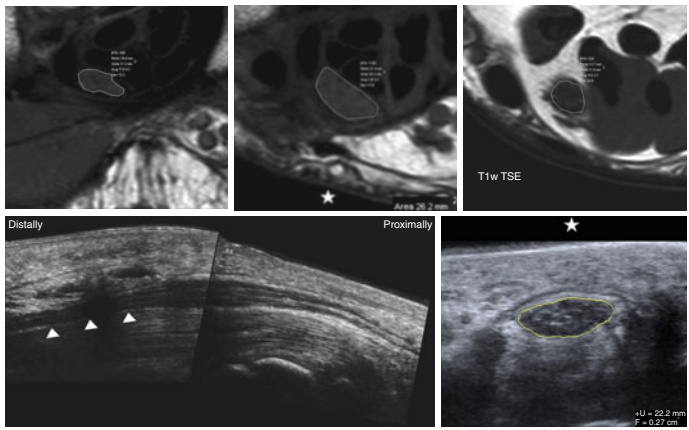

Fig. 6.16 Enlargement of the cross sectional area proximally to the site of compression on T1-weighted images and the corresponding high resolution ultrasound images (*asterisk*). *Arrowheads*: site of compression

in another study – higher than normal limits obtained from ultrasound studies [26, 135, 136]. As stated above, besides nerve CSA enlargement a second important characteristic of a nerve entrapment syndrome is the increased T2 hyperintensity on T2-weighted images affecting the nerve segment proximal to the site of compression. However in older studies, T2 hyperintensity alone was of low specificity (<40 %) what holds also true for the sensitivity and specificity of different other MR signs of carpal tunnel syndrome [137–138]. Latest reports provide normal values using diffusion tensor imaging (DTI) [139]. As mentioned above, patients with CTS are characterized by an increased ADC and decreased FA. Clinical improvement after surgery was reflected in a decreasing diffusivity (ADC), but not in anisotropy [123].

Ulnar neuropathy at the elbow (*UNE*): In a recent study the mean ulnar nerve size in symptomatic individuals was 0.12 cm^2 and 0.06 cm^2 in controls. When using a cut-off value of 0.08 cm^2, sensitivity/specificity yielded 95 % versus 80 %.

Moreover, the mean relative T2 signal intensity was increased in patients with ulnar neuropathy at the elbow [140]. Another report stated that T2 signal increase seems to be an accurate sign to determine the presence of UNE, and that nerve caliber enlargement may be used to discriminate severe from mild UNE with a resulting sensitivity of 83 % and specificity of 85 % after qualitative assessment [141].

Apart from what is stated in the chapter about high resolution ultrasound (Sect. 6.1.5), unfortunately, there are only case reports or small prospective or retrospective case series regarding MRN applied on rare entrapment neuropathies. In Guyon's canal syndrome (GCS), it has been recently shown that the T2-signal increase of distal ulnar nerve branches and, in particular, of the deep-motor branch is very accurate in the diagnosis of GCS, yielded a specificity of 90 % and a sensitivity of 89.5 %. These results correlated nonlinear with the motor-conduction-velocity to the first dorsal-interosseus muscle [142]. Due to the lack of systematic studies and the rarity of cases, no values of normal or pathologically changed nerve size, T2 hyperintensity etc. in other rare entrapment neuropathies currently exist. Diagnosis should be based on the qualitative MRN characteristics of nerve entrapment listed above. A more detailed overview is given in specialist literature [134, 143–145].

Similar to the high resolution ultrasound findings, MRN can provide additional information about anatomical nerve variations, underlying pathology such as entrapment by tenosynovitis, ganglion, hematoma, lipoma, or other space-occupying masses, and reasons for failed surgery [134].

6.2.6 Application to Nerve Injuries

For a detailed description of Sunderland's classification specifying different degrees of peripheral nerve injury and further theoretical details, please refer to Sects. 3.1 and 3.2 again.

In general, MRN exhibits analogous advantages to HRUS in patients with a traumatic nerve lesion [146, 147]:

- It may differentiate between minor lesions (Sunderland I–III) and major lesions (Sunderland IV and V), and it can demonstrate neuroma formation
- It localizes the lesion very accurately
- It may allow an assessment of the underlying pathology (damage by osteosynthesis, hematoma, or scar-tissue, etc.)

However, MRN may be challenging if there is metallic hardware in the area of injury [146].

Slight grade I lesions according to Sunderland may be difficult to see on MRN. In animal studies it has been shown that the contrast agent gadoflurine has the capacity to visualize these grade I lesions [125]. However gadoflurine is not approved for human use. According to recent literature [142], and similar to pronounced demyelinising entrapment, a focal nerve enlargement with accompanying T2 lesion might be seen. This has little significance because grade I lesions have a favourable outcome and tend to a spontaneous recovery.

However, there exists a systematic animal study which demonstrates the intraneural alterations following acute nerve traction injury to the sciatic nerve in rats, an injury that represents a grade II/III lesion according to Sunderland (peri- and epineurium preserved, endoneurium/axons as structure partially damaged). Notably at the site of the injury and less in the distal nerve segment, first, a hyperintensity of the fascicles on T2-weighted images, and, second, a nerve enlargement starting at the first day and peaking 7 days after injury were observed. The proximal portion of the injured nerve demonstrated only moderate signals in the same manner. Histological studies and electrodiagnostic testing could demonstrate that these findings represented the Wallerian degeneration. After nerve regeneration (re-innervation) these signals disappeared first in the proximal nerve segment, later in the distal nerve segment, and

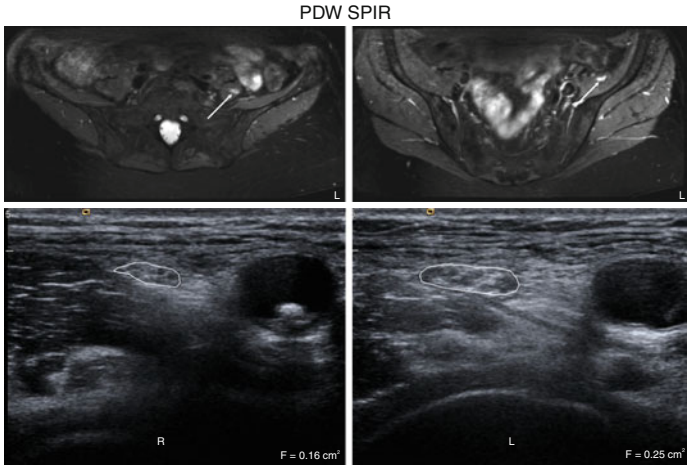

Fig. 6.17 Grade II/III lesion of the femoral nerve (*small arrow*) at the left side after traction injury with hyperintense lesion on PDW-SPIR images (*above*). Below the corresponding high resolution ultrasound images with preserved echotexture but enlarged cross sectional area and fascicle oedema at the *left side* compared to the healthy *right side*

finally at the site of the injury [67]. Based on our own clinical experience and published data from further authors, identical findings are to be seen in humans [146, 147]. An example is shown in Fig. 6.17.

Grade IV lesions are distinguished by a disruption of individual fascicles; they have the same MR-neurographic characteristics as grade II/III lesions as long as the continuity of the nerve is preserved [146, 147].

In the case of the most severe lesion (a grade V lesion), the nerve continuity is interrupted, and a gap occurs due to retracted nerve stumps. The hyperintense T2 signal turns into a hypointense T2 signal, representing the gap [146, 147].

Without previous surgical treatment in cases of grade IV/V lesions, neuroma formation is always observed. There are two neuroma types: the neuroma in continuity (in grade IV lesions)

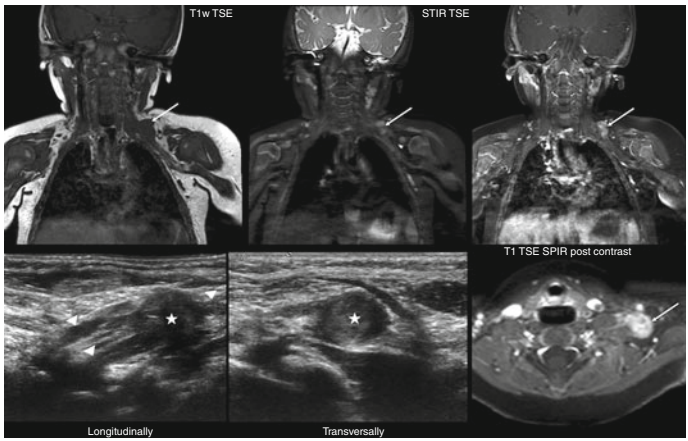

Fig. 6.18 Grade IV lesion with a neuroma in continuity of the brachial plexus after a childbirth injury in a 6-month-old girl. Note the hyperintense lesion on STIR images and the irregular contrast uptake on T1 SPIR images. *Small arrows*: neuroma in continuity. At the corresponding ultrasound images it is clearly seen that the cervical roots C5 und C6 (*arrowheads*) enter the neuroma (*asterisk*) and that a further part of the brachial plexus exits the neuroma (*arrowhead*)

and the stump neuroma (in grade V lesions). The neuroma in continuity is characterized by a baseball-shaped mass with nerve continuity on either side, similar to the appearance of peripheral nerve sheath tumors. If the nerve continuity is disrupted, a baseball-shaped end bulb or stump neuroma appears. All types of neuroma demonstrate a hyperintensity on T2-weighted images [147]. Some authors state that a neuroma is characterized on MRN by the presence of surrounding scarring, lack of a split fat or target sign, and by the absence of abnormal enhancement [146, 147]. However, we have often observed an irregular abnormal contrast enhancement in traumatic neuromas (Fig. 6.18).

As initially mentioned, diffusion tensor imaging (DTI) may play an important role in the evaluation of the extent of axonal damage and axonal regeneration in injured nerves in future [116].

6.2.7 *Application to Ganglion Cysts, Peripheral Nerve Sheath Tumors, and Other Intraneural Space-Occupying Lesions*

As already mentioned in Sect. 6.1.7, extra- and intraneural ganglion cysts and the slowly growing benign schwannomas and neurofibromas occur most commonly. Multilocular tumor manifestation can frequently be associated with neuro-cutaneous symptoms [148].

Ganglion cysts are mass lesions with well-defined margins and unilocular or multilocular shape (see also Sect. 13.3.8). On fat-suppressed T2-weighted image sequences, they are hyperintense and show septae in two cases out of three. On fat-suppressed T1-weighted images, post-contrast enhancement of the cyst wall can be demonstrated [149]. An intraneural ganglion is characterized by a filling of the affected nerve segment with synovia (Fig. 6.19) . A communication with a joint or a tendon sheath as source of the cyst can often be seen [150].

Benign tumors of the peripheral nerve sheath appear as ovoid, fusiform or rather round masses and have well-defined margins (Fig. 6.20). The nerve that acts as originating structure enters and exits the mass (see also Sects. 13.3.1 and 13.3.2). On T2-weighted images it demonstrates low signal intensity in the center and high signal intensity at the periphery. This typical sign is referred to as the “target sign”; it can also occur with multiple rings – known as the “fascicular sign”. Another typical sign is the “split fat sign”, seen on longitudinal T1-weighted images when the mass is surrounded by a layer of fat (Fig. 6.21). Benign nerve sheath tumors may present a variable or homogeneous enhancement on post-gadolinium imaging [151].

Unfortunately, these signs declared as typical may also be variably seen in malignant peripheral nerve sheath tumors. Malignancy is strongly suspected in the case of a fast growing mass with an early onset of pain and neurological abnormalities.

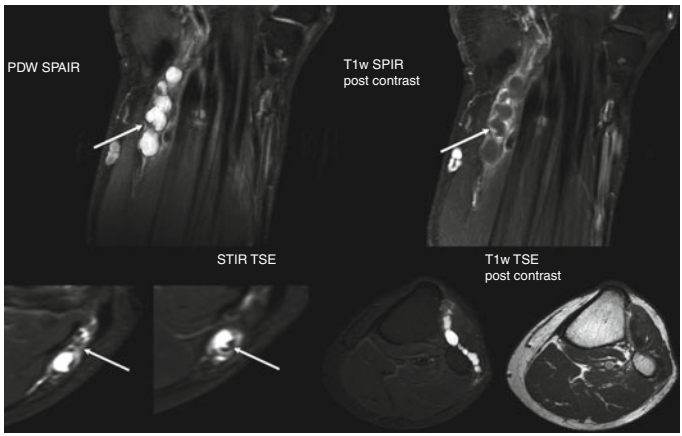

Fig. 6.19 Ganglion cyst (*small arrows*) of GUYON's canal (*above*). Note the hyperintensity of the cyst presenting septae on PDW images; furthermore, the contrast uptake of the wall of the cyst on T1 fat-suppressed post gadolinium images. *Below*: an intraneural ganglion cyst of the common peroneal nerve is shown with the same characteristics mentioned above. Note the hypointense fascicles "floating" in the hyperintense synovia (*small arrows*)

Signs of malignancy in MR findings are ill-defined or invasive margins, peritumoral oedema, and a diameter over 5 cm together with heterogeneous signal intensity on T1- and T2-weighted images [151–153]. If using modern DTI, benign peripheral nerve sheath tumors are characterized by higher ADC values ($1.1–2.0 \times 10^{-3}$ mm²/s), whereas malign peripheral nerve sheath tumors show low diffusivity values ($0.7–1.0 \times 10^{-3}$ mm²/s) [124]. Chhabra et al. 2013 [116] currently use three cut-offs in the clinical routine complementary to conventional imaging":

- ADC $\leq 1.0 \times 10^{-3}$ mm²/s → suspicious for malignancy
- ADC = $1.0–1.5 \times 10^{-3}$ mm²/s → indeterminate
- ADC $\geq 1.5 \times 10^{-3}$ mm²/s → usually benign"

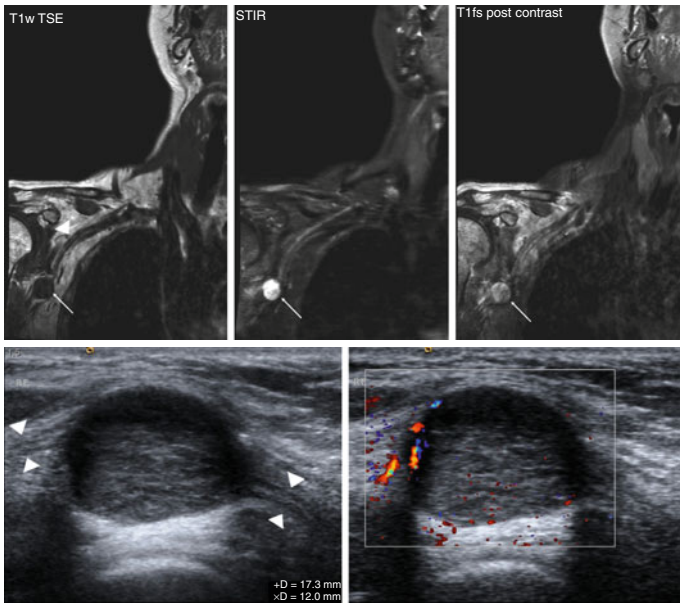

Fig. 6.20 Schwannoma of the radial nerve trunk at the axilla. Note the hyperintense lesion on STIR images and the contrast uptake on T1 fat-supressed images. The nerve enters and exits the mass lesion. *Below*: corresponding ultrasound images. *Arrowheads*: common radial nerve

Only the lowest ADC values, reflecting the cellular portion of the lesion, should be considered in the analysis. Higher ADC values may also be present in malignant peripheral nerve sheath tumors, representing necrotic or less cellular portions. However, it remains to be seen whether in the future DTI adds more value over contrast imaging [116].

A real diagnostic challenge is the differentiation of plexiform growing tumors like perineurioma, plexiform neurofibromas (Sects. 13.3.3 and 13.3.4) or tumor-like conditions such as amyloidoma (Chap. 14) from an immune neuropathy (Sect. 10.5).

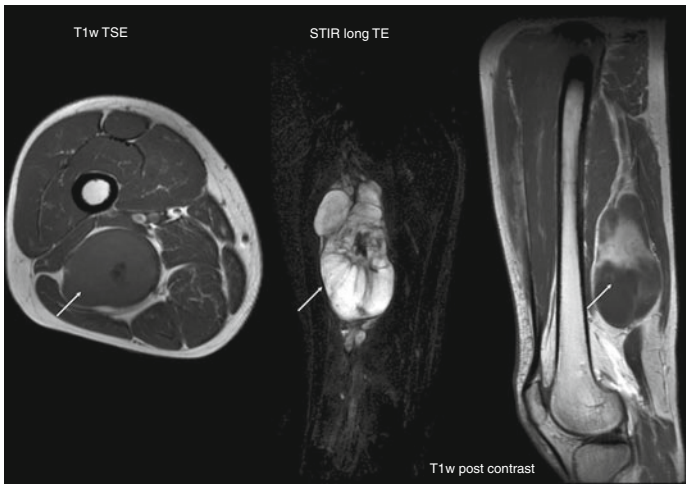

Fig. 6.21 Neurofibroma (*small arrows*) of the sciatic nerve in MRI. Note the split fat sign (*left*) and the target sign (*middle*). *Right*: sciatic nerve enters and exits the mass lesion. Characteristic are well-defined margins and an irregular contrast uptake

Rather frequently, biopsy of nerve fascicle leads to a definitive diagnosis only [84, 85]. All of these lesions may show a fusiform nerve enlargement on T1-weighted images with effaced or enlarged nerve fascicles as well as a T2 hyperintensity on appropriate MR sequences and a post contrast enhancement [154]. Advanced functional diffusion tensor imaging will probably better characterize the full extent and the internal architecture of these lesions in future [155–157]. An example is given in Fig. 6.22.

6.2.8 Application to Polyneuropathies

There is a striking lack of systematic studies regarding MRN findings in nerve disorders such as inflammatory, hereditary and

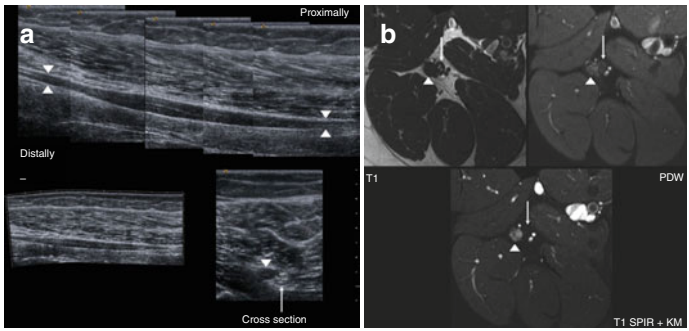

Fig. 6.22 Fusiform nerve swelling – diagnostic challenge: 19-year-old female with a 4-year history of progressive palsy of the left common peroneal nerve. (**a**) On ultrasound images: a high division of the sciatic nerve at thigh-level with a hypoechoic, fusiform swelling of the peroneal part (*arrowheads*) approaching a length of 70 mm and a normal appearance of the tibial part (*arrow*). (**b**) The same findings on MRN images: the peroneal part (*arrowheads*) shows a swelling and effacement of its fascicles on T1-weighted images, a hyperintensity on PDW images and strong gadolinium uptake on T1 SPIR images, whereas the tibial part appears normally (preserved fascicles with no enlargement on T1 images, no hyperintensity on PDW images and no contrast uptake on T1 – SPIR images). Fascicular biopsy revealed a perineurioma

other types of neuropathies. Until now, diagnosis is still mainly based on the anamnesis survey, clinical examination, electrodiagnostic testing as well as laboratory findings. MR neurography can demonstrate the abnormal nerves or nerve roots as focally or generalized diffusely hyperintense on T2-weighted images with or without focal or generalized nerve enlargement on T1-weighted images [151, 154]. Generally no contour changes are observed [151]. In some cases of chronic inflammatory demyelinating neuropathy, a focal nerve enlargement and a hyperintensity on T2-weighted images at the site of the conduction block was found, although only in 50 % with gadolinium uptake [130]. Some authors, however, reported a significant

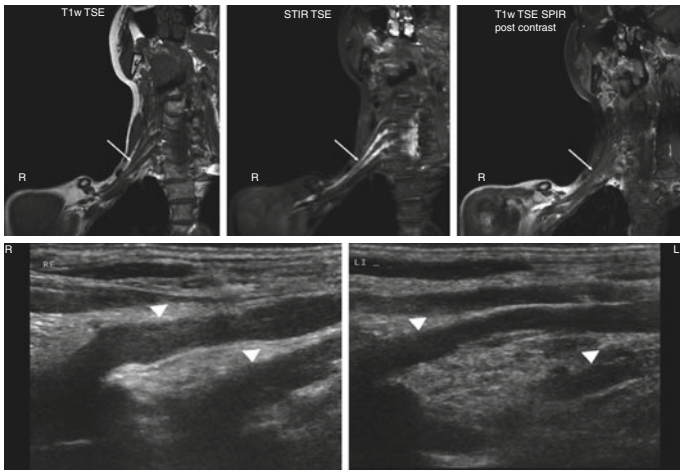

Fig. 6.23 Hypertrophy and hyperintense lesion of the roots C5 and C6 in a patient with chronic inflammatory demyelinating neuropathy on T1 and STIR images. Note the slight contrast uptake. *Below*: corresponding ultrasound images of the affected (*right*) and unaffected side (*left*). *Arrowheads*: roots C5 and C6

uptake of contrast agent in CIDP involving lumbar or cervical nerve roots as well as of the brachial or lumbosacral plexus [84, 154]. An example is shown in Fig. 6.23.

Among the different disorders characterized by affecting multiple nerves is the Parsonage-Turner syndrome, also known as "neuralgic amyotrophy" (see Sect. 10.3). It typically gives a sudden onset of severe shoulder pain, accompanied or followed by sensorimotor deficits with a "patchy" distribution pattern, whereby nearly every nerve of the brachial plexus may be affected. Several mechanisms, especially of autoimmune origin, have been discussed [157]. According to our own experience, patients may show a long-segmental or fusiform enlargement of the affected roots, of parts of the brachial plexus, of peripheral nerves, or of even single fascicles within a trunk on T1-weighted

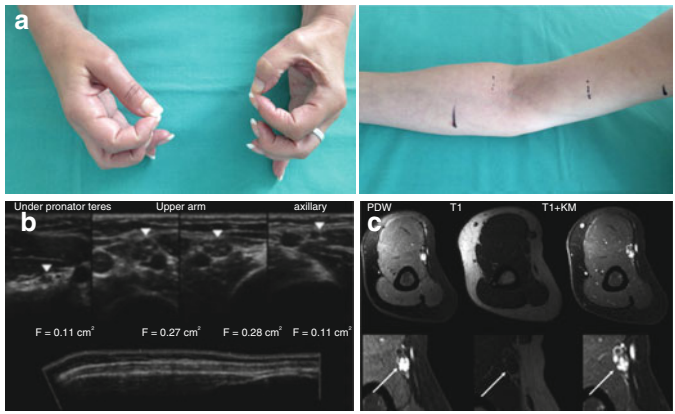

Fig. 6.24 Parsonage-Turner syndrome mimicking a palsy of the right anterior interosseus nerve. (**a**) Clinical findings. The swelling of the median nerve and its fascicles is located between the two *dotted lines*. (**b**) In high resolution ultrasound: a long segmental swelling of the median nerve and its fascicles (*arrowhead*) at the right upper arm can be demonstrated. (**c**) MRN findings, same case: effacement of the fascicles and nerve swelling on T1-weighted images, severe hyperintensity on PDW images and remarkable contrast uptake on T1 SPIR images. *Arrow*: median nerve

images with a corresponding hyperintensity on T2-weighted images. However, we found contrast uptake in a few cases only. Pham et al. [158] found in 20 cases of neuralgic amyotrophy mimicking an interosseus anterior syndrome a T2-lesion of the corresponding fascicle group within the median nerve at upper arm. However, further systematic studies are needed concerning autoimmune neuropathies including the Parsonage-Turner syndrome and chronic inflammatory neuropathies and their variants; research on gadolinium uptake and its time course will help to differentiate them better from other nerve lesions such as plexiform tumors (Figs. 6.24 and 6.25).

Hereditary sensorimotor neuropathies (CMT1) and multifocal motor neuropathies (MMN) may show a similar characteristic hypertrophy of the cervical roots and peripheral

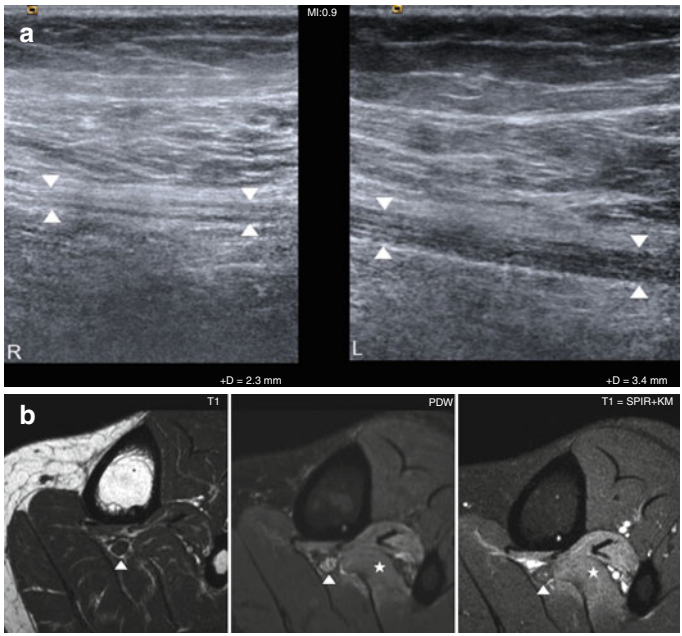

Fig. 6.25 Parsonage-Turner syndrome mimicking a palsy of the left distal tibial nerve. (**a**) In high resolution ultrasound: a long segmental swelling of the left distal tibial nerve and its fascicles (*arrowhead*) in comparison with the unaffected side. (**b**) MRN findings, same case: effacement of the fascicles and nerve swelling on T1-weighted images, moderate hyperintensity on PDW images and no contrast uptake on T1 SPIR images. The two hyperintense dots are veins. *Arrowheads*: tibial nerve. *Asterisk*: note the muscle denervation changes: hyperintensity on PDW and contrast uptake on T1-SPIR images

nerves (please refer to Sects. 10.4 and 10.5.2). In some cases of diabetic neuropathy with a distal symmetric affection, additionally, the proximal nerve portion demonstrated a hyperintensity of fascicles on T2-weighted images [130].

The diagnostic value of the imaging approach on neuropathies results from the capability to detect abnormalities in

different nerves in a single investigation. This statement holds true especially if a pure mononeuropathy is clinically presented but the anamnesis survey gives reason to suspect more than an entrapment. However, the same statement already applies to high resolution ultrasound techniques. For more details please refer to Chap. 10.

6.2.9 *Secondary Alterations of the Muscle Following Nerve Damage*

As already mentioned above, axonal loss leads to denervation of muscle substance. In the early stage, this reaction is characterized by an oedema. Subsequently, MR imaging shows a hyperintensity on T2-weighted images and contrast uptake on T1-weighted fat saturated sequences (Fig. 6.25b). Regeneration and axon re-growth is lacking after nerve injury muscular atrophy starts. This process is characterized by a remodelling of muscle tissue with connective tissue and fatty tissue. Together with a decrease of muscle diameter, this can be depicted with the appropriate sequences [112, 113, 115], again similar to our findings by means of ultrasound.

6.2.10 *Conclusions*

MRN provides some important additional information to assess the diagnosis and differential diagnosis of focal neuropathies. Especially deeply situated structures of the peripheral nervous system such as the lumbosacral plexus and the intrapelvic part of the sciatic nerve can be examined much more accurately than in high resolution ultrasound. We dispose of high contrast resolution and the capacity to administer contrast agents. However, MRN is not applicable in obese patients and in patients with a pacemaker or cardiac defibrillator, and it is difficult to apply

when dealing with children. The method is not available everywhere and is cost- and time-consuming [159].

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Chapter 7
Surgical Treatment of Focal Neuropathies

Götz Penkert

7.1 Technical Implications

The surgical treatment of focal entrapments predominantly consists of nerve relief and decompression. Entrapment syndromes are lesions in anatomically narrowed regions. First, normally existent anatomical structures can start to irritate the nerve. Second, the site where the nerve pierces a hard fascia can constrict the tissue. Third, the roof of a preformed tunnel can compress the nerve when additional structures within the tunnel develop chronic swelling. Subsequent surgery aims at removing the irritating structure, widening the piercing site, or transecting the ligamentous roof of the preformed tunnel. This kind of surgery remains restricted to the paraneural space. The epineurium and all of its contents remain untouched. Intraneural tissue reactions like fibrosis have to be avoided if at all possible.

In the 1970s, interfascicular neurolysis procedures were favored even in carpal tunnel symptom cases [1]. Thereafter it became quiet again concerning these tendencies; we must suppose that the considerations of Sect. 4.2 gave reason to its re-disappearance.

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Nevertheless a confrontation with an entrapment case which presents a preoperative severe loss of function as well as an unexpected impairment of electrical nerve conductivity is not rare and happens repeatedly. Occasionally during operation the surgeon can then find an unusual thickening and hardening of a nerve segment and has to decide immediately whether to leave the nerve behind with bad prognosis or to start a microneurolysis [2]. During the last few decades, scientific discussions have taken place on the option of stepwise neurolysis in these special cases of fibrosis resp. pseudoneuroma. The fact that such a procedure can increase pain, and even induce a neuropathic pain as previously demonstrated in Sect. 4.2 must not be neglected. Alternatively, when we leave fibrosis behind, chronically manifested motor deficits will probably not improve. Progressed ulnar nerve and tibial nerve neuropathies may intra-operatively present these unpleasant dilemmas.

Compared to entrapments, an iatrogenic focal nerve injury frequently affects for instance the spinal accessory nerve [3]. Further iatrogenic injuries to other nerves can follow screw removal procedures. Function deterioration can occur after bone injury followed by scars in the surroundings, as well as by scars after arterial puncture and hematoma. Sometimes, such injury related focal neuropathies then need more than a simple outside decompression, especially, if the thin spinal accessory nerve is involved as described in Sect. 8.1. If pain is not in the foreground and improvement of motor deficits is particularly required, neurolysis steps have absolutely to be taken into account. The surgeon has to decide immediately because subsequent approaches carry greater risks of worsening. The natural explanation is that the primary surgeon already leaves scar tissue behind. Thus, the primary surgeon bears the entire responsibility.

We therefore do not hesitate to describe the procedure of neurolysis, and in doing so, point out that it is a stepwise procedure [4]. It should be stopped immediately if the microscope shows that fascicles are swelling. The following different neurolysis

steps must not be engaged at any price in order to prevent any following neuroma pain. Under magnification, the epineurium in the intact part of the nerve proximal and distal to the involved area can be incised longitudinally. After that, the external sheath of the epineurium which surrounds the nerve trunk can be removed. As the third step, separating of fascicle groups from each other can start – "interfascicular neurolysis". Our goal has to be to achieve sufficient decompression to enable the axon sprouts to overcome the lesion and reach their target. We must always bear in mind that a fibrosis of connective nerve tissue exerts circular compressing forces on the fibers. As result, a grade II lesion according to Sunderland's classification occurs (see Sect. 3.1). The re-growing capacity of nerve axons starts when these forces are decreased. This factor forms the basis of "neurolysis." We expect increased criticism with regard to these considerations. However, when patients with progressed motor deficits and even without any injury in their history, and without pronounced pain, demand improvement, the surgeon has either to advise and apply or to deny such neurolysis steps. Gender neutral: The surgeon has the responsibility and has to make the final decision; it may be influenced by preoperative discussions with the patient about risks concerning pain. These reservations do not apply to surgeons dealing exclusively with efferent nerves like the spinal accessory, anterior, or posterior interosseous nerves. They are without any pain risk of course (Fig. 7.1).

7.1.1 Conclusions

Focal neuropathies to which we offer surgical treatment are a challenge: no doubt the patient appreciates the surgeon's immediate success; however, the surgical procedure which the physician chooses has primarily to be the suitable and correct one. Secondary corrections with re-exploration comparatively bear higher risk of deterioration. The surgical procedure itself is relatvely easy,

Fig. 7.1 Principles of interfascicular neurolysis which can be applied on severe nerve trauma. The epineurium is pealed away. The remaining interfascicular fibrosis is tried to be removed so that fascicles expand

whereas the pre- and intraoperative decisions to be made are of high responsibility.

7.2 Surgery in Bloodless Field

Do we need a tourniquet to provide a bloodless operating environment in nerve surgery? Our personal opinion is that we do not. Without any doubt, there is need of bloodlessness in hand surgery and in reconstructive surgery (e.g., in tendon transfers with soft tissue perforation from one incision to the next). However, guidelines for hand surgeons, neurosurgeons, and orthopedic surgeons recommend operating in bloodless field, especially with carpal tunnel syndrome and other easy entrapments of the forearm. However, they concede that the question of bloodlessness remains controversial at present. Nevertheless, the question gets more and more import forensically. Pitfalls after median or ulna nerve entrapment surgery almost inevitably lead to litigation. Before criticism arises concerning our opinion, please read the following citations:

1. Stewart in 1987:“Nonetheless, neuropathies following the use of pneumatic tourniquet still occur; sometimes they can be attributed to a faulty aneroid gauge and excessive pressure or to prolonged tourniquet application, but they may happen despite all normal precautions” [5].
2. Sunderland in 1991:“…. The much higher incidence of causalgia following high nerve injuries is due to wounding by high velocity missiles. The severity of the retrograde neuronal changes is known to be proportional to the severity of the injury and its proximity to the cell bodies” (one of the author’s listed nine “characteristic clinical features of causalgia”) [6].
3. Kline and Hudson in 1995: “If an extremity is operated on under tourniquet and the latter is inflated for 60 min or longer, the tourniquet should be left down for 20 min or more before NAP recording is tried. Ischemia and low wound temperature can block successful recordings. In several earlier cases in which recording were done under tourniquet, NAP traces were flat, and yet regeneration, as shown by either histological study or resected segment or subsequent clinical cause, was adequate” [7].
4. Birch in 1998:“Evidently, a paralysis is always produced when an occluding cuff is in position for 30 min or longer. It nearly always recovers spontaneously within a few minutes of the release of the pressure. Sometimes the paralysis is longer lasting; rarely, some of its effects are permanent. The reversible paralysis is that of transient ischemia; the long-lasting paralysis is that of damage to the myelin sheath; the permanent paralysis is that of damage to all elements of the nerve, including the axon, with ischemic damage to muscle” [8].

The term “causalgia” is today synonymous with complex regional pain syndrome (CRPS). The above-mentioned guidelines concerning the carpal tunnel syndrome declare that such a CRPS occurs extremely rarely. Nevertheless, in 7,000 endoscopic carpal tunnel releases, a CRPS was observed 10 times [9].

Further facets are:

- In a regional presentation of rehabilitation medicine in 2009, we were confronted with the fact that one quarter of patients with CRPS had undergone carpal tunnel syndrome surgery; in that special region, surgeons always work under a tourniquet.
- Nerve surgery is difficult when proximal approaches are needed, but it is easy in the forearm. Why should we have to apply a tourniquet where surgery is easier?
- We personally remained absolutely free of CRPS complications since 1981, with strict avoidance of a tourniquet.
- On the other hand: many CRPS I and II patients consulted us. In all cases some kind of surgery had been previously undertaken and always under tourniquet, e.g., osteosynthesis of distal radius fracture or ankle fracture, removal of osteosynthesis material, and release of tarsal tunnel syndrome. Our therapeutic idea to treat these terrible results consisted of nothing but peripheral nerve stimulation (PNS) such as representatively reported in 1996 [10].
- Rat experiments, meantime, underlined the fact that ischemia seems to be mainly responsible for inducing sympathetic reflex dystrophy [11].

Without doubt, several personal opinions have just been expressed, whereas randomized scientific evidence is still lacking.

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Chapter 8
Nerve Entrapment at Shoulder and Arm

Josef Böhm, Götz Penkert, and Thomas Schelle

8.1 Spinal Accessory Nerve

8.1.1 Anatomy

At the level of the skull base, the accessory nerve exits through the jugular foramen together with the internal jugular vein to which it is situated laterally. It leaves the jugular vein behind the digastric muscle and then runs on fibers of the levator scapulae downwards, where the nerve course is covered by the sternocleidomastoid muscle belly. At the lateral border of this muscle, three nerves arise quite superficially: the occipital minor nerve, the great auricular nerve (both nerves wrap around the lateral muscle border and run upwards), and the spinal accessory nerve which runs downwards and laterally into the posterior triangle of the neck. The exit point of these three nerves serves as an important landmark for finding the spinal accessory nerve surgically. In the whole posterior triangle, the nerve is situated subcutaneously, and it is therefore frequently in danger of being damaged during diagnostic extirpations of lymph nodes. Before entering the trapezius muscle it divides into several motor branches as functional supply.

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It is very important to remember that parts of the trapezius muscle are additionally supplied by motor branches of the cervical plexus, as, e.g. described by Allieu et al. (1982) [1]. Anatomical descriptions even exist to the effect that cervical plexus motor branches can substitute the accessory nerve completely. We have occasionally observed patients suffering from impairment of physical activities and with visible atrophy of the superior part of the trapezoid muscle following surgery on persistent lateral neck cysts. They are located more cranially than lymph nodes, and surgery may easily hit this cervical plexus supply. We remember two cases with preserved continuity of the spinal accessory nerve, although with severe motor deficits following a neck cyst surgery. Intraoperative nerve action potentials, during exploration of these cases, surprisingly revealed normal function of the accessory nerve indicating isolated damage to the plexus branches.

Nerve connections between the 11th nerve and the 1st and 2nd cervical roots do additionally exist [2]. They carry afferent fibers, but they are usually not involved in the typical focal spinal accessory nerve lesions we have to deal with.

8.1.2 Damaging Factors

For the most part, spinal accessory nerve lesions are of iatrogenic origin, and only quite rarely are they due to a perforating injury [3]. As mentioned above, superficial lymph node biopsies in the posterior triangle are the main damaging cause [4, 5]. Real nerve entrapments of spontaneous onset do not exist. We nevertheless include this iatrogenic nerve injury in our compilation because of its focal character as is to be seen in Fig. 8.1.

8.1.3 Clinical Symptoms

The main complaints consist of pain in the shoulder region and of a lowered position of the affected shoulder [6]. Subsequently,

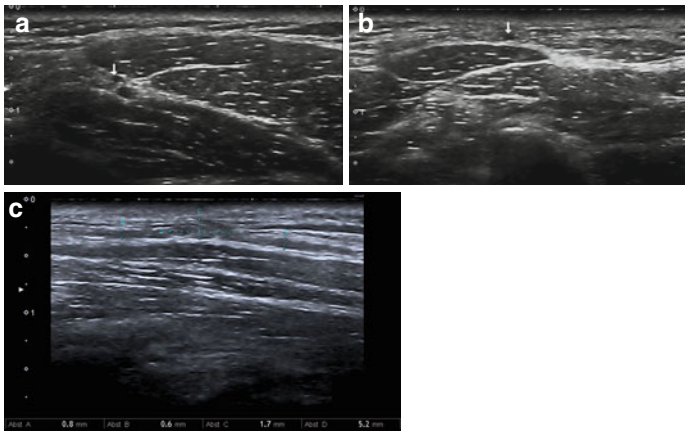


Fig. 8.1 Neuroma in continuity after lymph-node excision of the spinal accessory nerve. (**a**, **b** Cross-sections. **c** Longitudinal section). (**a**) Spinal accessory nerve (*arrow*) at the anterior border of the trapezius muscle. (**b**) Spinal accessory nerve (*arrow*) at the posterior triangle of the neck at a depth of about 3 mm beneath the skin surface. (**c**) Neuroma in continuity

the trapezoid deficit causes an asymmetrical aspect of both shoulders.

Why does pain occur despite the fact that the spinal accessory nerve exclusively carries motor fibers in only two-thirds of its lateral course? Overstretching forces to the brachial plexus have been discussed as etiology [6], but it seems more reasonable that pain is the result of overstretched fibrous ligaments or the result of overloaded muscles which have to compensate permanently.

The supraspinous and deltoid muscles abduct the humerus into a 90° position; then its further abduction requires rotation of the scapula [7]. This movement needs traction of fibers of the superior part of the trapezoid muscle and additionally of the levator scapulae and rhomboid muscles. The latter muscles have to compensate lost forces of the trapezoid fibers after a focal nerve lesion. The winging of the scapula is less distinct as in cases of serratus anterior muscle palsy following functional loss of the long thoracic nerve.

8.1.4 Electrodiagnostics

In order to differentiate major lesions, where a surgical repair is applicable, from minor lesions, electrodiagnostic workup of the spinal accessory nerve should include both nerve conduction studies and needle electromyography. Different techniques with updated normal values have recently been published in the literature [8, 9]. They allow examination of all segments of the trapezoid muscle (upper, middle and lower parts) separately. Using two different stimulation sites (posterior cervical angle at the midpoint of the posterior margin of the clavicular head of the sternocleidomastoid muscle for distal stimulation and the anterior-inferior angle of the mastoid process for proximal stimulation), a distal and a proximal motor latency. Additionally, a CMAP amplitude can be measured, moreover, the motor nerve conduction velocity can be calculated [9]. Thus, an early stage of lesion with only poor demyelization is already detectable. The motor nerve conduction velocity of the spinal accessory nerve in healthy individuals (upper segment of trapezoid muscle) is 65.7 ± 3.1 m/s and the CMAP amplitude 11.6 ± 3.5 mV [9]. Furthermore, as described in Chap. 5, needle electromyography helps to identify an axonal lesion by deriving evidence of denervation potentials in the trapezoid muscle. In the case of an additional absence of voluntary motor unit potentials or reinnervation potentials, a major lesion which probably needs surgical repair can be assumed [10].

8.1.5 Imaging

The superficial part of the course of the spinal accessory nerve is very easily accessible with high resolution ultrasound [11, 12]. The nerve runs across the posterior triangle of the

neck with either a straight (56 %) or tortuous (44 %) course at a depth of about 3 mm beneath the skin surface and penetrates the anterior border of the trapezius muscle. This point can be used as landmark. A mean nerve caliber of 0.76 ± 0.12 mm is reported in the literature [11]. Especially in the case of iatrogenic nerve lesions in the cervical triangle, high resolution ultrasound can depict the neuroma formation in continuity (Fig. 8.1) or, alternatively, a nerve transection [13]. On the other hand, the cisternal and intraforaminal course of the spinal accessory nerve is, meanwhile, best visualized by means of high spatial resolution b-FFE gradient echo MRI [14].

8.1.6 Treatment

The extracranial course of the spinal accessory nerve is never irritated or entrapped by pre-existing anatomical structures but is focally damaged due to mechanical factors of a perforating character. Such injuries leave behind a small region of scar tissue. When exploring the nerve, when exploring the nerve the area should initially be avoided, exposure should be started at a more central level of the nerve course, which means at the lateral margin of the sternocleidomastoid muscle. The exposure of the wrapping occipital minor and auricular major nerves acts as a landmark and is extremely helpful. This principle is almost mandatory as, in the case of nerve discontinuity, the proximal nerve stump rather frequently disappears behind the muscle belly of the sternocleidomastoid [7]. In order to follow these principles, the original biopsy skin incision which corresponds to skin creases situated transversely has to be extended. Vertical limbs must be added to the primary small horizontal incision to allow nerve exploration within healthy tissue above and below the scar area. Unfortunately, such incisions are cosmetically undesirable [15]. Millesi therefore

advocates several skin incisions situated parallel and with explicit regard to the skin creases,

Usually the nerve enters and exits an area of hard scar tissue. We then need a microscope and microsurgical instruments to answer the question of whether nerve continuity is evident or absent.

It may take about half an hour. The spinal accessory nerve is thin and of monofascicular structure at the level of the posterior triangle. In the case of persevered nerve continuity, the fibrosis involves the epineurium. This epineurium has to be longitudinally incised over the short distance of the lesion [7, 15]. Because of the small nerve caliber and its intraneural pattern, internal neurolysis steps are obsolete.

Cases with lost nerve continuity need nerve repair, very rarely achieved by end-to-end suture, mostly by one single auto-logous nerve graft. We have described the details of accessory nerve repair previously [15]. Although the focally damaged spinal accessory nerve lies superficially, just under the skin, exploration and repair remain a great challenge in peripheral nerve surgery for the following reasons: despite the little procedure that injured the nerve, we may even be confronted with almost complete nerve avulsions. The central avulsion then happens behind the sternocleidomastoid muscle. We are pleased when we find a small neuroma indicating the proximal nerve end. The same holds true in the periphery: the avulsion can hit the ramification of the nerve; distal nerve stumps do not develop neuromas. The identification of the distal nerve stump may therefore easily present more difficulties than the exposure of the proximal neuroma. Each surgeon now takes full responsibility for the primary surgical intervention. Secondary re-explorations lead to rather more difficulties and can fail completely due to extended scar tissue. As we have to expect a large field of varying pathologies – from short distance scar entrapment to long distance avulsion, spinal accessory nerve surgery should not be underestimated.

8.2 Brachial Plexus

8.2.1 Thoracic Outlet Syndrome (TOS)

8.2.1.1 Anatomy

The literature about anatomy, damaging factors, and treatment is so immense and differing in detail that we have to restrict our statements to personal experiences. Citations of literature will also be limited to examples which are representative of specific viewpoints.

We must define the neurogenic form of the syndrome as a nerve irritation syndrome. The five cervical nerve roots run together with the subclavian artery and vein between the scalene muscles laterally and downward. The two lower roots first rise a little through the upper thoracic outlet, and then unite to the inferior trunk which turns downwards. The interior margin of the first rib and sometimes, additionally, a cervical rib has thereby to be overcome. This anatomical situation is in itself completely normal because it is pre-existing in every human. An irritation syndrome of the inferior trunk can, however, start when anomalies or phylogenetically residual structures occur with fibrous ligaments within the scalene muscles, additional scalene muscles, fibrous ligaments between an elongated cervical transverse process and the internal margin of the first rib. Sometimes, if a cervical rib is present, such a fibrous band starts from its end, and runs obliquely through the thoracic outlet to insert somewhere at the first rib. Slim persons with a long and thin neck, predominantly women, are particularly prone to develop lower plexus irritation due to these structures. We should not forget that additional structures run transverse between the scalene muscles and fibrous bands: the subclavian artery and vein, and the transverse colli artery sometimes of large caliber. Pulsating forces within the arteries are quite able to press the comparably soft nerves against the fibrous ligaments. Due to

arm abduction, daily activities, or unfavourable positioning overnight, the pre-existing narrowness in the thoracic outlet increases so that vascular and neurogenic symptoms will easily be induced or will deteriorate.

8.2.1.2 Damaging Factors

In the thoracic outlet we are not confronted with a particularly narrow tunnel as elsewhere in limbs, but exclusively with an unfavourable combination of life-long pre-existing structures which leave behind a narrowness. The onset of symptoms is, therefore, usually independent of any incident or injury. Typical damaging factors are often unknown. Instead, symptoms start spontaneously. Patients almost never have a reasonable explanation as to why and exactly when discomfort came into their life. Therefore patients and physicians take a long time to realise that a thoracic outlet syndrome might be the answer.

8.2.1.3 Clinical Symptoms

Patients' complaints consist of electric-current-like sensations almost always irradiating into the ulnar aspect of the hand. Sometimes they observe these disturbing sensations during special arm positions or movements. In cases that have progressed further, patients sustain atrophy of the intrinsic hand muscles. Permanent numbness is not in the foreground, but it is discomfort by disturbing dysesthesias instead. As most of these patients with thoracic outlet syndrome don't present any objectively noticeable deficits, physicians often suspect that patients are exaggerating their symptoms.

The onset of symptoms is usually unrelated to any trauma; they start spontaneously, and they increase chronically. Nevertheless, very rarely, the patient is able to remember a certain period of time when the symptoms actually emerged.

Sometimes he even discusses a potential incident which he then suspects is responsible, like having carried heavy moving boxes on his shoulder in the past.

The typically nonspecific symptoms may be presented bilateral. On the other hand, the existence of a cervical rib on only one side does not really dispose which side is symptomatically affected. As fibrous ligaments are already mentioned as being responsible for symptom origin, the side and size of a cervical rib that has been examined in the patient's X-ray do not determine which is the affected side of a plexus irritation. We even observed the opposite situations more frequently with a cervical rib on one side and symptoms due to fibrous bands on the other. Nevertheless, bilateral symptoms are possible with and without cervical ribs, with and without the enlargement of the seventh vertebra's transverse processes, but always in cases of very slim persons.

From the neurological viewpoint it may be important that the Trömmner reflex runs through the inferior trunk, and that the Roos testing (paresthesias during finger motion with arms abducted, elbow joints 90° flexed, and hand palms to outside) turns out pathologically on the affected side. In our own experience, we particularly rely on the findings during supraclavicular palpation: this maneuver presses nerves against the disturbing ligamentous structure and immediately increases the amount of dysesthesias. We are convinced that this sign is a real aid in reaching the diagnosis. It indicates the location of a slightly degenerating and regenerating process like the Tinel sign. It even indicates the affected structure of the brachial plexus, namely, predominantly, the inferior trunk. By doing so, the patient observes dysesthesias exactly in the sensitive distribution area of the affected trunk. This area is similar to a dermatome, and completely differs from an autonomous peripheral nerve distribution area; for instance, numbness or dysesthesias of the ulnar nerve present an exact border in the middle of the fourth digit, whereas inferior trunk irritation also involves the third digit, sometimes with a border in the middle of the third

digit. Of course, it should not be a matter of discussion that a cervical disc protrusion C6-7 has to be excluded before any thoracic outlet surgery is indicated.

8.2.1.4 Electrodiagnostics

As mentioned above, thoracic outlet syndrome is a controversial topic in the literature. Besides TOS resulting from vascular compression, a neurogenic form occurs. The neurogenic TOS can be further subdivided into a disputed and a true form. The majority of neurogenic TOS belongs to the disputed form (95 %), whereas the true neurogenic TOS is very rare and accounts for less than 1 % of cases [16]. The true neurogenic TOS is characterised by an affection of the inferior trunk of the brachial plexus together with objective diagnostic findings, whereas the disputed neurogenic form of TOS produces only subjective symptoms like chronic pain. In contrast to carpal tunnel syndrome and ulnar neuropathy at the elbow, unfortunately, there are no standardized guidelines regarding the EDX in the diagnosis of TOS at the moment. One reason is certainly its rarity. In this section we will focus on the EDX in the diagnosis of the neurogenic forms only. Regarding vascular TOS, please refer to the specialist literature. Taking into account the anatomy and pathophysiology, the main goal of EDX is to demonstrate a lesion of the inferior trunk, and, on the other hand, to exclude other pathologies which are similar to TOS, e.g. neuropathy of the ulnar nerve or affection of nerve roots C7 and C8. This easily succeeds in advanced stages of true neurogenic TOS with a pre-existing axonal lesion, but those cases where only a transient compression without permanent damage of neural structures occurs are rather difficult to detect (disputed neurogenic TOS). The registration of f-waves of the ulnar and median nerves in neutral position and after provocative maneuvers is unspecific, and a reduction of amplitudes of motor-evoked potentials after provocation was reported in two cases only [17, 18]. The intramuscular anterior scalene block may serve as a diagnostic tool

for disputed neurogenic TOS; it temporarily blocks and paralyzes the muscle and allows the first rib to descend, thereby decompressing the thoracic outlet. Pain relief following this diagnostic intervention may serve as a positive predictor when surgery is considered [19]. Demyelinising nerve damage (true neurogenic TOS) instead increases the chances of obtaining electrically abnormal findings. A pathologically changed sensory nerve conduction study of the medial antebrachial cutaneous nerve (sensory latency over 2.4 ms, latency difference of 0.3 ms or more, amplitude under 10 μV, and amplitude ratios of 2.0 or more) is therefore suggested as the most reliable approach. Considering patients with true neurogenic TOS, 40 out of 41 had at least one of these four diagnostic criteria [20]. Furthermore, it can simultaneously exclude a supraganglionic root lesion and a neuropathy of the ulnar nerve. It has to be valued superior to the simple registration of sensory nerve action potential of the ulnar nerve from the fifth finger, which is pathological in the case of true neurogenic TOS as well as in the case of ulnar neuropathy [20]. Other tests have been described in the literature. Fractionated motor nerve conduction studies both of the ulnar and median nerves together with registration of the corresponding f-waves may be helpful. Reduction in segmental motor nerve conduction velocity to <85 m/s of either the ulnar or median nerves across the thoracic outlet should corroborate the clinical diagnosis [21]. A similar test is the registration of compound muscle action potential latencies of the abductor pollicis brevis and abductor digiti minimi muscles after magnetic stimulation of the brachial plexus. Compound muscle action potential latency of the ABP over 11.04 ms and of the ADM over 10.60 ms can indicate a true neurogenic TOS [22]. Another useful diagnostic tool is the registration of dermatomal and mixed nerve somatosensory evoked potentials (SEPs); however, the abnormality rate for both ulnar and C8 dermatomal SEPs was 100 %-significant only in a small group of advanced disease with severe muscular atrophy, whereas in earlier clinical stages, this rate dropped significantly to 50–67 % [23]. If primary demyelization is followed by secondary axonal loss, or if axonal damage primarily occurred,

needle electromyography can easily demonstrate these changes by analysis of the distribution pattern of denervation in several muscles (see Sect. 5.6.2). In true neurogenic TOS the abnormalities occur in all muscles supplied by the root C8, and they do not occur in the paravertebral musculature of the cervical spine [24, 25]. This phenomenon helps to differentiate TOS from root C7 and C8 lesions due to cervical disc protrusion. In summary, needle electromyography together with registration of sensory nerve action potential of the medial antebrachial cutaneous nerve plays the key role in the diagnosis of true neurogenic TOS.

8.2.1.5 Imaging

High resolution ultrasound and 3 T-MR imaging provide an important diagnostic tool to detect pathological irritations of the brachial plexus and the cervical nerve roots, whereas EDX is nonspecific. High resolution ultrasound is a cost effective alternative to MR imaging outside the spinal cord [26]. Both high resolution ultrasound and MR are able to define the underlying pathology of brachial plexus involvement, e.g. of neoplasia, traumatic, or inflammatory origin, or due to congenital bone variations and fibromuscular anomalies [26, 27]. Using the transverse processes of the cervical vertebra as a landmark, it is easy to identify the cervical nerve roots with high resolution sonography. Taking into account that the transverse process of C7 has only a posterior tubercle, it becomes easier to determine the root level. In most individuals, this landmark will allow to start the examination of the whole course of nerve roots C4–C7 from the foramen to the interscalene space, and, furthermore, of its trunks above of the clavicle. In non-obese individuals, the root C8/TH1 and the brachial plexus below the clavicle are mostly visible [28]. Normal values of CSA (nerve roots C5–C8 and brachial plexus above of the clavicle) have been published only in a few studies to date [29–31]. At present, experience regarding differential diagnosis between true and disputed neurogenic TOS starts using high resolution ultrasound. Figures 8.2 and 8.3 illustrate imaging

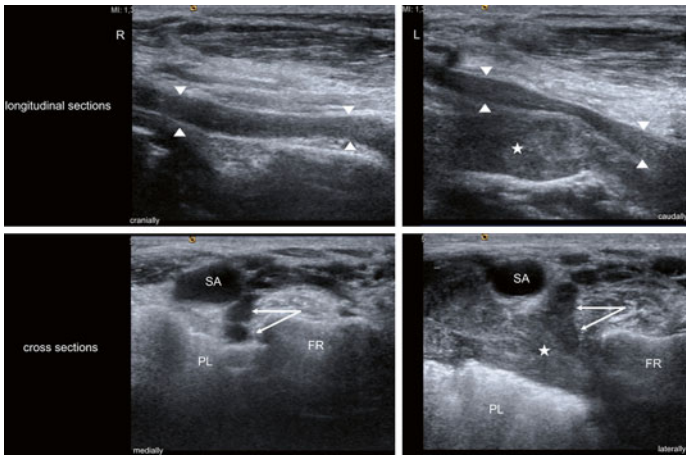


Fig. 8.2 Pancoast-tumor (*asterisk*) compressing nerve root C8/Th1 (*arrows*, *arrowheads*). *Right*: unaffected side, *left*: affected side (*SA* subclavian artery, *PL* pleura, *FR* first rib)

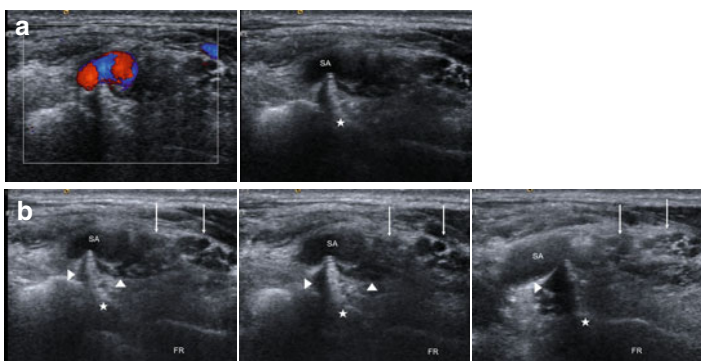


Fig. 8.3 Fibrous band at the medial edge of the medial scalene muscle (*asterisk*) compressing nerve root C8/Th1 (*arrowheads*) and subclavian artery (*SA*). (**a**) Compression of subclavian artery (colour-Doppler and greyscale images). (**b**) Image sequence (from cranial-left to caudal-right) showing the compression of nerve root C8/Th1 (*arrowheads*). *FR* first rib, *arrows* parts of brachial plexus supplied by roots C5-C7

abilities. According to our experience in 6 cases, fibrous bands and cervical ribs always demonstrated the same appearance wedge/sickle-shaped with hyperechoic edge of the scalene muscles as well as with a hypoechoic and thickened inferior brachial plexus trunk. External compression with the probe led to an indentation of the subclavian artery (Fig. 8.3). MR imaging additionally provides the opportunity to demonstrate the T2 nerve lesion in the case of entrapment (see Chap. 6) and the administration of contrast agent. This may be helpful in differentiating between entrapment from other types of pathology such as neoplasm or inflammatory immune neuropathies and to localize the site of entrapment precisely – supraclavicular, infraclavicular, and subclavicular entrapment possibilities have at least to be considered [26, 27].

8.2.1.6 Treatment

As already outlined in the introductory remarks of this book, all young patients want relief of pain symptoms. Besides new diagnostic means, we primarily continue to rely on the Tinel sign which can be elicited when the nerves are pressed by the physician's fingertips against the irritating structure in the supraclavicular triangle.

The literature is full of advice on how to approach the proximal part of the brachial plexus in the case of mechanical irritation. The history of this development can be derived from Atasoy's extensive presentation [32]. The thoracic outlet syndrome is defined as a focal irritation. Consequently, the approach has to remain as small as possible. Personal experiences over more than two decades regarding the disputed and true neurogenic cases are as follows. First rib resection as often advocated is not mandatory; this also holds true in cases with associated vascular symptoms which can even consist of artery embolism or venous incidents like pulmonary embolism in patient's history. Inferior trunk damage following rib resection as sporadically observed means an intolerable complication; it has therefore to be avoided in all cases. Partial cervical rib

resection was necessary in about 50 %. Complete scalenotomy is unnecessary because muscle fibers do not compress nerves. However, ligamentous fibers hidden between muscles have to be identified and transected completely. In a single case of middle trunk compression, an elongated transverse process of C6 was shortened. When the internal margin of the first rib is particularly sharp, it can be rounded off by means of punches in order to reduce its additional irritating role. The supraclavicular approach was always sufficient to achieve vascular and nerve decompression and thus relief of symptoms. However, there is one restriction to be stated: all cases we operated on in this period always had neurogenic symptoms and, infrequently, additional vascular ones. Pure vascular thoracic outlet syndromes were not included in our series because, in our region, they are included in the field of vascular and thorax surgery.

Theoretically, three different approaches of different extensions to the thoracic outlet region are available, and, of course, for several decades all of them have been considered to be equally successful: the superior approach through the lateral neck triangle [33–35], the transaxillary approach which needs first rib resection [36, 37], and the posterior subscapular approach which is restricted to recurrent cases [34, 38]. Few authors recommend a combination of these options [39]. The whole history of recommendations since 1861 can be followed up – as already mentioned – in the comprehensive contribution by Atasoy in 1996 [32].

The main objections to the transaxillary approach are possible damage of the inferior trunk and a rather hidden localized cervical rib if existent [40], to the supraclavicular approach damage of the phrenic nerve [40]. Probably due to the surgeon's experience, these complications remain rare. Nevertheless, in reality, they are intolerable. Another intolerable complication is reported by Millesi et al. in 2007: sliding capacities of roots and trunks have to be preserved. The fat tissue which surrounds the nerves allows this sliding during extensive arm motion. This tissue has to be kept aside and never removed [41]. Secondary bleedings leave behind scar tissue and cause severe adhesions. A postoperative suction drainage of 2 or even 3 days is therefore mandatory in our

experience. Intra-operative looping of roots and nerves irritates mechanically; it could cause the surgeon to induce damage on previously untouched and normally functioning nerves. We should remember that the myelin sheath of the nerve fiber is rather sensitive; its damage may end up as grade I lesion in accordance with Sunderland's classification detailed in Sect. 3.1.

A small skin incision, 2–3 cm parallel to the clavicle, gives enough space. The skin in the neck region is extremely flexible and elastic so the actual approach site can easily be extended by two self-holding retractors. Of course, these retractors should not touch any nerve structure. Particular care has to be taken with the expected course of the phrenic nerve at the opening of the exposure. Adjacent, we find the above-mentioned package of loose fat tissue which we enter by using spreading movements of our scissors. We don't cut through the tissue both in order to identify small venous vessels which have to be coagulated and individually transected and in order to avoid injury to the important structures lying underneath. By doing so, the omohyoid muscle is isolated within the fat package and can be looped and kept away. Above the muscle, by palpation with the finger tip, it will be helpful to feel and identify where superior and middle trunks run laterally and downward. A little more medial, the anterior scalene muscle can be exposed. By longitudinally spreading movements a careful opening of its surrounding membrane finally leads to the phrenic nerve. Spreading movements of scissor tips must always remain parallel to the expected structure. Instruments that keep nerves such as the phrenic nerve away must always to be placed in such a way that avoids harm or even kinking. During the surgery, all these instruments have to remain under permanent control. We use Langenbeck retractors and never loop nerves. The phrenic nerve acts as border: the whole exposure remains lateral to it because, if medial to it, the surgeon is in danger of damaging the internal jugular vein and the vagus nerve. The superior trunk – previously identified by fingertip – is now carefully isolated far enough, with the middle trunk next to

it. At this point, both trunks can be slightly retracted upwards and lateral. Under them, palpation with fingertip will help, first, to find the irritating structure such as a fibrous band, and, second, to identify the subclavian artery which runs parallel to and under the clavicula, but sometimes also abnormally cranial convex curved. Before transecting or even removing any structure which you suppose to be responsible for the symptoms, a reliable identification of the inferior trunk and its division into the two centrally running roots C8 and T1 is mandatory. In most cases, the inferior trunk is flattened, and it rides across the band to lateral. Anatomical variations at root level, e.g. pre- and post-fixation of the brachial plexus, or a missing medial trunk are confusing [42, 43]. Prior to transecting the disturbing fibrous band, and sometimes, additionally, to opening a hypertrophic supra-pleural membrane you must have identified and isolated each structure. Along with this most important step, you can take a punch either to round off the internal margin of the first rib or to remove a cervical rib piece by piece, the latter however as extensively as possible. Rough bone edges left behind can cause new irritations. Before starting rib removal, its periost has to be carried off in order to skeletonise the bone. This procedure avoids injury to the parietal pleura and therefore reduces risks of pneumothorax decisively. Retracting instruments to keep nerves away again have to be under your permanent control. The suction in your left hand can aid as retractor in addition to instruments operated by your assistant.

As previously mentioned, a suction drain for 2 or even 3 days is necessary to exclude secondary bleeding and scarring. Before wound closure, the fat tissue is replaced into the supraclavicular groove. In addition, within this space are many small lymph vessels from which extravasations of lymph liquid will occur; this quickly stops when the wound sticks under suction (Fig. 8.4).

We therefore prefer hospitalization of these patients for no less than 4 days. In only one case a pneumothorax complication and the need for thorax drainage extended hospitalization. We

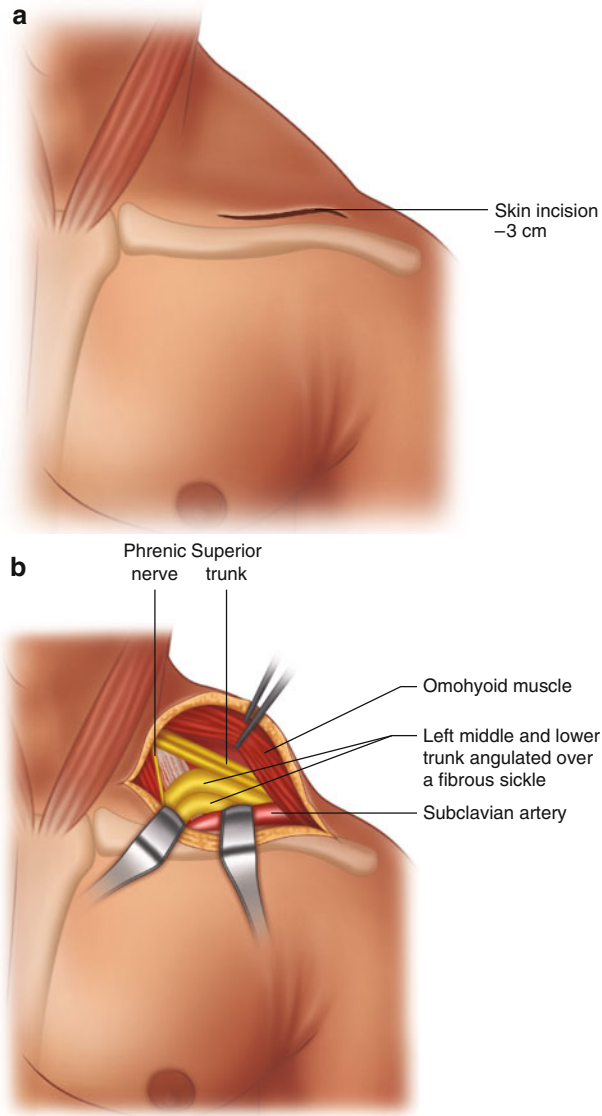


Fig. 8.4 Surgical treatment of TOS. (**a**) Supraclavicular skin incision. (**b**) Exposure of nerve trunks. (**c**) Transection of a disturbing fibrous "sickle"

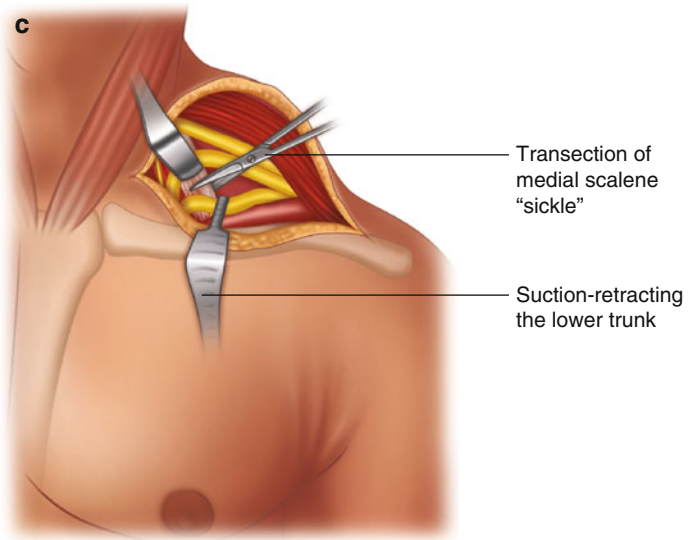


Fig. 8.4 (continued)

have had no cases of phrenic nerve lesion nor recurrences. In one of our recent patients, a female physiotherapist with a fuller figure, the elicitable pain disappeared at once; however two days later there was a sudden onset of a neuropathic pain in the neck and lateral triangle region. We are aware of criticism but these selected patients are in permanent contact with each other via the internet; thus problems come to light very quickly nowadays.

8.3 Costaclavicular Syndrome

We must assume that a compression of nerves and vessels between clavicula and first rib is a more or less historical idea. At first, it was described by Falconer and Weddell in 1943 [44]. They reported about soldiers who sustained symptoms in their arms after carrying heavy loads on their shoulders. The therapy they advised

was first rib resection. Following Falconer's first paper, literature presented a few further papers in 1949 and 1962 [45, 46].

However, our own experiences derived from conventional arteriography and modern MR-angiography demonstrated every time that a vessel stenosis during functional positioning of the arm occurred more proximal than the costaclavicular space is located. This phenomenon actually indicates that we always dealt with a classical thoracic outlet syndrome. Subsequently, our approach was the supraclavicular one without first rib resection as described in the thoracic outlet chapter. Falconer and Li described very comprehensively 50 years ago that they indeed preferred this approach and even succeeded in doing a first rib resection via the supraclavicular space; in their paper, the authors recommended to place the surgeon's fingertips between clavicula and first rib and to depress the shoulder downwards and backwards as an important maneuver to consider first rib resection in selected cases [46]. However, we think that these considerations are more or less historical ones.

The only realistic focal brachial plexus neuropathy that can occur in the costaclavicular space in any case, is the one related to hypertrophic callus after clavicula fracture. The treatment then consists of the reduction of the callus only or, in the case of a pseudoarthrosis, of a secondary stabilization of the clavicula.

8.3.1 Electrodiagnostics and Imaging

Please refer to Sect. 8.2.1.

8.4 Hyperabduction Syndrome

Comparable to the costaclavicular syndrome, the question again rises as to whether the hyper-abduction syndrome really exists. We found the first description in the literature by

Wright in 1945 [47]. Radiating dysesthesias were reported as depending on hyper-abducted arms during work and general daily activities.

We have experience with one such case. A few years ago, a young man presented repeated discomfort irradiating downwards into the sensitive area of the radial nerve, indicating an irritation of the posterior cord. A characteristic Tinel sign could be elicited in the infraclavicular groove as the only available guiding symptom to suggest surgery. At that time, functional neuroimaging was still not on the way.

The nerve irritation at cord level of the brachial plexus originates from the attachment of the pectoral minor muscle tendon at the coracoid process of the scapula. Therefore, surgery achieves relief by transecting this tendon. The skin incision can be angled at 90° to the clavicula and leads downward towards the axilla; the exposure should try to define the margins of the pectoral major and deltoid muscles with the cephalic vein in between. This vein indicates where you can easily enter into a sheath of fat tissue. Through this fatty package the pectoral minor muscle runs belly transverse to the course of the pectoral major muscle fibers. Of course, the pectoral minor muscle should then be isolated in its entire circumference. By doing so, you are able to identify the cord structures of the brachial plexus which lie underneath the pectoral minor muscle and parallel to the skin incision. You are still allowed to transect the tendon without any risk to the nerves.

This single surgery was done before qualified imaging experience as described in Chap. 6 came up. The indication to operate on the patient was exclusively a clinical one.

8.4.1 Electrodiagnostics and Imaging

Please refer to Sect. 8.2.1.

8.5 Long Thoracic, Dorsal Scapular and Thoracodorsal Nerves

Mechanically caused focal lesions of these three nerves are rare and comparatively unknown. The nerves play a role in selected cases of trauma-related brachial plexus lesions as axon donor (the dorsal scapular nerve) or special target nerves. Iatrogenic lesions rarely occur after extensive axillary lymph node extirpation. Nerve repair then requires such a great deal of effort that reconstructive surgery is usually preferred.

In particular, when a patient's history is characterised by sudden and severe pain with diffuse involvement of the shoulder region, then by pain disappearance, and next by motor deficits and quick atrophy with discomfort and inability to control and move the shoulder, the diagnosis of an inflammatory brachial plexus disease (Parsonage-Turner syndrome) has to be suggested as we describe in detail in Sect. 10.3. The onset of motor deficits is often outlined as occurring overnight. The symptoms may affect one of the three nerves separately or all three nerves together, and, additionally, the axillary (circumflex) nerve. MRI then reveals rapid fatty muscle substance degeneration and significant signal alterations of brachial plexus nerves. This important differential diagnosis must always be considered very seriously. Surgery should then be unconditionally avoided.

8.5.1 Clinical Symptoms

Independent of these diagnostic considerations, weakness of the serratus anterior muscle due to a long thoracic nerve lesion is considered a severe discomfort in patients' eyes. Active arm abduction then stops at 90° because active forces to turn the inferior margin of the scapula outward to support further abduction have been lost.

8.5.2 Electrodiagnostics

As stated above, a lesion of long thoracic, dorsal scapular and thoracodorsal nerves is rather uncommon, therefore reports about EDX and imaging of the three nerves remain limited. Nevertheless, all three nerves can be examined using Erbs´s point for surface stimulation and by means of surface or needle recording electrodes placed over or in the muscles supplied by these nerves (long thoracic nerve – serratus anterior muscle, thoracodorsal nerve – latissimus dorsi muscle, dorsal scapular nerve – rhomboid muscles). Thereby, a distal motor latency and amplitude of the compound muscle action potential can be derived. Generally, the upper limit of normal decrease in amplitude from one side to the other is 50 % [48]. The normal values of distal motor latency depend on the recording technique and also on the distance between stimulation and recording electrode. For details please refer to Le Monaco et al. [48]. In addition, needle electromyography can demonstrate the axonal damage in the specific muscles mentioned above, and it can especially help to detect the comparably frequent inflammatory neuropathies which likely involve the three nerves – e.g. a Parsonage-Turner syndrome, or, alternatively, a C5 root lesion [49].

8.5.3 Imaging

Unfortunately, there are only a few reports about imaging of the three nerves. The long thoracic and dorsal scapular nerves were visible under ultrasound assessment within or superficial to the middle scalene muscle after division of the root C5 in 90 % of cases. The dorsal scapular nerve pierces the middle scalene muscle similar to the musculocutaneous nerve which pierces the coracobrachialis muscle. The nerves were located at depths similar to the perceived C6 nerve root, but a reliable distinction between the two nerves was only possible with the aid of

electrostimulation [26, 50]. No knowledge exists of normal values of the CSA of the three nerves, nor of ultrasound imaging trials of the thoracodorsal nerve at the moment. Recently normal values of the anterior-posterior diameter of the long thoracic nerve (1.6 ± 0.3 mm) as well as examples of pathological involvement (e.g. Parsonage Turner Syndrome) have been described in a small retrospective study [51]. MR imaging may also become effective in recognising neuropathies of the shoulder girdle involving one or several of the three nerves [26].

Accordingly, trials of EDX and imaging on the long thoracic, dorsal scapular and thoracodorsal nerves will still continue at centers with specific research interests at the moment.

8.5.4 Treatment

The long thoracic nerve is the one and only nerve which is said to suffer from entrapment near the thoracic outlet region. The nerve arises at root level from C5 to C7, and runs posterior to the superior trunk downwards where it pierces the medial scalene muscle from outside to posterior. There are not previously published papers by Nath et al. in 2007 which report on 50 cases of long thoracic nerve decompression and neurolysis with post-operative significant improvement in 46 cases [52, 53]. The authors describe their usual approach as through the supraclavicular groove for exposure of roots and primary plexus trunks. They identify the long thoracic nerve posterior and lateral to the superior trunk. They add an external and internal microsurgical neurolysis. They mention that the nerve has several fascicles, and that it can also divide early into two or three branches at the location where the nerve traverses the middle scalene muscle. Its muscle fibers were finally transected as far as the situation required. This series seems to be the largest in literature. The rate of postoperative significant improvement of scapular winging within 1 day to 3 months is reported as 88 %. A few patients had a postoperative seroma similar to our experiences following thoracic

outlet surgery, a relatively minor important detail which nevertheless emphasizes that suction drainage should be left for long enough. Similar experience with four cases of long thoracic nerve neurolysis was described by Disa et al. in 2001 [54]. The authors also stated that reconstructive surgery by means of muscle transfer would be the method of choice if recovery failed to appear or if nerve repair was being considered after nerve injury.

It is correct that Nath and his co-authors mention the Parsonage-Turner syndrome as one of considerable etiologies of functional loss of the long thoracic nerve (see also Sects. 6.1.8, 6.2.8 and 10.3). Half of their patients experienced pain; unfortunately, the paper does not reveal any detail of its onset, time course and localization. It should not be neglected that the long thoracic nerve mainly consists of efferent fibers. Nobody knows whether the few afferent fibers from muscle spindles are somehow able to generate pain sensations which the patient then feels, a question which is discussed when referring to the interosseus anterior nerve in the work of Birch and co-authors in 1998 [55]. The selection of appropriate cases to operate on after occurrence of severe scapular winging was determined again (in Nath's 50 cases) according to patient history: particular attention was focussed on "injuries or any associated events" [52, 53]. It remains to be debated how to choose between inflammatory plexus neuritis and long thoracic nerve entrapment. Our personal impression was that our patient histories predominantly presented similarity to a plexus neuritis disease. It would be of immense help if surgeons could compare their series of operated and non-operated cases with each other in order to detect guiding clinical details which may help with the differential diagnosis. The development of modern imaging probably techniques will further contribute to the remaining diagnostic uncertainties. Findings of long thoracic nerve entrapments by imaging seem impossible because of the extremely thin nerve caliber and its hidden localization posterior to the superior trunk. Instead, indications of nerve entrapment can obviously be derived clinically, whereas differential-diagnostic evidence of plexus neuritis can be visualized by means of imaging.

8.6 Suprascapular Nerve

The suprascapular nerve can be involved in the field of focal neuropathies very frequently so we have to deal with it rather intensively.

8.6.1 Anatomy

The nerve arises from the superior trunk in the supraclavicular groove, turns to lateral and posterior, and runs through the incisura scapulae. The incisura has a ligamentous roof, the transverse superior scapular ligament. The nerve then has a kink of 60° to run downward where it divides into the supraspinous and infraspinous branches. The supraspinous branch turns again to lateral and supplies the supraspinous muscle which is situated in the supraspinous groove. Its tendon blends into the fibrous capsule of the shoulder joint which forms the rotator cuff. The infraspinous branch runs through the spinoglenoid notch into the infraspinous groove to supply the infraspinous muscle. Its tendon also blends into the fibrous rotator cuff. The course of the inferior branch has an additional roof between the superior and inferior fossa, the transverse inferior scapular ligament.

8.6.2 Damaging Factors

The main location of a focal nerve entrapment is the incisura scapulae with its transverse ligament as roof. Intensive sports, particularly volleyball or weightlifting, but also special exercises during ballet, can involve the nerve at the superior ligament where it kinks upwards during special maneuvers. Antoniadis et al. reported on their experiences with 28 cases in 1996; they outlined similar patients' histories [56].

8.6.3 Clinical Symptoms

Most complaints consist of weakness during abduction in the shoulder joint. For the rest, further complaints are more varied, e.g. impairment during daily activities and sports in the affected shoulder region. Clinical findings then are weakness of initial upper arm abduction via the supraspinous muscle and weakness of a rotation outwards via the infraspinous muscle, in a position of a 90° flexed elbow joint. Behind the scapula we sometimes find a small area of numbness; very rarely the patient presents pain in this area. It should not be neglected that a separate infraspinous branch entrapment can occur under the ligamentous roofs in the spinoglenoid notch [57]. The resulting clinical finding is then presented as outward-rotation weakness only without pain or numbness. Never to be neglected is the important differential diagnosis to a partial rotator cuff tear which has similar clinical symptoms but presents with severe pain [58]; considerable pain that the physician induces by palpation of the rotator cuff area leads to the supposition of such degenerative periarthrosis symptoms, whereas if pain is not present during palpation, such a periarthrosis can be rather reliably excluded. MRI of the shoulder joint area is then definitively able to describe the conditions of the supra- and infraspinous tendons in the rotator cuff. Electromyography of the affected muscles with pathological results instead acts as an aid to diagnosing an alternative suprascapular nerve entrapment.

No less frequently, a so-called ganglion turns out to be the origin of nerve symptoms. Its location is predominantly the suprascapular notch, and there it sits, occupying space along with the irritating ligamentous roof of the incisura scapulae. Moreover, and rather rarely, such a ganglion can be situated within the spinoglenoid notch and can alone cause entrapment of the infraspinous branch [59]. Logically, imaging as a mandatory procedure leads to these findings and helps to decide about different treatment modalities.

8.6.4 Electrodiagnostics

In suprascapular neuropathy, the highest abnormality rate could be derived from nerve conduction studies (88 %), whereas needle electromyography showed pathological alterations in only 33 % of cases. Surface stimulation was always performed at Erb's point, and compound muscle action potentials together with distal motor latencies were recorded from the supraspinous and infraspinous muscles. Therefore, in the majority of studies, concentric needle electrodes have been used [60]. The upper limit of distal motor latency to the supraspinous muscle was 2.71 ms, while data for the infraspinous muscle yielded an upper limit of distal motor latency of 3.42 ms in healthy individuals. Much more important than the distal motor latency prolongation are changes in the compound muscle action potential configuration in comparison to the healthy side [60]. The same test can also be performed with surface recording electrodes. The upper limit of distal motor latency is then 3.7 ms to the supraspinous and 4.2 ms to the infraspinous muscle, respectively [61]. In an advanced disease with axonal damage, needle electromyography shows denervation potentials either in both muscles, or only in the infraspinous muscle [62]. Furthermore, and again rather important, needle electromyography can exclude other pathologies, similar to suprascapular entrapment neuropathies, like a C5 lesion or an inflammatory affection of the upper brachial plexus (e.g. Parsonage-Turner syndrome; see Sects. 6.2.8 and 10.3) by means of examination of the muscles of the cervical spine or other muscles, e.g. the deltoid muscle, supplied by the root C5 but not by the suprascapular nerve. Abnormalities in needle electromyography and/or nerve conduction studies exclusively in the infraspinous muscle indicate entrapment neuropathy at the spinoglenoid notch, whereas the involvement of both muscles is compatible with entrapment neuropathy at the suprascapular notch [62].

8.6.5 Imaging

In order to apply peripheral nerve blocks the suprascapular nerve was depicted using high resolution ultrasound within the supraspinous fossa or at the suprascapular notch, however, at this site, the nerve is located deep under the trapezoid and supraspinous muscle bellies, and hence it is hardly visible there [63]. It can be visualized at the division of the nerve root C5, running posterior over the middle scalene muscle and then passing underneath the omohyoid muscle at a level slightly above the clavicle (Fig. 8.5). At this point

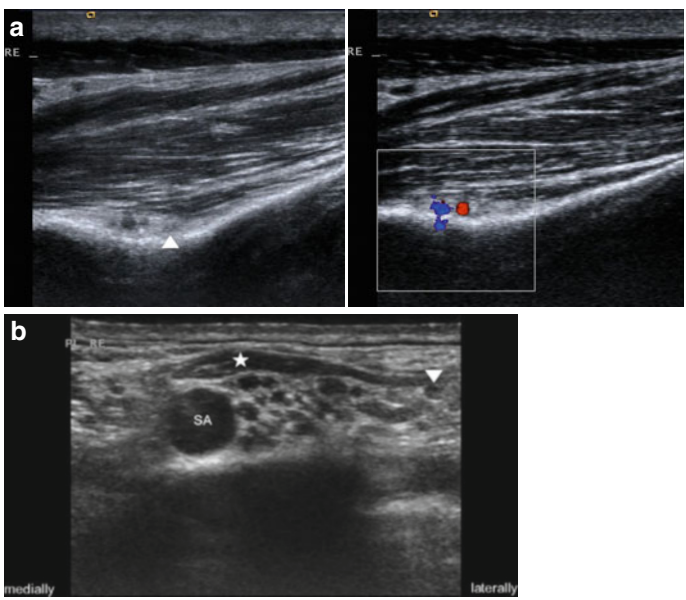


Fig. 8.5 Normal anatomy of the suprascapular nerve in HRUS. (**a**) Greyscale and colour-Doppler images of the suprascapular nerve (*arrowhead*) and the accompanying vessels at the level of the suprascapular notch below trapezoid and supraspinous muscle bellies in a slender individual. (**b**) Suprascapular nerve (*arrowhead*) at level of the supraclavicular brachial plexus below the omohyoid muscle (*asterisk*), *SA* subclavian artery

diagnostic nerve blocks can be performed precisely [64]. However, this site is not the location of the classical entrapment. Its normal CSA ranges from 1.9–2.0 mm^2 [65].

Therefore 3 T MRI should be preferred over high resolution ultrasound if a suprascapular nerve entrapment is suspected. MRI is especially able to demonstrate a T2 lesion at and above the site of entrapment (regarding spinoglenoid notch or suprascapular notch) and the fatty atrophy in the muscles supplied by the suprascapular nerve in the case of a chronic axonal loss. Moreover, MRI reveals special entrapment causes such as anomalous or thickened transverse scapular ligament, space-occupying lesions like a ganglion cyst or a soft tissue tumor (Fig. 8.6). It differentiates severe and painful traction injuries occurring with retraction of a large rotator cuff tear. In addition, mimicking conditions as mentioned in the EDX section in Chap. 5 can be excluded using MRI [62, 66, 67].

8.6.6 Treatment

The literature is full of recommendations on how to approach the suprascapular nerve and notch. Nerve entrapments are predominantly located under the superior transverse ligament, which the surgeon has to transect. The suprascapular artery is mostly lying upon the ligament whereas the nerve runs beneath. It may rarely happen that the supraspinous nerve branch runs together with the artery on the ligament. These details have to be clarified before ligament transection starts. Different approaches to the suprascapular notch are described in the literature: a lateral approach with the patient in a prone position [68], from above with skin incision transverse to the margin of the scapula and with respect to skin creases, additional detachment of trapezoid muscle fibers from the clavicula [69], transmuscular approach from behind with patient in supine position, fibers of the supraspinous muscle belly are split [56, 70], as with

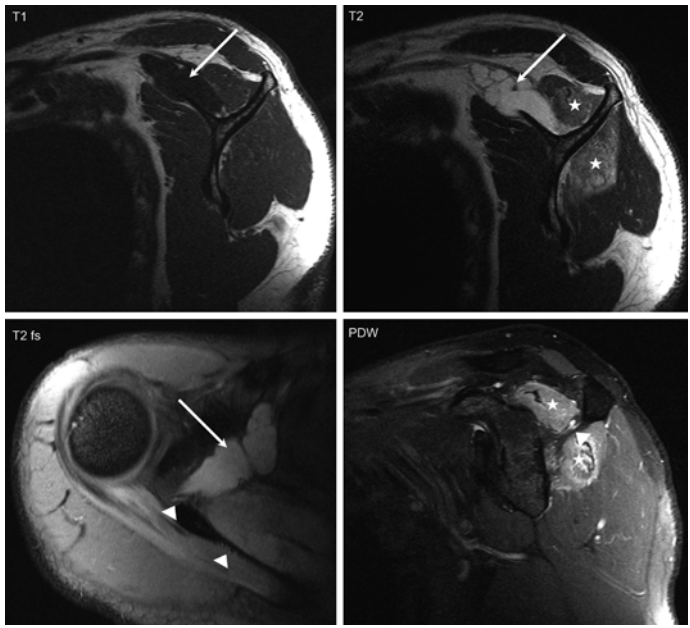


Fig. 8.6 MRI images of suprascapular nerve pathology caused by a ganglion-cyst (*arrows*) compressing the nerve at level of the suprascapular notch. Particularly on PDW images the nerve appears hyperintensive (*arrowhead*). Note also the denervation changes both of the supraspinous and infraspinous muscles (*asterisk*) compatible with a compression at the suprascapular notch (Images courtesy of Bergit Boy, MD and Marc Ewing, MD (department of radiology, Friederikenstift Hannover))

the next sub-periostal approach, patient again in supine position, supraspinous muscle belly kept upwards [71, 72]. We personally prefer the latter for two reasons: first, a transmuscular approach with splitting of the supraspinous muscle belly may damage microscopically small intramuscular branches of the supraspinous main nerve branch, and, second, early identification of the infraspinous branch and its course through the spinoglenoid notch is allowed. The frequent muscle belly atrophy facilitates

the approach [73], and it provides enough space to follow the infraspinous branch to proximal in order to identify the suprascapular branch which runs more to lateral. The suprascapular notch usually lies more laterally than was at first thought. Early on we use the microscope after skin incision parallel to the spina scapulae, and after transection of the fascia near the spina scapulae, consequently at the moment when the supraspinous muscle fibers come into view, and when the subperiostal approach starts. The downwards running infraspinous nerve branch is the first nerve structure you can identify at the bottom of the supraspinous groove. It is embedded in fat tissue under the muscles fibers. Spreading movements of the scissor tips have to be as careful as possible so as not to injure the nerve. As a last step, transection of the superior transverse ligament is carried out.

Isolated infraspinous nerve branch entrapments have been reported [57, 74]. Incision, patient's positioning and approach along the infraspinous surface of the spina scapulae comparable to the approach along the supraspinous surface should be suitable.

Pretty often, ganglia arising from the glenohumeral joint may be found to be similar to these manifestations near the tibiofibular joint. The pathogenesis of these ganglia remains controversial, but there is more and more evidence that occurrence of intra- and extraneural ganglia is connected with the articular nerve branch, here the branch to the glenohumeral joint. Therefore, the surgeon has to address and ligate this connection between joint and ganglion [74]. Nevertheless, a high rate of recurrence is known among surgeons at every location where the ganglia can occur. Our last three cases with multi-cystic ganglia in the supraspinous fossa were punctured CT (computed tomography)-guided. Re-puncturing, if necessary, is less risky than reoperation.

Not to be ignored is the rare possibility that ganglia may also be located within the spinoglenoid notch [74]. Imaging then has

to decide between puncturing or surgical approach through the supraspinous or the infraspinous fossa. In any case, imaging is mandatory before suprascapular nerve entrapment surgery starts, namely, because of the relatively high frequency of space occupying ganglia in the region near the glenohumeral joint.

After open surgery, the fascia of the supraspinous or infraspinous grooves has to be reattached at the spina scapulae. The suction drain can be removed 1 day later.

8.7 Axillary (Circumflex) Nerve

The axillary (circumflex) nerve can rarely suffer from a focal entrapment within the so-called quadrilateral space. Most axillary nerve lesions are instead related to trauma such as subcondyle humerus fractures or dislocations with luxation of the glenohumeral joint. The resulting severe functional loss needs to be examined after a few months and very frequently nerve repair by grafts.

Alternatively, frequent functional involvements happen in the context of the inflammatory Parsonage-Turner syndrome as described in detail in Sect. 10.3. Imaging modalities for assessing the differential diagnosis are discussed in Sects. 6.1.8 and 6.2.8. Initially, very severe shoulder girdle pain is noted, then weakness. Recovery takes several months.

8.7.1 Clinical Symptoms

The few entrapments that were observed as single cases and that were reported in the literature must be mentioned here [75, 76]. Over-trained muscles are described as origin. The quadrilateral space is formed medially by the triceps, laterally by the humerus bone, above by the teres minor muscle and below by the teres

major muscle. Patients' complaints consisted of pain and numbness in the distribution area of sensory axillary fibers at the lateral aspect of the shoulder. The symptoms could be provoked by lateral rotation of the shoulder or hyperabduction. With the aid of today's modern imaging techniques, functional MR-angiography might delineate occlusion of the posterior circumflex humeral artery in abduction and lateral rotation of the shoulder joint.

8.7.2 Electrodiagnostics and Imaging

Lesions of the axillary nerve can be assessed by means of motor nerve conduction study using Erb's point for surface stimulation. Placing the surface recording electrodes over the deltoid muscle, the upper limit of distal motor latency is 5.4 ms and the lower limit of compound muscle action potential is 4,6 mV with an upper limit of normal decrease in amplitude from one side to the other of 54 % [61]. Furthermore in the case of axonal loss, needle electromyography helps to differentiate from other conditions (upper brachial plexus lesion, C5 root lesion) due to analysis of the denervation pattern in different and neighbouring muscles. Additionally, myography can demonstrate whether reinnervation has started or not [49].

Ultrasound imaging of the axillary nerve can be performed via a posterior approach at the proximal part of the upper arm. Within the neurovascular space bordered by teres minor, deltoid, and triceps muscles as well as the shaft of the humerus, the nerve can be localized and successfully blocked with local anaesthesia beside the posterior circumflex artery (Fig. 8.7). However, it is rather difficult to visualize the small nerve directly, and hence there are no normal values of CSA available [77]. Recently a new sonographic approach has been described turning the upper-arm in a position of abduction and eversion. Traumatic lesions of the axillary nerve could be assessed using this method [78]. For that reason, 3 T-MR imaging seems to be the better approach to examine the whole course of the axillary nerve and its damaging factors [79].

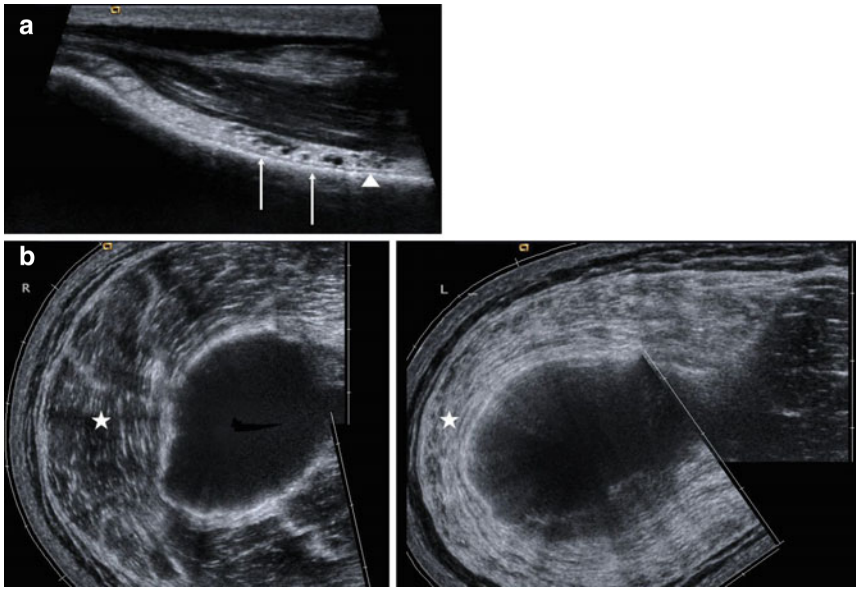


Fig. 8.7 (**a**) Ultrasound imaging of the axillary nerve performed via a posterior approach at the proximal part of the upper arm. Within the neurovascular space bordered by teres minor, deltoid, and triceps muscles as well as the shaft of the humerus, the axillary nerve (*arrowhead*) and the accompanying vessels (*arrows*) are clearly to be seen. (**b**) Denervation changes of the left deltoid muscle (*asterisk*) compared to the unaffected right side in a patient after motorcycle accident (panoramic view)

8.7.3 Treatment

The suitable surgical approach can best be derived from the atlas of peripheral nerve surgery by Kline et al. [80]. Nevertheless, the quadrilateral space is much more deeply located than thought when looking at illustrations of atlas specimens. Each patient reported in the literature was a sportsman with hypertrophic musculature. Experience with the approach from behind to the axillary nerve is described in our previous book, referring to trauma related nerve lesions where a bi-portal approach is always applied [81].

8.8 Musculocutaneous Nerve

With regard to the purpose of this book, a short overview will be sufficient because entrapment pathologies of the musculocutaneous nerve are unknown. Most of its fibers arise from root C6, and run within the lateral cord which divides at the level of the median fork. The musculocutaneous fibers turn to lateral and pierce the coracobrachialis muscle. Its fibers supply the biceps and brachialis muscles, and they contain sensory fibers for the lateral cutaneous nerve of the forearm.

The nerve has great importance in the case of brachial plexus reconstructive surgery as the target nerve of primary value. Injuries which hit the nerve in its upper arm course are either of a perforating character or happen in the context of proximal humerus fractures. Exploration and nerve repair is then almost ever indicated.

Difficulties can arise when the surgeon is confronted with the anatomical anomalies that were described in Sect. 2.1 and which we found in several cases: the musculocutaneous fibers do sometimes not separate from the tateral cord proximal the shoulder joint level, but instead arise from the median nerve still

at upper arm level. Similar observations are also described in literature [82]. We found these anomalies in search of the musculocutaneous nerve as the target in brachial plexus repairs.

The musculocutaneous nerve can quite frequently be severely be affected by inflammatory neuropathies such as the Parsonage-Turner syndrome as described in Sects. 6.2.8 and 10.3. Surgical decompression without some injury in the patient's history is actually never indicated because, as mentioned, entrapment-related spontaneous deterioration of musculocutaneous function never occurs.

8.8.1 Electrodiagnostics

Lesions of the musculocutaneous nerve can be assessed with a motor nerve conduction study using Erb's point for surface stimulation. Placing the surface recording electrodes over the biceps brachii muscle, the upper limit of distal motor latency is 5.6 ms and the lower limit of compound muscle action potential is 4.0 mV with an upper limit of normal decrease in amplitude from one side to the other of 33 % [61]. In addition, the terminal branch of the musculocutaneous nerve (lateral antebrachial cutaneous sensory nerve) can also be examined with a sensory nerve conduction study [83]. Furthermore, in the case of axonal loss, needle electromyography helps to differentiate from other conditions (upper brachial plexus lesion, C6 root lesion) due to analysis of the denervation pattern in different muscles. Additionally, electromyography can demonstrate whether a reinnervation takes place or not [49].

8.8.2 Imaging

The whole course of the musculocutaneous nerve can easily be visualized with high resolution ultrasound. At axilla level, the

nerve enters the coracobrachialis muscle and runs within it (Fig. 8.8). After exiting the coracobrachialis the nerve lies between the brachialis and the biceps brachii muscles. Even its terminal branch (lateral antebrachial cutaneous sensory nerve) may possibly be visible, piercing the superficial fascia at elbow level in non-obese individuals (Fig. 8.9). The mean CSA of the

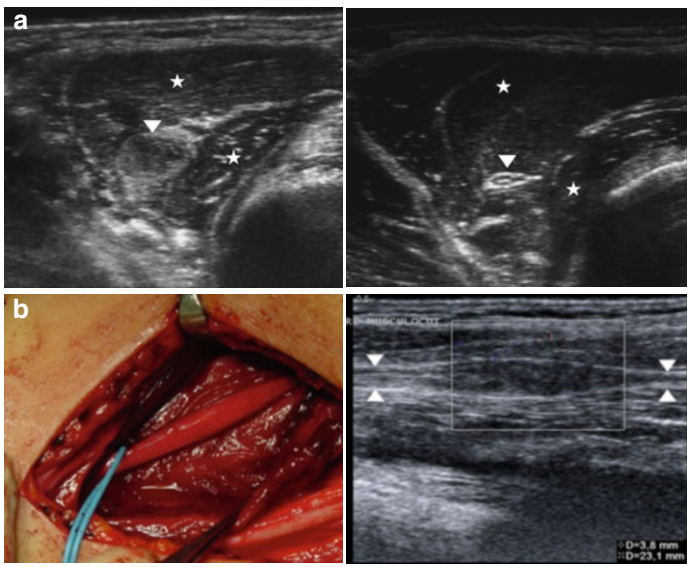


Fig. 8.8 (**a**) Cross sections of musculocutaneous nerve (*arrowhead*) piercing the coracobrachialis muscle (*asterisk*) at the axilla. Note the destroyed echotexture and nerve swelling as well as muscle denervation alterations after major lesion in a young man after motorcycle accident at the left side. In comparison to that at right side normal nerve structure preserved. (**b**) Corresponding intraoperative images and longitudinal sections of the pathologically altered nerve. Note the focal, fusiform, non-vascularised nerve enlargement, representing a neuroma in continuity (Intraoperative image courtesy of Ralph Schoen(MD), department of neurosurgery, Hospital Dessau-Rosslau)

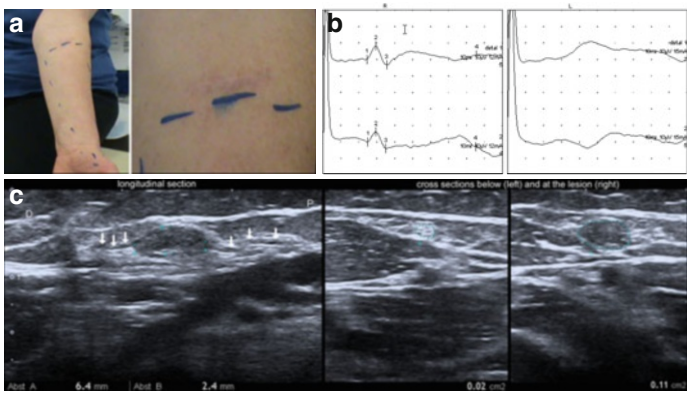


Fig. 8.9 Major lesion of the left lateral antebrachial cutaneous sensory nerve after suicide attempt. (**a**) Clinical representation of numbness below the scar. (**b**) Nerve conduction studies revealed an absent sensory nerve action potential (SNAP) of the left lateral antebrachial cutaneous sensory nerve. (**c**) Ultrasound images demonstrating the neuroma in continuity and the normal appearance of the nerve below the lesion below the lesion (*arrows*: lateral antebrachial cutaneous nerve enters and exits the neuroma in continuity)

musculocutaneous nerve was reported as 2.5 ± 0.4 mm^2. Owing to its small size and out-of-plane course, the MCN may be more reliably depicted with ultrasound imaging than with MR imaging [84]. It should be mentioned that many anatomical variations of the musculocutaneous nerve were observed [85]. Ultrasound can show them before performing a nerve block or another intervention. Taking into account the scientific literature and our own experience with different traumatic lesions, a neuroma especially in continuity is easily detectable using sonography [84]. Moreover, we could observe a single case of external compression both of the median and musculocutaneous nerves at the axilla due to an arm prosthesis (Fig. 8.16). If ultrasound imaging is not available, 3 T-MR imaging may be a sufficient alternative, especially if administration of contrast agent is necessary [79].

8.8.3 Treatment

Focal musculocutaneous entrapment lesions commonly do not occur, whereas traumatic lesions of this nerve appear rather frequently. But the treatment of nerve injuries is not a main purpose of this book, therefore we refer here to nerve trauma literature [80].

8.9 Radial Nerve at the Upper Arm

8.9.1 Anatomy

After separation of the axillary (circumflex) nerve from the posterior cord, remaining nerve fibers continue as radial nerve into the medial aspect of the upper arm. Behind the humerus, the nerve transverses through the spiral groove to the lateral side of the upper arm. A few centimeters distal it pierces the lateral intermuscular septum. Two or three proximal branches which supply the triceps muscle arise from the radial nerve below the armpit, whereas, after running transverse behind the humerus, the first motor branch runs to the brachio-radial muscle. Knowledge of these anatomical details helps to interpret clinical findings and to assess the location of the nerve lesion.

8.9.2 Damaging Factors

In terms of entrapment neuropathies, focal neuropathies at upper arm level occur rather rarely. The radial nerve may be easily compressed against the humerus bone within the spiral groove. Popular terms to describe this mechanism are “saturday night palsy” or “park bench palsy”. Be that as it may, an entrapment behind the humerus is always related to a special incident, and it never occurs spontaneously.

Quite another and extremely rare mechanism of nerve lesion can affect the radial nerve at the site where it pierces the lateral intermuscular septum. We have observed a case of spontaneous nerve rotation with hourglass-like aspect intraoperatively. Literature is full of single case reports of nerve rotation. Affected nerves were the radial nerve but also the posterior interosseus and anterior interosseus nerves [86, 87]. The single case of rotated radial nerve trunk we experienced had no special incident as possible explanation in its history [88]. Nevertheless, a spontaneous origin seems to be inconceivable. The latest scientific findings perhaps suggest a real context with hereditary forms of the Parsonage-Turner syndrome to which we refer in Sects. 6.2.8 and 10.3.

Trauma- or injury-related radial nerve lesions are commonly due to humerus fractures. Almost always the triceps muscle function remains preserved despite severe injury to the extremity. It will surely become of great interest if, particularly behind the humerus, imaging techniques and further improvements can distinguish between nerve continuity or discontinuity; furthermore, question of intraneural reactive scarring on one hand, or of the nerve gap distance after disruption on the other, will become an issue in future.

8.9.3 Clinical Symptoms

Loss of triceps muscle function indicates a highly proximal lesion, preservation of triceps function but loss of brachio-radial muscle function indicates a lesion between the two branching levels. Therefore, the clinical differentiation concerning the level of lesion should not be difficult. Electromyography supports the clinical assessment, and it can moreover differentiate between partial or complete functional loss, a question of great importance for deciding adequately whether surgical treatment is indicated.

8.9.4 Electrodiagnostics

Electrodiagnostic workup in radial neuropathy of the upper arm includes motor and sensory nerve conduction studies, needle electromyography and motor and sensory nerve conduction studies of the ulnar nerve and the median nerve in order to confirm the isolated neuropathy of the radial nerve and to exclude other kinds of neuropathies. A fractionated motor nerve conduction study with recording from the extensor indicis proprius (EIP), stimulating at forearm, at elbow, as well as below and above the spiral groove bilaterally, is recommended. With surface electrodes the normal CMAP recorded from the EIP is 2–5 mV. In the case of axonal loss 3–5 days after the lesion, a decreased distal CMAP results when comparing the involved side with the contralateral one. Measuring CMAP with surface electrodes often results in an initial positive deflection; therefore, this testing only allows approximations to solve the question of axonal loss. It presents additional difficulties when side-to-side comparison with neighboured radial muscles is tried, because the EIP is not a well-isolated muscle. Compared to the median and ulnar nerve, the radial nerve has a less straight course; this fact results in difficulties in the distance measurement. Together with the initial positive deflection of the CMAP, it can lead to considerable inaccuracies in measuring true conduction velocities. Therefore, the conduction velocity is not of real value concerning the radial motor function, but a focal conduction block between the proximal and distal sites and the determination of the relative CMAP amplitude are useful to assess eventual axonal loss [89].

In contrast to motor conduction evaluation, study of the sensory superficial radial nerve (SRN) is easy to perform. The most common technique is the antidromic excitation of the nerve 10 cm proximally on the distal radius while recording takes place distally from the first digit with ring electrodes. Comparing the SNAP on both sides is important. A decreased or absent

SNAP suggests axonal damage at some level of the superficial radial nerve fibers. An interesting phenomenon in exclusively or predominantly proximal demyelination cases can be a normal SNAP although the patient reports marked numbness in the distribution of the superficial radial nerve [89]. Normal SNAP of the SRN may also occur in incomplete axonal proximal radial nerve lesions which spare fibers of the SNR on one hand [90], and particularly in the case of exclusive axonal loss in the posterior interosseus nerve (PIN) on the other.

Needle electromyography examination is required for a relatively accurate determination of lesion location and differential diagnosis. In PIN abnormalities the lesion will be limited to those muscles innervated by the posterior interosseus nerve (extensor indicis proprius, extensor digitorum communnis and extensor carpi ulnaris). In radial neuropathies at the spiral groove, brachioradialis and the long head of the extensor carpi radialis are involved in addition to PIN-innervated muscles. If the lesion is at the axilla, the triceps muscle is included in the lesion. A far proximal lesion of the posterior cord shows additional abnormalities which include the deltoid muscle (axillary nerve) and latissimus dorsi muscle (thoracodorsal nerve). In plexus brachialis lesions the cutaneous antebrachii lateralis nerve may also be involved. A C7/C8 radiculopathy will show abnormalities of the cervical paraspinal muscles and of non-radial-innervated C7/C8 muscles (pronator teres, flexor carpi radialis, interosseus dorsalis manus I) and a normal neurography of the sensory SRN.

8.9.5 Imaging

High resolution ultrasound can visualize the origin of the nerve injury, and it helps to locate the lesion by examining the entire nerve course from the axillary region down to the wrist. The nerve is easily detected in a transverse scan at the lateral

aspect of the mid-humerus and can be followed from that point upward and downward [91]. Color Doppler sonography allows the visualization of the accompanying deep brachial artery, which serves as an anatomical landmark to identify the nerve. HRUS was meanwhile even proven to be useful for accurate evaluation of patients with nerve palsy associated with humeral shaft fracture [91]. Electrodiagnostic studies are instead less able to localize the site of proximal radial neuropathies [92]. Following a trauma, HRUS can assess primary lesions related to the trauma (e.g. a major lesion concerning Sunderland Grade V with discontinuity of the nerve after transection or severe traction) or secondary reactive alterations during the healing process of the humerus fracture (e.g. callus compression or nerve riding on a metal plate). Comparable cases are shown in Figs. 6.8b and 8.10b. In our experience, the typical compression syndrome among drinkers ("Saturday night nerve palsy") causes either no changes detectable by high resolution ultrasound or a segment of the radial nerve at the site of compression with normal nerve caliber proximally and distally to it (Fig. 8.10a), whereas we never detected a focal entrapment at upper arm level with spontaneous onset so far. However, important is the fact that, in some non-traumatic and non-entrapment radial nerve palsies, HRUS can demonstrate either unspecific long-distance thickening of nerve fascicles and caliber changes (e.g. as a result of brachial plexus neuritis or diabetic mono-neuritis multiplex) or specific morphological changes such as peripheral nerve sheath tumors, tumor like lesions, or even a torsion or rotation neuropathy (see Chap. 12). Figure 8.10b illustrates nerve damage due to trauma, Fig. 8.10c a malign nerve sheath tumor, and Fig. 8.11 a nerve torsion. We experienced the curious radial nerve torsion neuropathy as characterized by an hourglass-like stenosis with congestion of the nerve more proximally from it. Similar changes have been described by other authors [93].

3 T-MR neurography is also suitable for the diagnosis of radial neuropathies. Especially in early stages of the lesion it already shows a T2 lesion of fascicles [94, 95]. The advantage is that this type of imaging makes it possible to administer

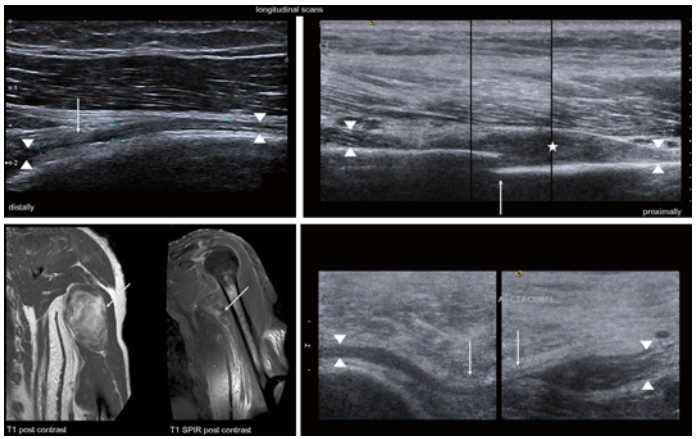


Fig. 8.10 Several lesions of the radial nerve at upper arm. (**a**) Saturday night palsy in HRUS. Longitudinal scan along the radial nerve (*arrowhead*) at the spiral groove shows spindle-like swelling of the radial nerve (caliber 2 mm, proximal of it only 1.2 mm) above the site of compression (*arrow*). (**b**) HRUS, fracture of the humerus (*arrow*) with a bone fragment piercing the radial nerve (*arrowhead*) with secondary development of a neuroma in continuity (*asterisk*). (**c**) MRI, Malign peripheral nerve sheath tumor of the radial nerve in a patient with Neurofibromatosis type I before and after resection (*left*). HRUS; scarring after excision-induced nerve entrapment (*arrows*) of the radial nerve (*arrowheads*) (MR images courtesy of Karsten Stock, MD (department of radiology, Dessau-Rosslau))

contrast agent if a tumor is suspected. However, the majority of cases do not need such expansive means.

8.9.6 Treatment

Spontaneous radial nerve trunk entrapments behind the humerus are not to be expected as just mentioned. In our experience, surgical exposure following a supposed nerve compression was never indicated. The same holds true in the literature.

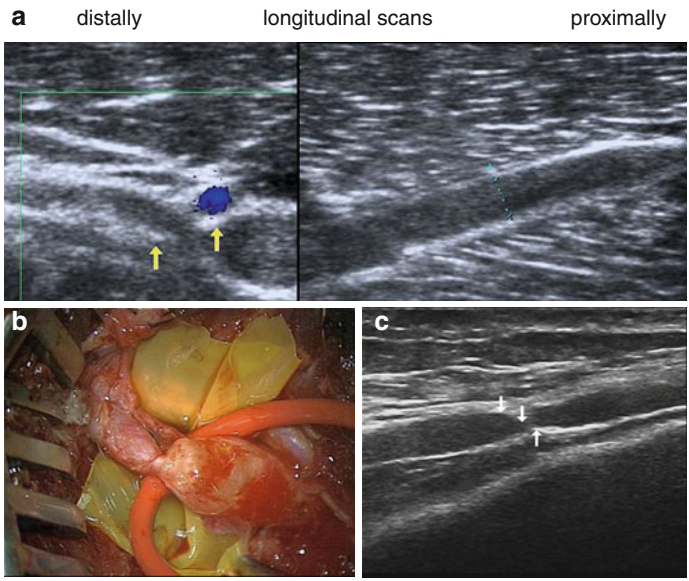


Fig. 8.11 HRUS, multiple torsions of the radial nerve. (**a**) Substantial caliber change and hourglass-like constriction (*arrows*) with deep brachial artery just above the constriction as well as long-segmental enlargement of nerve diameter proximal to the constriction (3.6 mm). (**b**) Corresponding intraoperative image. Only after removal of adventitia the twisted constriction and proximal bulging of the nerve is clearly visible (Image courtesy of Thomas-Nicolas Lehmann, MD (department of neurosurgery, Hospital-Bad Saarow/Germany)). (**c**) A second distally located spontaneous torsion of the radial nerve in the same patient month after surgical repair of the torsion depicted in (**a**) and (**b**) (*arrows*: site of nerve torsion)

The very rare nerve rotation cases have hardly an incident in patient history, and in contrast to missing causative factors, a complete functional loss mostly from the brachio-radial muscle downwards. The symptoms occur suddenly, and nerve exploration, e.g. via a longitudinal incision at the lateral aspect of the lower half of the upper arm, is unavoidable. To facilitate the

exposure, the surgeon can start at the level of the elbow skin crease. A direct approach between the brachio-radial and biceps muscle bellies and the biceps tendon is suitable to identify the radial nerve most easily. From that point, the exposure has to progress in a proximal direction. An hourglass-like nerve aspect, if it really has been found, needs a microneurolysis under microscope and mostly a nerve repair by grafts as demonstrated in Chap. 12 and [88]. It should be noted that graft repair of the radial nerve carries the best prognosis of all extremity nerves.

8.10 Posterior Interosseus Nerve

8.10.1 Anatomy

Several aspects of anatomy are of great clinical importance and must be known: the radial nerve runs downwards between the biceps tendon and the brachioradial muscle, the superficial radial nerve branch separates at different levels, the posterior interosseus branch enters the supinator channel under a tendon-like arcade which is referred to as the arcade of Frohse. This is the most common site of focal nerve entrapment. Different names, depending of the site of compression, are common: "PIN syndrome" or "supinator syndrome" [96]. The entrapment results in pure motor deficits. It is characterized by loss of ability to extend the digits as well as radial wrist deviation due to weakness of the extensor carpi ulnaris but preserved wrist extension because the short radial wrist extensor remains supplied. This muscle mostly gets a small branch that separates from the superficial nerve branch or from the main radial trunk. A very small sensory branch runs downwards supplying a small skin area between the first and second digit on the extension side.

"Radial tunnel syndrome" is sometimes said to be a distinct clinical entity without motor weakness and occurring as a result

of repetitive motion injuries to the elbow [97]. The term "algetic supinator tunnel syndrome" is unfortunately confusing and should not be used [98]. It is associated with a tendinosis such as epicondylitis humeri radialis.

8.10.2 Damaging Factors

Focal compression sites from proximal to distal include fibrous bands from the radio-capitellar joint, the tendinous edge of the extensor carpi radialis brevis muscle adjacent to the radial recurrent artery and its branches (leash of Henry), the arcade of Frohse (tendinous proximal end of supinator), and a fibrous band at the distal end of supinator muscle [96, 99]. Entrapment neuropathies of the PIN occur frequently. The patient normally does not remember any incident and not even the time of weakness onset. Sometimes a lipoma compresses the nerve branch against the arcade of Frohse. It remains controversial as to why such lipomas emerge exactly under the supinator muscle. They are accidentally found during surgery, or they are evaluated by HRUS (high resolution ultrasound) or MRI (Multi-Resonance-Imaging) when done pre-operatively. PIN palsy can be a very rare complication of rheumatoid arthritis affecting the elbow joint. Eighteen cases of PIN palsy due to rheumatoid arthritis have been reported so far in the literature [100]. Trauma related PIN palsies result from radius fractures or its osteosynthesis. The functional loss is then almost ever complete.

As a differential-diagnostic problem with the PIN syndrome, with slowly proceeding palsy and without numbness, the rare multifocal motorical neuropathy (MMN) has to be kept in mind. It can quite easily simulate an entrapped motor nerve as described in detail in Chap. 10. PIN torsion (rotation) as a spontaneously occurring incident has been sporadically reported in literature [86, 101]. We observed one case with multi-segmental torsions of the radial nerve, including one incident in the supinator tunnel. Not to be

ignored is the fact that severe PIN palsies can generally be the single manifestation of a Parsonage-Turner syndrome, sometimes even with nerve torsion (see Sects. 6.2.8 and 10.3; Chap. 12). We have experienced in two such patients initial and heavy shoulder pain as important anamnesis factor and, following these attacks, a relatively sudden weakness.

8.10.3 Clinical Symptoms

Lesions of the posterior interosseus nerve as related to entrapment etiologies occur spontaneously and deteriorate slowly. Numbness is never noted. Wrist extension remains preserved to some extent because of functional continuity of the short radial extensor muscle. The physician observes a deviation of the wrist to radial because the ulnar wrist extensor is included in the impairment. This combination of preserved and impaired functions has to lead to the correct diagnosis. In case of forearm fracture and secondary osteosynthesis, complete functional loss results rather frequently. It will then become an important challenge of imaging techniques to answer questions about nerve branch continuity or discontinuity. However, osteosynthesis material might limit its capabilities.

8.10.4 Electrodiagnostics

Electrophysiological techniques exist to assess the conduction through the radial tunnel. In one of these techniques, motor latencies of the brachioradialis and extensor carpi ulnaris muscles were compared after stimulation of the radial nerve either at the axilla or above the elbow. The recording was performed with a coaxial needle electrode after percutaneous axillary stimulation and with surface electrodes after near-the-nerve elbow stimulation. A latency difference between these two

muscles of 1.3 ms represented the posterior interosseous nerve conduction in the radial tunnel; the normal range of the side-to-side difference was limited to only 0.4 ms after elbow stimulation [102]. In another technique the radial nerve was stimulated at the elbow between the biceps brachii and brachioradialis muscles at three different positions of the forearm (neutral, maximal forearm supination and maximal forearm pronation). During the recordings the forearm had been in the required position for 30 s. In controls the inter-positional latency was less than 0.12 ms, and in patients with the clinical diagnosis of radial tunnel syndrome was 0.44 ms. Following radial nerve decompression, differential motor latencies in the test group decreased below control values. A differential latency of ≥0.30 ms was considered indicative of PIN syndrome [103]. In our opinion, however, those techniques are unfortunately of little practical value due to complicated application of the first method described and due to the questionable replication of findings using the latter technique. We need further studies to explain the mechanisms of conduction slowing during short periods of assumed positional compression.

The best technique to localize the lesion is needle EMG which helps to differentiate from other conditions (upper brachial plexus lesion, C7 root lesion, a more proximal situated lesion of the radial nerve trunk) due to analysis of the denervation pattern in different muscles. It can again demonstrate whether a reinnervation takes place or not. Pure PIN lesions have a normal superficial radial nerve SNAP.

8.10.5 Imaging

The whole course of the PIN can be rather easily visualized with high resolution ultrasound. To identify the posterior interosseous nerve, we begin imaging the nerve in the transverse plane at the brachioradialis muscle. There we intend to show the bifurcation of the radial trunk into its superficial and deep components. The deep branch penetrates the supinator muscle and continues

as the posterior interosseous nerve. According to our experience, at the entry site under the supinator muscle, the posterior interosseus nerve changes its tubular shape into a more flattened one. As a consequence, its separate fascicles are located side by side one behind the other. This localized appearance may mimic a compression [104]. The normal value of the posterior interosseus nerve derived from a prospective study is 2.3 ± 1.3 mm^2 [105]. A cut-off value of >1.5 mm of the anterior-posterior diameter at the level of the arcade of Frohse should be specific detecting a PIN-syndrome [106]. However, sonography can detect space-occupying lesions such as lipomas or ganglion cysts beside the nerve (Fig. 8.12c). Comparable to other entrapment neuropathies, swelling of the nerve proximally to the

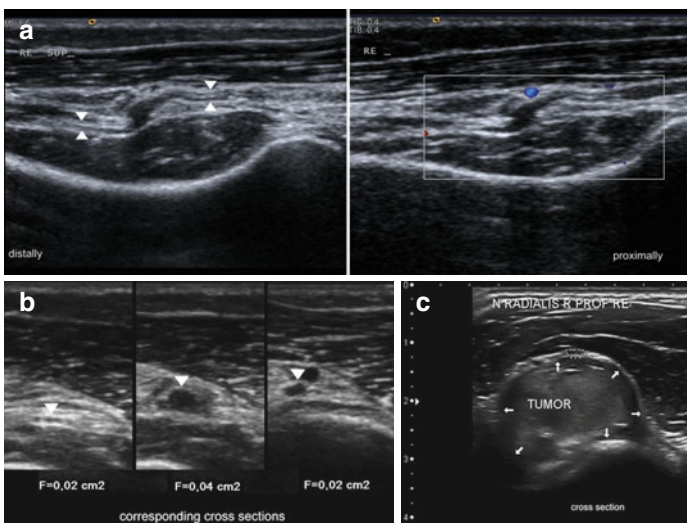


Fig. 8.12 Pathologies of the posterior interosseus nerve in HRUS. (**a**) Entrapment of the PIN (*arrowheads*) at the entry of the supinator muscle, left greyscale image. On the *right* colour-Doppler image of the arcade of FROHSE (*blue dot*) is clearly to be seen. Note the abrupt nerve swelling at the entry of supinator muscle. (**b**) The corresponding cross sections. (**c**) Ganglion-cyst compresses the PIN (*arrows*: margin of the ganglion – cyst). Courtesy Dr. L. Schelosky, Switzerland

compression site can at least be detected (Fig. 8.12a, b). According to the literature, focal nerve swelling just proximal to a fibrous band [107] respectively before entering the supinator muscle [108], or an extended swelling due to a tight band of aponeurotic origin [109] could be demonstrated by sonography and later surgically verified. The main diagnostic criterion is, of course, a swelling of the nerve proximal to the arcade of Frohse if existent [110]. Prospective studies are needed to establish diagnostic criteria for PIN syndrome based on nerve dimensions. If ultrasound imaging is not available, 3 T-MR imaging may be a sufficient alternative. MR neurography findings include enlarged and abnormally T2 hyperintense posterior interosseous nerves with regional muscle denervation edema (AE) in the forearm compartment [111].

8.10.6 Treatment

Surgical treatment of entrapment neuropathy of the posterior interosseus nerve is done via a skin incision, preferably S-shaped and including the elbow crease. We start at the distal lateral upper arm region. Blunt exposure between the brachio-radial muscle and the biceps easily leads to the radial nerve. From that point we proceed to distal. Great care has to be taken with exploration of the superficial and deep branches, and particularly with the small motor branch supplying the short radial extensor muscle. This branch has to be preserved unconditionally; otherwise the patient will lose his preoperatively unimpaired wrist extension. Intra-operative electrical stimulation equipment is therefore mandatory if posterior interosseus nerve exposure is started. Further exploration to distal aims at transecting the arcade of Frohse in order to extend the opening where the nerve disappears under the supinator muscle. The muscle fibers themselves do not compress the nerve and should therefore be preserved. In the case of a lipoma or a ganglion under the muscle, it is depicted during transection of the arcade of Frohse. In such a special case it is necessary to split the supinator muscle belly, to

take a microscope, and to remove the lipoma with preservation of continuity of the interosseus nerve. The assistant will help by retracting overlying soft tissue, but the surgeon is responsible for preserving the above-mentioned small motor branch to the radial wrist extensor. Small vessels always run transverse to the interosseus nerve course in front of the arcade of Frohse. They have to be identified early enough, coagulated and transected independently whether the operation is done with or without bloodlessness. We recommend a suction drain for 24 h because re-bleeding will surely disturb the nerve function (Fig. 8.13).

Posterior interosseus nerve repair after traumatic nerve damage is comparatively difficult. We use a second incision situated longitudinally at the dorsal aspect of the middle forearm. The extensor muscle fibers are split in order to approach the interosseus membrane on which the final branches and the distal stump of the interosseus nerve lie. The bi-portal approach has the advantage that the supinator muscle and the overlying muscles remain untouched. Grafts for nerve repair are drawn through the supinator tunnel and nerve coaptation under the microscope takes place proximally and distally via both separated approaches. As the time period that can be allowed for the removal of osteosynthesis material of the lower arm is at least 1 year, and as nerve repair, in contrast, does not allow waiting for this length of time, the osteosynthesis has to remain in place for the whole life in these cases.

8.11 Superficial Radial Nerve

8.11.1 Anatomy

The superficial radial nerve branch passes, as already mentioned, above the supinator muscle and then deep into the brachio-radial muscle. At the level where watch straps are normally worn, the nerve pierces the lower arm fascia and runs downwards

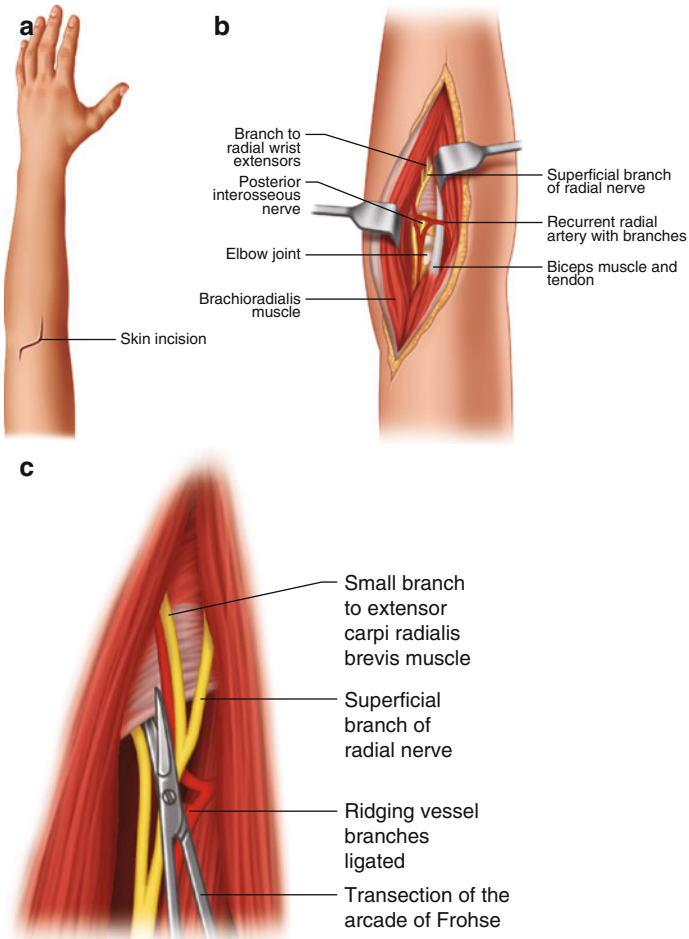


Fig. 8.13 Surgical treatment of posterior interosseus nerve entrapment. (**a**) Skin incision. (**b**) Exposure, note the branch to medial wrist extensors separates before the supinator tunnel. (**c**) Transection of the roof of the supinator tunnel

superficially to supply the main part of the wrist's and digits' skin at their dorsal aspect. The subcutaneous course makes it vulnerable to perforating injuries, whereas the site of piercing makes it sensitive to entrapment.

8.11.2 Damaging Factors

Pure entrapment neuropathy – "Cheiralgia paraesthetica" – is due to tight bands around the wrist. In contrast to that, perforating injuries may hit the subcutaneously situated nerve and its several skin branches as resulting from cutting injuries or iatrogenically from vessel canulations. Another cause of compression and an important differential diagnosis is the de Quervain tenosynovitis. Swelling of tendons of the abductor pollicis longus as well as the extensor pollicis longus and brevis muscles may compress the nerve. The Finkelstein test is positive in both cases and creates assessment difficulty if pain is the only symptom. On the other hand, numbness in the distribution area of the superficial radial nerve as well as neuropathic pain symptoms clearly indicates nerve involvement.

8.11.3 Clinical Symptoms

The nerve consists exclusively of sensory fibers. Therefore, pain is the leading symptom which usually radiates into the whole distribution skin area. The resulting discomfort is severe, and daily activities are massively inhibited.

8.11.4 Electrodiagnostics

There are two major antidromic stimulation techniques that have been described for this nerve. In the first antidromic technique,

the recording surface electrode is positioned over the superficial radial nerve at the wrist and the second electrode over the second metacarpal bone on the dorsum of the hand. The stimulation can be performed 10–14 cm proximal to the wrist over the lateral margin of the radius. The onset latency is 1.8 (10 cm) – 2.4 (14 cm) ± 0.3 ms and the amplitude of the SNAP is 31 ± 20 μV [115]. In the second antidromic technique, a ring electrode is positioned proximally over the proximal portion of the first digit and distally near the thumbnail. Because the terminating fibers are small, the resulting SNAP is lower (12 ± 8 μV) if compared to that of the above-mentioned method [116]. Stimulating the nerve proximally in the ante-cubital fossa and distally 10 cm proximal to the recording site at the base of the first digit allows one to determine the distal sensory latency and a forearm sensory nerve conduction velocity (SNCV) (62.1 ± 4.2 ms, left/right difference <7 m/s) [117]. Unfortunately the diagnostic and localizing value of electrodiagnostic studies of the superficial radial nerve was found to be low because, in 30 % of the cases, the SNAP was absent or the amplitude was diminished [118].

8.11.5 Imaging

The radial nerve is easily visualized with high resolution ultrasound as a deep and superficial branch between the brachioradialis and brachialis muscles at the elbow. Although small in size, the superficial branch can be followed without problems down to the radial styloid; there in our experience, the underlying cause of nerve pathology could be detected, e.g. neuroma after trauma [112] or a granulomatous nerve infiltration or hypervascularity due to perineuritis in a case of cutaneous sarcoidosis (see Chap. 14). Its normal cross sectional area is 0.02–0.03 cm^2 [119]. Besides assessment of neuroma formation of the nerve, other authors reported about neurinoma. In our experience, compressive de Quervain tenosynovitis is also detectable [120]. Two such examples are shown in Figs. 8.14 and 8.15.

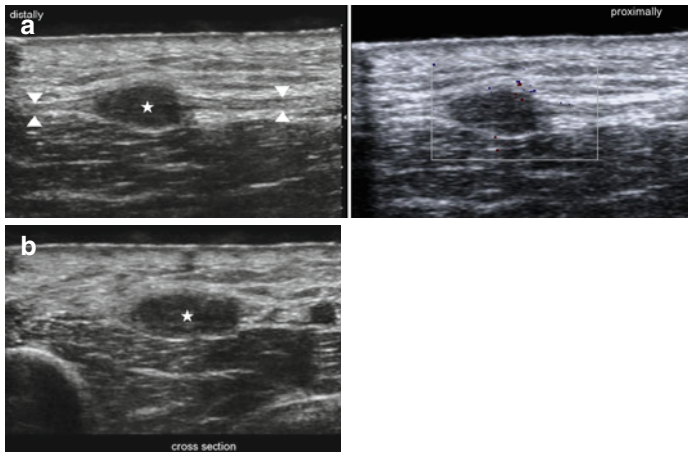


Fig. 8.14 HRUS, neuroma in continuity (*asterisk*) of the superficial radial nerve (arrowheads) after iatrogenic vessel puncture. (**a**) Greyscale and colour-Doppler images in longitudinal sections. (**b**) Cross section

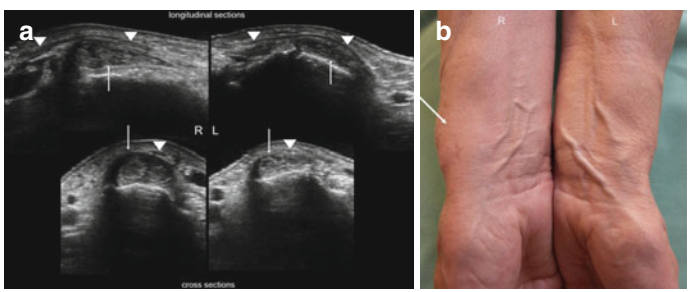


Fig. 8.15 (**a**) HRUS, de Quervain tenosynovitis at the right side (R) compressing the right superficial radial nerve (*arrowheads*). Arrows point on the tendons of the long thumb abductor and the long and short thumb extensor. On the right, swelling and halo is clearly to be seen. Unaffected left side (*L*) is shown for comparison. (**b**) Clinical aspect

8.11.6 Treatment

Depending on the kind of lesion, treatment modalities are easy or extremely difficult. The entrapment pathology needs to mark the point of maximum of the Tinel sign pre-operatively, and then to approach the piercing point of the nerve. The site where the small nerve squeezes through the fascia has to be extended a little. The skin incision is about 2 cm long.

Nerve injury however remains a great challenge. Imaging of the neuroma is easy, but various attempts to treat them are discussed in the literature [112]: nerve stump ligation to stop re-growth of fascicles, re-placing the neuroma in muscle substance after partially shortening the nerve, re-placing the nerve stump into bone, and neuroma resection with reconstruction of nerve continuity by graft [113].

We have been confronted with a great number of patients suffering from pain more than ever following such procedures. Our own experiences with nerve repair by grafts show they resulted in neuropathic pain. The superficial radial nerve distribution is extremely susceptible to resulting in neuropathic pain (see Sect. 4.2). It should be remembered that the neuropathic pain types occur suddenly, they have an early onset, and they achieve steady state at once. Patients may present such kinds of symptoms immediately after the injury or immediately after the first surgical trial. Our experience is that further surgery in the periphery can lead to a severe aggravation of the pain. We therefore prefer peripheral nerve stimulation (PNS) as the method of choice and gave up other peripheral trials completely. The identification of the superficial radial nerve succeeds at elbow level via the already described approach to the supinator tunnel. Intraoperatively, electric stimulation is needed to find the exclusively sensible target nerve [114]. Incidentally, three nerve compression cases were successfully neurolized, whereas one case with already preoperatively existent neuropathic pain remained unchanged. We cannot do anything else but remind everyone to assess the pain type that is present pre-operatively.

8.12 Median Nerve at the Elbow (Pronator Teres Syndrome)

8.12.1 Anatomy

The median nerve starts as a median fork which carries fibers from the lateral and medial cords. It runs downwards in the medial aspect at the upper arm together with the brachial artery. Some literature says that in 1 % a supracondyle process exists which proceeds downwards as a Struther's ligament to the medial epicondyle. Thus, an osteofibrous channel is formed which fixes the nerve during supracondyle humerus fractures [121]. Other authors remain doubtful whether this structure really exists [122]. At elbow level, the median nerve is situated between the brachial muscle and the common head of forearm flexor muscles. It passes between the two heads of the pronator teres muscle.

8.12.2 Damaging Factors

Two rare cases of median nerve compression at levels below the axilla are illustrated in Figs. 8.16 and 8.17. In the first case, entrapment of the upper median nerve was caused by a poorly customized arm prosthesis followed by axonal loss due to the compression of both the median and musculocutaneous nerves (Fig. 8.16). In the second case, intravenous administration of recombinant tissue plasminogen activator after a minor stroke led to a diffuse subepineural haemorrhage in the median nerve compressing its perineurium and fascicles (Fig. 8.17). These examples illustrate that a median nerve entrapment between axilla and elbow commonly needs a severe causative factor.

The nerve is also vulnerable in the case of supracondyle humerus fractures and puncture procedures through the elbow

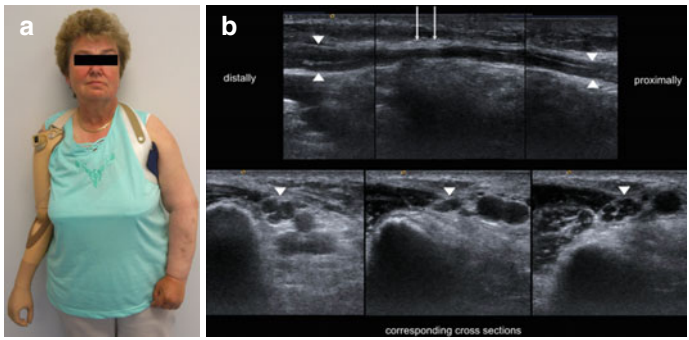


Fig. 8.16 (**a**) Entrapment of median nerve at the axilla by a poorly customized arm prosthesis followed by axonal loss due to the compression of both the median and musculocutaneous (not shown) nerves. (**b**) Corresponding ultrasound images demonstrating a flattening (*arrows*) of the median nerve (*arrowheads*) at the site of compression as well as a nerve swelling proximally and distally to the lesion

crease. Accident or incident related lesions may of course also occur near to the pronator teres. However, the question seems to be justified whether a spontaneously occurring entrapment – the so-called "pronator teres syndrome" – really exists. Millesi only mentions this syndrome by citing literature [121]. Stewart raised doubts with his opinion that nerve compression within the pronator muscle is a "matter of speculation" [123]. Birch describes three of his own cases which were operated on without any motor recovery [124]. He discusses compression possibility at the margin of the aponeurosis of the biceps (lacertus fibrosus). Gelberman writes that there is a "specific inciting event occasionally" [125]. This expression gives reason to believe that

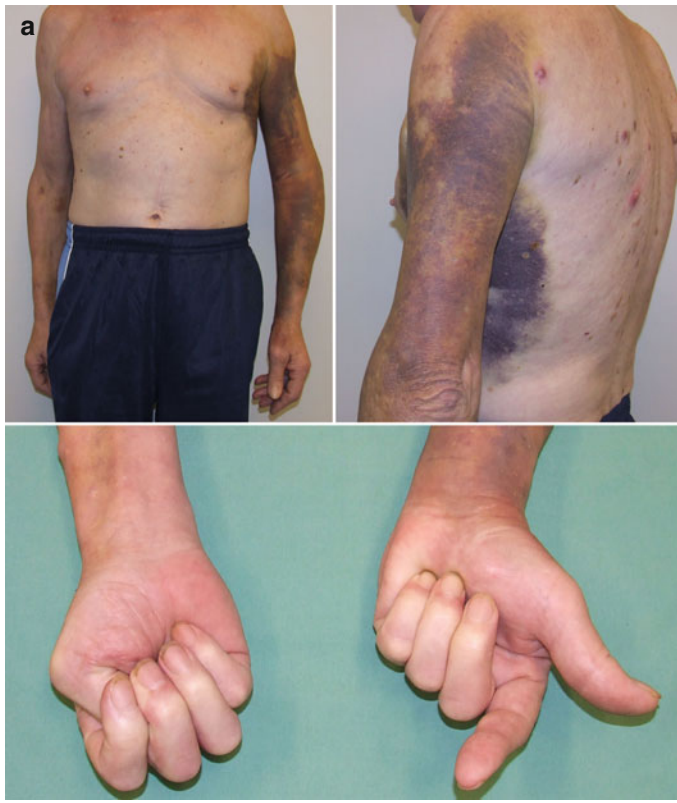


Fig. 8.17 (**a**) Diffuse hemorrhage after intravenous administration of recombinant tissue plasminogen activator in patient with minor stroke leading to a proximal palsy of the median nerve. (**b**) Corresponding greyscale and colour-Doppler images demonstrate a subepineural bleeding (*arrow*) within the median nerve sheath (*arrowheads*) compressing its perineurium and fascicles

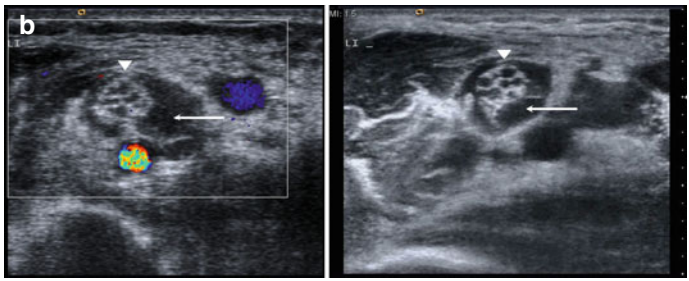


Fig. 8.17 (continued)

entrapment cases with spontaneous onset may sometimes occur. Vogel stated in 2001: "This syndrome is extremely rare; I never observed it during 30 years electromyography practice" (translated from German [126]). We observed a case of an 11-year-old boy developing a supracondyle process after a supracondyle humerus fracture compressed the median nerve between its bony surface and scar tissue (Fig. 8.18). To summarize, injuries to the median nerve mimicking a "pronator teres syndrome" are common, but the existence of real entrapment is disputed.

8.12.3 Clinical Symptoms

Pain occurs within the medial aspect of the forearm, and typical nerve symptoms involve the entire median nerve distribution, the three and a half digits, and all the median nerve supplied forearm and hand muscles. A positive Tinel sign has to be evoked at the side of the expected compression structure with irradiating paresthesias into the three and a half digits.

8.12.4 Electrodiagnostics

The electrodiagnostic distinction of various proximal focal median neuropathies is very challenging. The expected pathological findings for proper localization are inconsistent. In 72 patients with "pronator teres syndrome" only 30 % showed a decreased

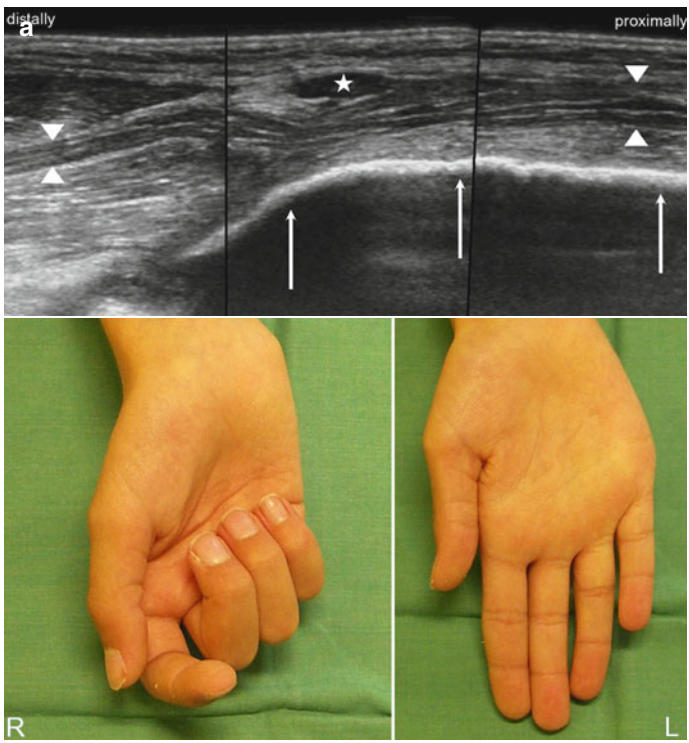


Fig. 8.18 (**a**) Proximal palsy of the right median nerve in an 11-year-old boy after supracondylar humerus fracture. The median nerve (*arrowheads*) is entrapped between a supracondyle process (*arrows*) and scar-tissue (*asterisk*) in HRUS. (**b**) "Supracondyle process" (*arrow*) developed after osteosynthesis (X-ray photograph courtesy of Karsten Stock, MD (department of radiology and neuroradiology, DessauRosslau Hospital/Germany))

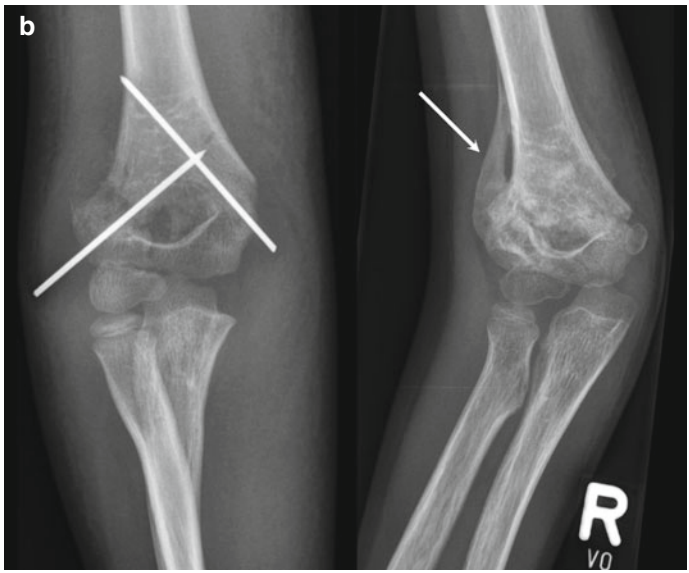


Fig. 8.18 (continued)

median forearm velocity, 65 % had abnormal median sensory factors of either abnormal conduction velocity or amplitude, and 70 % gave pathological EMG findings [127]. Electrodiagnostic studies should exclude other causes of the complaints, e.g. the presence of a concomitant carpal tunnel syndrome, cervical radiculopathy, or brachial plexus lesion or neuritis.

8.12.5 Imaging

The whole course of the median nerve at the elbow and proximal forearm can easily be visualized by means of high resolution ultrasound. Sonography can detect space-occupying lesions, or in the case of a supposed entrapment neuropathy, it should normally demonstrate a focal nerve swelling proximally to the

compression site as we would generally expect in entrapments (Figs. 8.16, 8.17 and 8.18). MR neurography can be used as an alternative method. MR imaging may show denervation-associated changes in specific muscles innervated by the affected nerve. The analysis of the de-innervated muscle distribution, under knowledge of the nerve innervation pattern, can help determine the nerves involved and the levels of nerve lesion [128].

8.12.6 Treatment

First of all, our experience is completely restricted to iatrogenically caused nerve lesions or fracture-related nerve injuries. Spontaneous entrapments were never transmitted. The skin incision should be S-shaped and starts over the palpable groove between the bellies of biceps and brachial muscle. Care has to be taken not to injure the brachial artery which partly covers the nerve course; nerve branches to the pronator teres muscle have to be identified and preserved unconditionally. The lacertus fibrosus has to be transected, and the neurolysis proceeds up to the pronator teres muscle slit. Sometimes, tendon-like ligaments form the margins of the pronator teres slit. These ligaments have to be incised to decompress the median nerve's entrance into the deep forearm compartment. Again, doubts remain as to whether a spontaneous entrapment really exists.

8.13 Anterior Interosseus Nerve

8.13.1 Anatomy

The anterior interosseus nerve branch separates from the median nerve trunk perhaps 1 cm above the pronator teres muscle slit, turns a little to radial and supplies the flexor pollicis longus and flexor

digitorum profundus II and III muscles. Being very peripheral, it also supplies the pronator quadratus muscle. The nerve branch does not contain any sensory fibers for skin supply but, of course, does contain afferent fibers from muscle spindles.

8.13.2 Damaging Factors

If a focal nerve entrapment exists, it has to be localized to the site of fibrous margins of the heads of the pronator teres. Earlier literature discussed a compressing thrombosis of the anterior interosseus artery or of ulnar collateral vessels, an aberrant radial artery, or an accessory head of the flexor pollicis longus muscle as causative factors [129, 130]. On the other hand, a far more proximally located nerve lesion still within the main median nerve trunk was sometimes observed [131]. Cases of nerve rotation unrelated to a special incident located within a sector of the median nerve trunk, even a few centimeters proximal from the medial epicondyle, were described [132]. The paper summarises 19 of these curious cases with hourglass-like constrictions. As we describe in Chap. 10, palsy with sudden onset (perhaps even with nerve rotation in images) generally has to be considered as an autoimmune inflammatory disease, probably in the context of Parsonage-Turner syndrome (syn. "brachial plexus neuritis"; see also Sects. 6.2.8 and 10.3; Chap. 12). Kiloh and Nevin already discussed this fact in their original paper, the first description of the anterior interosseus nerve syndrome [133]. Figures 6.24 and 8.19 show these rare examples as they present in modern imaging techniques. Further reports fit their primary idea exactly [134, 135]. Initial heavy pain must lead to suspect an inflammatory disorder. In a study, out of 21 cases, 12 presented with complete AIN lesion and 9 with incomplete AIN lesion. Pain was present in 15 cases out of the 21. Only five cases were related to compression and 16 cases to inflammatory lesion. The clinical course remained unknown in 10 cases. In the other 11 cases, spontaneous recovery occurred

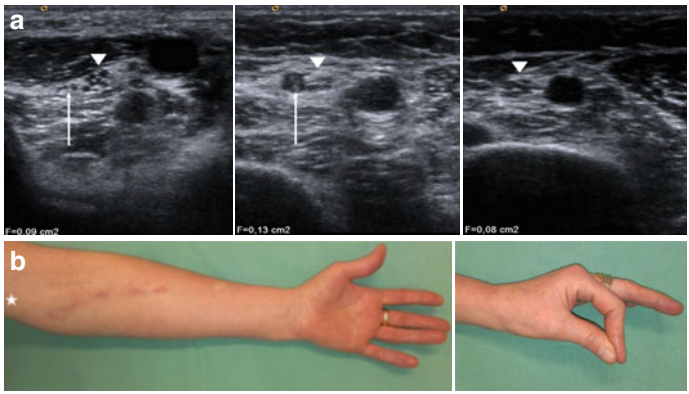


Fig. 8.19 Parsonage-Turner syndrome mimicking entrapment of interosseus anterior nerve. (**a**) On the *left side*, the fascicle group of the anterior interosseus nerve (*arrow*) just separates from the main trunk of the median nerve (*arrowhead*) below the pronator teres muscle at level of the elbow. At the distal third of the upper arm, this fascicle group (*arrow*) shows a hypoechoic swelling within the main trunk of the median nerve (*arrowhead*, *middle*). However, at the distal third of the upper arm, the median nerve (*arrowhead*) shows no abnormalities (*right*). (**b**) Neurosurgical decompression performed before ultrasound scan yielded no clinical improvement. *Asterisk* points on the location of fascicular swelling

10 times; and the mean recovery delay then was 14.3 months. The author concluded that, when no traumatism is obvious, no surgery should be considered for the first 12–16 months as late spontaneous recovery can still occur [136].

Four cases we have previously operated on ourselves remained without any really visible cause of palsy origin. Half of them improved postoperatively, the other half did not respond. One young woman operated on for a thoracic outlet syndrome with postoperative disappearance of her preoperatively long standing symptoms called our office 2 months later and complained of suddenly being unable to pinch the tips of her thumb and index finger against each other. When asking about pain, because plexus

surgery had been done 2 months earlier within the thoracic outlet, she described a 3-day period of extremely extensive pain in the shoulder girdle. After that, a sudden weakness occurred in a kind of a complete functional loss without any numbness and without any more pain. In the blood serum, antibodies against variola virus became evident. Another male patient entered our office and reported his history of a suddenly occurring extremely intensive pain in the elbow and forearm region with a duration of about 2–3 weeks. This pain period followed the primarily occurred incapability to flex his fingertips of the first and second digit. About 3 months later a neurosurgeon operated on him; 2 cm of the nerve were resected under the pronator teres arcade and repaired by a 2.5-cm graft. Microscopy of the nerve segment revealed oedema and lymphocytes within the perineural soft tissue.

To summarize the section, there are completely different causative factors of anterior interosseus syndrome – Kiloh-Nevin syndrome – to be kept in mind: beside rare compression neuropathies frequent autoimmune inflammatory diseases perhaps at the level of the brachial plexus or perhaps in the periphery with or without the curious occurrence of nerve rotations which, perhaps, will turn out as a special entity of the Parsonage-Turner syndrome (see Sects. 10.3; Chap. 12).

8.13.3 Clinical Symptoms

We have already described how a patient is suddenly unable to pinch the thumb and index fingertips against each other. Numbness is missing because the anterior interosseus nerve does not contain any sensory fiber with nociceptive function. For the same reason, usually we wouldn't expect severe pain in an entrapment although nobody can confirm that afferent muscle spindle fibers cannot generate a kind of pain sensation. Perhaps with improving imaging experience, we can better distinguish between three etiologic types of anterior interosseus

nerve palsies: an inflammatory disease with sudden onset and severe pain at the beginning, a nerve rotation with sudden onset and with or without pain (see Chap. 10), and a slowly progressing weakness which is mechanically caused and without pain.

8.13.4 Electrodiagnostics

Because the AIN is a pure motor nerve, SNAP of the median nerve is normal. The main electrodiagnostic studies include pronator quadratus (PQ) needle examination and AIN conduction study to PQ (recording with needle electrode). In controls, PQ latency was 4.1 ± 0.56 ms and the compound muscle action potential (CMAP) amplitude was 14.7 ± 4.3 mV. In patients, PQ latency was altered in 8 cases whereas CMAP amplitude, spontaneous activity, and recruitment pattern were abnormal in 18 cases. In the three remaining cases, diagnosis was assessed with needle examination of the flexor pollicis longus (FPL) and/or the flexor digitorum profundus of the second digit (FDP 2) [136]. Techniques using surface electrodes [137] are of poor value in true AIN cases since other median nerve innervated forearm muscles likely retain normal latencies and amplitudes and mask AIN lesions [136]. As anatomical variants for the EMG examination, the Martin-Gruber anastomosis should be considered (the AIN conveys fibers to the ulnar nerve, as described in Sect. 2.1), and in 30 % of cases the AIN can partially innervate the flexor digitorum superficialis muscles [129, 138]. Multifocal motor neuropathy (MMN) can mimic an AIN lesion. In these cases, electrodiagnostic evaluation demonstrated multiple conduction blocks, and, simultaneously, intravenous immunoglobulins were efficient in the treatment [139]. Rupture of the flexor pollicis longus and flexor indicis tendons can sometimes mimic an AIN lesion [140]. In the case of this important differential diagnostic constellation, the patient should present a normal conduction study and EMG examination.

8.13.5 Imaging

The sonographic examination of the anterior interosseous nerve is very difficult because this nerve is too small and located too deeply in the forearm to allow an adequate evaluation. If possible, one should look for a local swelling in comparison with the contralateral side [141]. We were confronted with a patient with sonography clearly bilaterally visualizing segmental swelling of a fascicle group within the median nerve at the upper arm just proximally to the elbow and not at the forearm in a case of Parsonage-Turner syndrome. The clinical and electrophysiological pattern was definitively a bilateral AIN syndrome. One unilateral example is illustrated in Fig. 8.19. The diagnosis of anterior interosseous neuropathy may also be demonstrated by muscle atrophies. MR-neurography (MRN) is a useful investigation in the diagnostic workup of AIN syndrome. It may demonstrate the abnormal AIN between the flexor digitorum superficialis and profundus muscles on the one hand, as well as denervation alterations in muscle substance of flexor digitorum profundus, flexor pollicis longus, and pronator quadratus (most common) [142–144] on the other. Axial T2 SPAIR shows high signal intensity (similar to vessels) of the AIN [145]. The radiologist should be aware that other patterns of muscle oedema in AIN injury (e.g. involvement of the flexor carpi radialis) may reflect variability in innervation [143]. Isolated increased signals in the pronator quadratus muscle are not necessarily related to an underlying nerve injury but are to proximal nerve inflammation [146]. In non-AIN compression, MRI often demonstrated muscle oedema patterns that were atypical for the AIN distribution. Differential diagnostic considerations then have to include a rupture of the flexor pollicis longus (FPL) tendon as mentioned above, a brachial plexus neuritis, the amyotrophic lateral sclerosis (see Sect. 10.2), and a possible compression of the proximal median nerve comparable to the two reported cases in the previous section [143].

8.13.6 Treatment

Provided a mechanical focal compression was considered, we formerly recommended a direct surgical approach via the middle third of the forearm. This approach became apparent as a failure in our eyes because causative factors must be expected within the pronator teres slit or proximally of it. Therefore, and in accordance with other authors, the approach eventually chosen will now be the same as that for the so-called "pronator-teres-syndrome". Of course, patients with initial pain in their anamnesis should not be selected as surgically suitable, because we must suppose a nonsurgical etiology as explained in the previous four sections. The remaining question will be how to diagnose a nerve rotation early. These curious cases need microsurgery, neurolysis, or even nerve repair; before imaging capacities, it was said that neurolysis has to include a fascicular inspection within the median nerve trunk to proximal in order not to overlook more proximal locations of AIN. We hope that ongoing experience in imaging will improve our chances to find the correct differential diagnosis at once. No other focal nerve lesion depends so much on imaging before the appropriate treatment decision is taken. However, if such surgery is really indicated we cannot wait 1 year or longer as recommended elsewhere [136].

8.14 Carpal Tunnel Syndrome

The median nerve entrapment at wrist level is well known because it is the most widespread spontaneous nerve compression syndrome that can occur in humans. Therefore, we do not try to repeat the huge mass of details of symptoms and treatment modalities. The list of literature one might have to refer to is enormous and could fill half this work. We try to restrict space given to important facts that have to be kept in mind.

8.14.1 Anatomy

In contrast to most of the focal entrapment neuropathies described so far, the carpal tunnel syndrome occurs within a narrow channel that exists from birth. Median nerve and additional tendons pass through this channel. Treatment consequences seem simple, but rare variations of nerve anatomy on one hand and variations of the thenar muscle supply on the other can provide reasons for unexpected difficulties.

1. One variation consists of a trans-ligamentous course of the thenar branch (Fig. 6.5a). We observed this variation ourselves a few times. After the publication of Lanz in 1977 we must always keep further variations in mind [147]. Moreover, and not to be neglected, is that, in addition to the variations discussed in this paper, we may be suddenly be confronted with a persistent median artery [148] (Fig. 8.20a, e).
2. A second problem may arise from different motor innervations of the human-thenar and hypo-thenar muscles [149]. Everybody knows that motor fibers supplying the thenar muscles can partly or exclusively run within the ulnar nerve as already described in Sect. 2.1.

The question of whether modern keyhole techniques which are applied on carpal tunnel release will be able to consider these anomalies safely remains controversial.

8.14.2 Damaging Factors

Most carpal tunnel syndromes are not related to any intraoperatively visible cause. In normal cases, with a spontaneous onset of symptoms, the cause is probably swelling due to chronic inflammatory tendovaginitis (Fig. 8.20d).

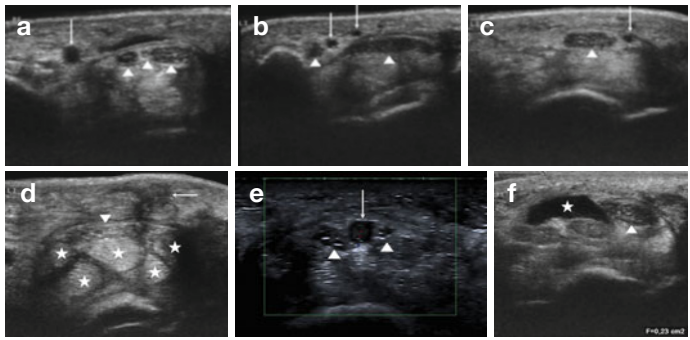


Fig. 8.20 HRUS, several anatomical variations of the median nerve and secondary pathologies resulting in carpal tunnel syndrome. (**a**) High division of median nerve with a smaller ulnar portion and a small persistent median artery between the two portions (*arrowheads*). Above the flexor retinaculum the hypoechoic belly of the palmaris brevis muscle appears. *Arrow* points on the ulnar artery. (**b**) A doubled ulnar artery (*arrows*) is located close to the median nerve (*right arrowhead*). *Left arrowhead* points on the ulnar nerve. (**c**) Palmar branch (*arrow*) of the median nerve (*arrowhead*) is additionally entrapped under the flexor retinaculum. (**d**) Tenosynovitis of the flexor tendons (*asterisks*) with space occupying effect within the carpal tunnel compressing the median nerve (*arrowhead*). *Arrow points* on a scar caused by previous carpal tunnel surgery. (**e**) High division of the median nerve (*arrowheads*) with a thrombosed persistent median artery (*arrow*). (**f**) Abnormal enlarged muscle belly of the second superficial finger flexor (*asterisk*) within the carpal tunnel compressing the median nerve (*arrowhead*)

On the other hand, special incidents or even injuries can easily become responsible for a carpal tunnel syndrome either with a sudden onset or with a strikingly quick progress: intraneural haemorrhage [150] (Fig. 6.5c), thrombosis within the persistent median artery [151] (Fig. 8.20e), tumor-like conditions such as synovial cysts, a chondroma or amyloidosis [152] (Fig. 6.5b), distally located lower arm fractures, a fact that has indeed been known for decades but is almost always disregarded by emergency surgeons [153]. Obese women are most likely to develop carpal tunnel syndromes and there is no reason to leave them behind with pain.

8.14.3 Clinical Symptoms

Symptoms are diffuse, may radiate up to the shoulder, and consist of pain, paresthesias, temporary or permanent numbness. The symptoms are particularly disturbing during the night. Strangely, they don't remain restricted to the three and a half radial digits as we would normally expect. The clinical differential diagnosis between a median nerve neuropathy at elbow level and wrist level depends on the examination: if the median nerve innervated forearm muscles are weak you can decide on a proximal nerve lesion. The thoracic outlet syndrome may come into consideration in rare cases, but please refer to the details in Sect. 8.2.1. Sometimes, a cervical root compression C6 has to be excluded, but its symptoms radiate from the spine into the periphery and the C6-dermatoma includes the extension side of the hand and the radial digits. Inflammatory diseases may rarely simulate a carpal tunnel syndrome such as, e.g. chronic inflammatory demyelinating polyneuropathy (CIDP). In combination with further existing focal entrapments, one has to take into account the hereditary tendency to multiple compression syndromes (see Chap. 10); all these syndromes remain rare, but must be kept in mind, at least when fruitless surgery has been carried out.

8.14.4 Electrodiagnostics

Diagnostic workup of median neuropathies in the carpal tunnel includes motor and sensory nerve conduction studies of the median nerve and the ulnar nerve in order to confirm the diagnosis and to exclude other kinds of neuropathies. A motor conduction study of the median nerve recording from the thenar muscle (distance to the stimulation site 6.5 cm) including measurement of distal latency (pathologic value >4.2 ms) shows poor sensitivity (65 %) but high specificity (98 %) [156].

Sensory conduction studies of the median nerve between the wrist and second or third digit to assess the sensory nerve action potential (SNAP) and the conduction velocity (SNCV) (N>47 m/s) also has a poor sensitivity of 65 % [156]. If results are normal, comparison of median and ulnar (N>44.6 m/s) sensory conduction between wrist and ring finger (difference >8 m/s is pathologic, sensitivity of 89 %, specificity of 98 %) can be done. Alternatively, comparison of median and ulnar sensory latency to the ring finger (sensitivity of 85 %, specificity of 97 %) does exist [156]. If these studies still do not allow an unambiguous diagnostic statement, the investigation of individual nerve segments, which is the most sensitive but rather time-consuming method, is recommended [157]. Overall, the sensitivity of standard tests (median digit-wrist sensory conduction velocity and wrist-thenar distal motor latency) was 83.5 %. Comparative/segmental tests disclosed abnormal findings in a further 11.4 % of cases, providing CTS electrodiagnosis in about 7 out of 10 "standard negative" cases. The overall sensitivity of the electrodiagnostic protocol was 94.9 % [157]. Median-to-ulnar motor conduction comparisons in the diagnosis of median neuropathy at the wrist, such as the median-thenar to ulnar-thenar latency difference (TTLD) and the median-thenar to ulnar-hypothenar latency difference showed high diagnostic sensitivities of 95–98 % and 85–88 %, respectively [158].

Comparison of median motor nerve distal latency (second lumbrical) to the ulnar motor nerve distal latency (second interosseus) revealed highly contradictory results with sensitivities of 56 % [147] to 97.5 % [159]. On the other hand, combining the standard tests yielded a sensitivity of 98.5 % [157]. CTS literature reviews provided convincing scientific evidence that median sensory and motor nerve conduction studies are valid and reproducible clinical laboratory studies, and they are said to confirm a clinical diagnosis of CTS with a high degree of sensitivity (>85 %) and specificity (>95 %) [156]. In accordance with the German guidelines for CTS, needle electromyography

of the abductor pollicis brevis muscle is only required in the case of technical problems such as innervation anomalies, thenar aplasia, advanced muscle atrophy, or pathologically increased stimulation thresholds [160]. In the case of incorrect diagnosis of CTS, different alternative origins of the complaints were later found such as polyneuropathy, ulnar nerve lesions, cervical radiculopathy, rheumatoid disorders, multiple sclerosis, syringomyelia or a motor neuron disease [161]. Nevertheless, for further details refer to Sects 5.3.1 and 5.3.2 (see Figs. 5.1 and 5.4).

8.14.5 Imaging

High resolution ultrasound can locate a lesion of the median nerve by examining the entire nerve course in the carpal tunnel, and even distally it can demonstrate the digital nerves very exactly. In addition to qualitative criteria of a compression neuropathy, several measurement methods are used such as various cut-off values of the nerve cross section proximal to the compression site, or the wrist-to-forearm ratio, which is considered to be abnormal over 1.4 [162] (Fig. 6.4). Evidence-based guidelines after a systematic review were meanwhile developed from cross-sectional area measurements in CTS. Measurement of median nerve cross-sectional area at the wrist is accurate and may be offered as a diagnostic test for CTS (Level A) [163]. Four articles met class I level of evidence [164–167]. The CSA "cut-off" ≥ 8.5 mm^2 leads to a sensitivity of 97 % and a specificity of 98 % [166] and a "cut-off" ≥ 10 mm^2 leads to a sensitivity of 82 % and a specificity of 87 % [167]. In a meta-analysis of 28 studies with 3995 wrists, a "cut-off" ≥ 9 mm^2 resulted in a sensitivity of 87 % and a specificity of 83 % [168]. A common anatomical variation is the bifid median nerve (Fig. 8.20a, e). The area of each portion of the nerve was added together, and a single cut-off of 12 mm^2 was used. The accuracy improved

when the difference between the median nerve area measured at the wrist and proximal forearm was calculated, whereby a difference of 4 mm or greater was used to diagnose CTS [169]. There are additional features of CTS such as palmar bowing of the retinaculum, an indentation of the median nerve at the proximal edge of the retinaculum, and the flattening ratio among others, which may be noted with sonography, but their value for the diagnosis of CTS has not been sufficiently studied [170]. Impaired vascularization of the median nerve in CTS was demonstrated and quantified by colour Doppler [171, 172]. Even if this method seemed to have promise for the future, currently the lack of standardization, comparative studies to the above-mentioned routine methods, availability of high resolution ultrasound devices, and the extra time required are arguments against current use as a routine method. Sonographic measurement parameters ("cut-off" CSA and wrist-to-forearm ratio) cannot determine severity because they do not show enough significant correlation with clinical severity in CTS or with electrodiagnostic severity [173]. Sonography can help to differentiate the so-called idiopathic CTS from a variety of local conditions with external nerve compression (e.g. ganglia, tumors, tenosynovitis, accessory muscle tissue and vascular anomalies [170]. It can provide preoperatively relevant additional information by detection of anatomic variants, e.g. proximal division of the median nerve, persistent median artery, atypical course of the thenar branch, and ulnar artery very close to the median nerve (Figs. 8.20 and 6.5) [174]. Some of them cause a potentially increased risk of iatrogenic nerve injury, especially in endoscopic procedures [175, 176]. Finally, it can help to explain poor outcome postoperatively such as incomplete transection of the retinaculum flexorum (Fig. 8.21) [177], postoperative nerve lesions with neuroma formation (Fig. 8.22), persistent compression due to scar tissue, thickening of the epineurium or epineural fibrosis, hematoma, or infection [178, 179]. Therefore, sonography should be performed in every patient at risk [170].

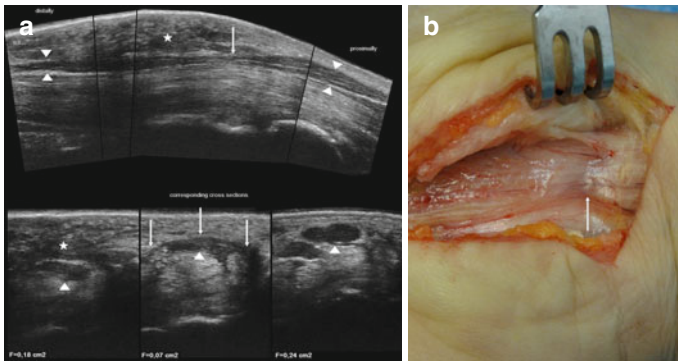


Fig. 8.21 Failed carpal tunnel surgery in HRUS. (**a**) Longitudinal section and corresponding cross section of the median nerve (*arrowheads*) within the carpal tunnel. Median nerve is compressed by an incompletely released flexor retinaculum (*arrows*) and scarring (*asterisk*) at the proximal part of the tunnel. Distally the released nerve is covered by scar tissue (*asterisk*). Note also the focal hypoechoic nerve swelling and CSA enlargement proximally to entrapment. (**b**) The image demonstrates the corresponding intraoperative findings (*arrow*: site of compression)

MRI is quite suitable in the diagnosis of carpal tunnel syndrome [180] as we try to show in Figs. 6.15 and 6.16. However, in our experience, sonography solves the vast majority of questions and is less expensive and less time-consuming.

8.14.6 Treatment

As the narrow space within the carpal tunnel can hardly be easily enlarged, surgery is usually the correct solution. Nevertheless, ultrasound guided corticosteroid infiltrations can achieve short periods of improvement in special situations, in particular, percutaneous needle release (“splinting”) as comparably applied

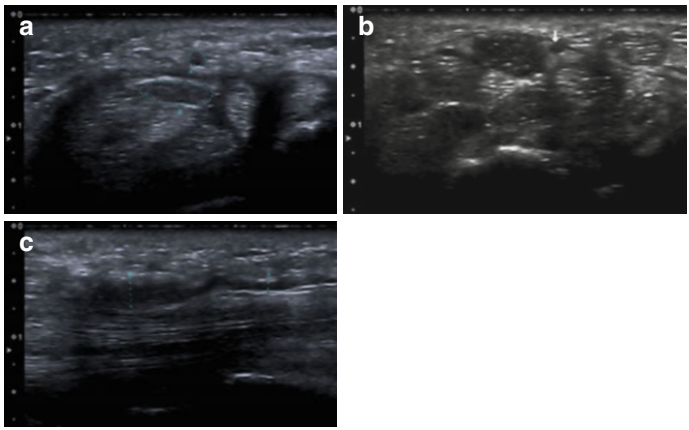


Fig. 8.22 HRUS, major lesion (Sunderland V) of the palmar branch of the median nerve after carpal tunnel surgery. (**a**) Distally a swelling of palmar branch (*A*) and median nerve (*B*) appears. (**b**) Proximally both the median nerve and its palmar branch (*arrow*) are covered by the fascia antebrachii. (**c**) Longitudinal section revealing the stump-neuroma of the palmar branch

on Dupuytren contracture cases should not be neglected. Idiopathic cases of only the demyelinising form with DML <6.2 are definitively suitable as the two neurologists among the authors point out according to their experience.

The goal of surgery is transection of the transverse carpal ligament. As a disadvantage, the flexor tendons lose their hypomochleon. After some weeks following surgery patients often complain of reduced force when flexing their fingers [154].

We prefer the open operation to overcome the above-mentioned anomalies early on. Nevertheless, many surgeons have become familiar with the one-portal and two-portal endoscopic techniques. Meanwhile, studies and reports exist supporting the already 12-year-old opinion that there are no significant advantages or disadvantages between open and endoscopic surgical procedures [154]. It is interesting that surgeons who use

endoscopy almost always say that the learning curve is much longer than that of the open operation. Therefore, open surgery is mandatory to be learned first, and, for this reason, the open technique should never be abandoned.

Considering the original purpose of this book we do not intend to address experts but physicians who are starting their training. We try to give a short description of our own method: use short general anaesthesia or axillary plexus anaesthesia, avoid local anaesthesia because anatomy then gets more difficult, use incisions of not more than 2 cm between thenar and hypothenar, start at the level of the distal transverse wrist crease, use a small self-holding retractor to hold back the subcutaneous fat tissue, let the longitudinal fibers of the palmar fascia come into view, incise longitudinally, use the retractor to keep away its incised margins, let either fat tissue or transverse located muscle fibers come into view, remain vertical and transect these fibers (vertical is necessary in order not to enter the Guyon's canal), replace your retractor more deeply, let the transverse silvery ligament fibers come into view, use the knife to transect layer of layer of these fibers parallel to the expected median nerve fascicles and transverse to the ligament fibers (doing so use very slight pressure so as not to enter the tunnel compartment forcibly – with enough retractor force, the incised ligament margins move apart), now use closed scissor tips to probe the tunnel compartment, lift up the distal end of the incised skin in the palm, use your scissors and transect piece by piece parts of the carpal ligament after bluntly probing the same distance within the carpal tunnel, retract the closed scissors tips with slight force upwards so that you can feel remaining ligament parts (they still need to be transected to complete your goal). By doing this final procedure piece by piece, you will avoid injuring the arterial palm arch. Now change direction and start the same procedure to proximal, and proceed with the transection until you palpate your scissors' tips 1.5 cm above the distal transverse wrist crease. Remain a little bit ulnar absolutely on the ulnar side to

avoid damage to the sensory palmar nerve branch. Then, finally, insert a small drain without suction bottle and close the skin only. Some bleeding can flow out into the bandage; the paraneural tissue responds on a hematoma with scarring. The drain is removed the next day without any pain.

Criticism can arise when the open method is said to be more secure to avoid injuring the above-mentioned anatomical anomalies. In any case, the trans-ligamentous course of the motor thenar branch comes into view early. Hand surgeons extend their incisions with regard to these anomalies. On the other hand, we endorse endoscopy. Incision sizes required thus remain controversial. We limit ourselves to our own experiences which have been without problems since 1999. There were four of our own cases with incomplete ligament transection which were re-operated after persistence of symptoms and unimproved electrodiagnostic testing. One additional case, a recurrence, became evident by an epineurium fibrosis, perhaps resulting from slight inflammation from a concomitant tenosynovitis

Another question remains – how to deal with nerves affected by scarring progressing after trauma – fractures in the neighbourhood of the carpal tunnel or, for instance, perilunate luxation into the carpal tunnel. Internal neurolysis as an adjunct to the carpal tunnel release was advised in 1973 in these cases [155], although never in the case of normal entrapments. Meanwhile, 6 years ago, guidelines in our region heavily opposed this adjunct. The method of choice probably lies somewhere between these different positions. When we read Curtis and Eversmann's paper carefully, arguments against gentle neurolysis in selected cases should vanish: "Throughout the procedure, the posterior or dorsal aspect of the median nerve remains undisturbed, and the median nerve is not elevated from its bed and areola tissue within the carpal tunnel. This precaution is taken to preserve whatever microcirculation may be present on the posterior surface of the nerve proper". Everybody can read that the authors do not recommend an interfascicular

neurolysis. Surgery has – as always – to remain adapted to the individual nerve situation.

8.15 Ulnar Nerve at the Elbow

Focal irritations of the ulnar nerve at elbow level represent the second most common focal nerve entrapment after carpal tunnel syndrome. Whereas clinical assessment and surgical relief of the latter syndromes are well defined procedures, treatment and surgery of the ulnar neuropathy is still debated and, at the same time, surgical problems occur more frequently and can also be the reason for unexpected litigation. The clinical and surgical approach to patients with ulnar neuropathy is consequently characterized by more restraint.

8.15.1 Anatomy

The ulnar nerve is unfavourably located within a bony groove (retrocondylar groove) where it is covered by an aponeurosis with transverse fibers (cubital tunnel retinaculum, Osborne's ligament). Before entering the retrocondylar groove, and about 4–5 cm above it, the Struther's arcade may cover and irritate the nerve course [181]. A few centimetres more distally but still above the medial epicondyle, a bony process is said to exist very rarely with a ligament which connects the tip of the bony spur to the medial epicondyle (Struther's ligament). Whereas the arcade exists quite frequently, the supracondylar process with the ligament is increasingly unlikely to exist [122]. When exiting the retrocondylar groove the ulnar nerve enters a compartment between superficially located flexor muscles including the pronator teres and a deeper situated muscle layer (flexor digitorum profundus muscles). Between

both muscle layers, an aponeurosis also covers the course of the ulnar nerve. This aponeurosis is of importance because most focal ulnar neuropathies originate from the nerve's entrance site underneath the aponeurosis (Fig. 8.24a).

8.15.2 Damaging Factors

In contrast to many other entrapments, ulnar neuropathy literature is full of opinions about different factors which cause the nerve lesion: E.g. interrelation between fractures of the elbow or joint deformities due to severe rheumatoid arthritis and a delayed neuropathy on one hand (Fig. 8.24c); single or multiple episodes of slide traumas that were not severe enough to cause bony alterations on the other hand; moreover, multiple episodes of minor pressure associated with elbow flexion such as, when leaning the elbow on hard desks. Another possibility is the presence of space occupying soft tissue masses within or around the retrocondylar groove such as ganglia or lipomas, or an anomalous muscle within the groove (ancoaneus epitrochlearis muscle) (Figs. 8.25a, b and 8.26c, d, f), as an extreme rarity, with the above-mentioned supracondylar process with its ligament – in up to or around 1 % of cases. And finally, the nerve can be liable to snap around the epicondyle with the risk of suffering from more extended compression episodes with the elbow in a flexed position [182].

However, in the meantime, damaging factors became more of a consideration: it is said to be sufficient to distinguish between primary – or idiopathic – and secondary – or symptomatic – forms of ulnar neuropathies only. Unfortunately, this simple classification influenced discussion about the correct surgical procedure in one single direction as we will describe later. However, we tend to express our hope that, in future, high resolution ultrasound imaging will help to differentiate more effectively between possible causative factors early enough. We also hope that the resulting types of surgery can be then adapted

to our imaging findings, and that the amount of litigation will thus be reduced.

We must never forget that the retrocondylar groove is a location which permanently exposes the nerve to unfavourable influences: flexion and extension movements of the elbow joint on one hand and repeated episodes of pressure on the other. No other nerve location is as dependant on functioning tissue to provide passive motility: motility of the main nerve trunk within its surroundings and motility of fascicle groups against each other within the epineurial sheaths [41]. Any damage to paraneurial or perineurial tissues and their fibrosis will deteriorate gliding abilities of longitudinally orientated fibers against each other. It seems conceivable that small nerve axons then suffer from tissue shrinkage if the adjacent joint is extended and flexed. We are convinced that intraneural damage plays a role that is underestimated in ulnar neuropathies; and, meanwhile, we know that high resolution ultrasound can depict these kinds of tissue reaction preoperatively, and in problem cases postoperatively as well. These imaging results are verified by previous statements.

8.15.3 Clinical Symptoms

Symptoms depend on the degree of nerve lesion, of course. They mostly begin with paresthesias and numbness over the hypothenar and over the flexion side of the one and a half ulnar fingers. The area of numbness includes the small area of the skin branch which arises from the ulnar nerve a few centimetres proximal to the wrist. It supplies the dorso-ulnar aspect of wrist and hand. This area allows to differentiate clinically between ulnar neuropathies at elbow or wrist level: if the dorso-ulnar numbness is absent we have at least to suppose a distally located nerve entrapment (see Sect. 8.16).

A pathognomic Tinel sign can mostly be elicited when we palpate the ulnar nerve within the retrocondyle groove. It is extremely rare that it is missing. Its frequent occurrence indicates that intraneural alterations probably differ in the case of carpal tunnel syndrome, because there a Tinel sign is extremely rare. We should explain this difference with our arguments about early alterations of gliding tissues adjacent to joints undergoing extensive motion, and also with the presumption that axonal de- and regeneration occurs earlier at this location than within pre-formed narrow channels.

Probably for the same reason, patients' complaints consisted much more frequently of decreasing myokinetics of the intrinsic hand muscles. Several daily activities were often reported to be impaired such as household activities, writing, or eating. In contrast to that, pain was rare; pain is almost never in the foreground of patients' discomfort. When however pain is a complaint, it is described as always occurring in combination with long-time elbow flexion.

8.15.4 Electrodiagnostics

Diagnostic workup of the ulnar neuropathy at the elbow should include motor and sensory nerve conduction studies of the ulnar nerve and the median nerve in order to confirm the diagnosis and to exclude other kinds of neuropathies. A fractionated motor nerve conduction study with three different stimulation sites (wrist, below ulnar groove, above ulnar groove) and a distance of at least 10 cm is recommended [183]. The recording electrode is placed on the abductor digiti minimi muscle. Recording over the first interosseus dorsalis muscle together with the application of an additional stimulation point at the elbow level may improve the sensitivity of this technique up to 98 % [184]. In the case of a demyelinizing lesion, fractionated motor nerve conduction studies can localize the lesion at elbow level if the following conditions are met [185]:

- "Focal slowing of the motor conduction velocity over 16 m/s at the elbow-segment in comparison to the motor nerve conduction velocity at the forearm-segment and/or
- Decay of the proximal CAMP by more than 20% – and/or abnormal temporal dispersion –, whereas the distal CAMP at the forearm remains normal" [185]

According to other authors, even a focal motor nerve conduction velocity slowing >10 m/s at the elbow-segment is considered to be pathological [183]. However, German guidelines recommend the above-mentioned higher value to avoid false positive results, since fractionated motor nerve conduction studies may be error prone and are operator dependent [186]. In a more recent study, the 45° of elbow flexion was found to be the position of least variation in motor and sensory nerve conduction velocities between the across elbow and below elbow segments [187]. Sensory nerve conduction studies may help to differentiate infra-ganglionic lesions (e.g. ulnar neuropathy at the elbow or lower brachial plexus lesion) from supra-ganglionic ones of root C8 (see Sect. 5.3). However, the sensory nerve action potential obtained after antidromic recording and stimulation at the wrist in the early stages of ulnar neuropathy can still remain normal despite clinical symptoms, and therefore it is less specific [184]. Here, comparison with the unaffected side may sometimes help to detect subtle changes of SNAP and of sensory nerve conduction velocities. In the late stages of the disease, massive secondary axonal loss often follows the focal demyelination. As a consequence, the site of the nerve damage can no longer be determined by means of fractionated motor nerve conduction studies due to generalized reduction of the CMAP amplitude and a slowing of motor nerve conduction velocity at all stimulation points. In this case, needle electromyography is able to demonstrate an axonal lesion, especially of the flexor carpi ulnaris muscle, as a sign of proximal affection of the ulnar nerve [188]. However, high resolution ultrasound here provides a more accurate alternative which is superior to

EDX as we will describe in Sect. 8.16. In order to differentiate other conditions (lower brachial plexus lesion, C8-root lesion) from an ulnar neuropathy at the elbow, sensory nerve conduction studies of the medial cutaneous antebrachii nerve and needle electromyography of several muscles supplied by root C8 – but not by the ulnar nerve – may be helpful [49]. For supplementary details please refer to Sects. 5.3 and 8.2.1.

8.15.5 Imaging

As mentioned above, high resolution ultrasound provides a more accurate way to locate a lesion of the ulnar nerve by examining the entire nerve course from the axillary region down to the wrist. In contrast to that, EDX cannot differentiate between ulnar neuropathies at the elbow and more proximally located pathologies. A case after failed surgery on ulnar neuropathy was recently examined in HRUS postoperatively. It revealed a Parsonage-Turner syndrome with the involvement of the whole proximal ulnar nerve trunk (see also Sects. 6.2.8 and 10.3). If the anamnesis survey had been conducted correctly, this surgery could have been avoided. Therefore HRUS or MR-neurography is the first choice if a high lesion comes into consideration (Fig. 8.23).

HRUS signs of ulnar neuropathy at the elbow are as follows:

1. Cross sectional area proximal to the compression site at the elbow $\geq 0.10\ cm^2$
2. Humeral-cubital ratio of both cross sectional areas >1.4
3. Qualitative signs of a nerve entrapment as described in Chap. 6 [189, 190]

In addition to nerve conduction studies, HRUS increases the sensitivity and specificity of the diagnostic workup [191]. Moreover, HRUS is able to demonstrate multiple or atypical sites of entrapment and other damaging factors like luxation of the ulnar nerve around the medial epicondyle, an additional

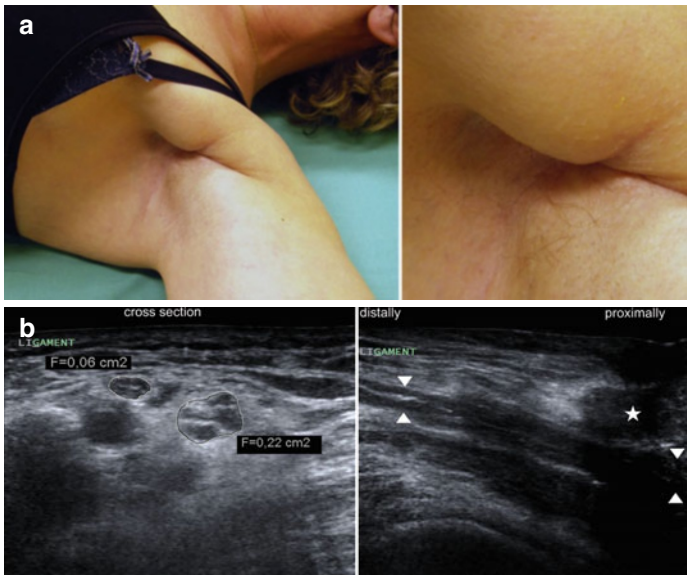


Fig. 8.23 Proximal lesion of left ulnar nerve at the axilla in HRUS. (**a**) Patient with history of breast-cancer developed a painful sensorimotor palsy of the left ulnar nerve. Note the scarring at the axilla that makes it impossible to fully elevate the left arm. (**b**) Note the pathologically changed CSA of the ulnar nerve (0.22 cm^2) in comparison with the median nerve (0.06 cm^2) on the *left image*. The *right image* shows a longitudinal section with a metastasis (*asterisk*) compressing the ulnar nerve (*arrowheads*)

ancoaneus epitrochlearis muscle, or a space occupying lesion (e.g. ganglion cyst, exostosis, peripheral nerve sheath tumor), factors that may influence the therapeutic strategy [192]. Furthermore, years after elbow fracture, the ulnar grove can undergo degenerative changes of the bony substance, leading to a nerve compression over a long distance and a more ventral nerve course. This condition – a "late ulnar palsy" – can be easily diagnosed with HRUS [193]. On the other hand, HRUS is

also valid to detect generalized neuropathies affecting nerve segments outside locations where entrapments used to occur (e.g. multifocal chronic inflammatory demyelinating neuropathy – CIDP, Sect. 10.5.1). They are characterized by a segmental nerve swelling or by the swelling of the entire nerve with fluctuations of fascicle thickness, "fascicular remodelling" or "masking" of the fascicular echotexture [194–196]. Finally, according to our experience of failed surgery, HRUS is able to detect the possible cause of the problems, e.g. epineural fibrosis, distal kinking after transposition, postoperative "snapping ulnaris syndrome", persistent compression due to scar tissue. Several of the factors described above are shown in Figs. 8.24, 8.25 and 8.26.

MRI is equally suitable in the diagnosis of ulnar neuropathies at the elbow. Especially in the early stages of the disease, it shows a T2 lesion of the fascicles [197]. On the other hand, it allows to administer contrast agents if a tumor is suspected. However, in the majority of cases this expansive examination is not required.

8.15.6 Treatment

As already mentioned, treatment modalities will and should be debated in future. Our personal opinion is that, because of the new and easily available imaging techniques, the appropriate surgery will again be subject to discussion. Basically, there exist three types of surgery:

1. In situ decompression, including endoscopic decompression [185, 198]
2. Decompression with subcutaneous transposition
3. Decompression with sub-muscular transposition [199, 200]

Assmus and co-workers have provided a huge overview about 137 papers dealing with ulnar neuropathy [185]. Based

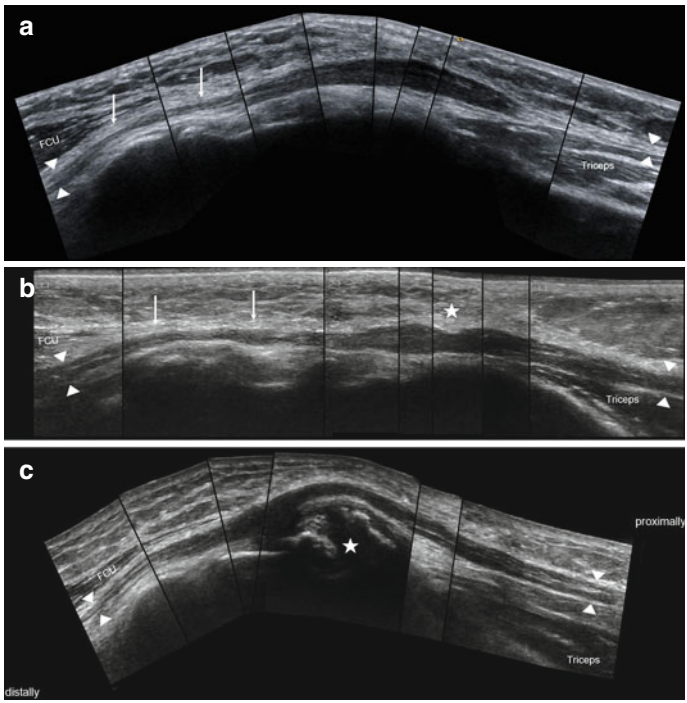


Fig. 8.24 Spectrum of ulnar neuropathies at the elbow in HRUS (I), longitudinal panoramic image reconstructions. *FCU* Flexor carpi ulnaris muscle. Triceps: Medial head of the triceps muscle. Arrowheads: Ulnar nerve. (**a**) Most typical form with compression below the flexor aponeurosis (*arrows*) and proximal swelling at the retrocondylar groove. (**b**) Multiple compression sites (flexor aponeurosis = *arrows* and retrocondylar groove = *asterisk*). (**c**) "Late palsy of the ulnar nerve" after a history of supracondylar humerus fracture with degenerative changes of the retrocondylar groove (*asterisk*)

on the evaluation of all the literature mentioned and on their own experience, the following recommendations for surgery were outlined:

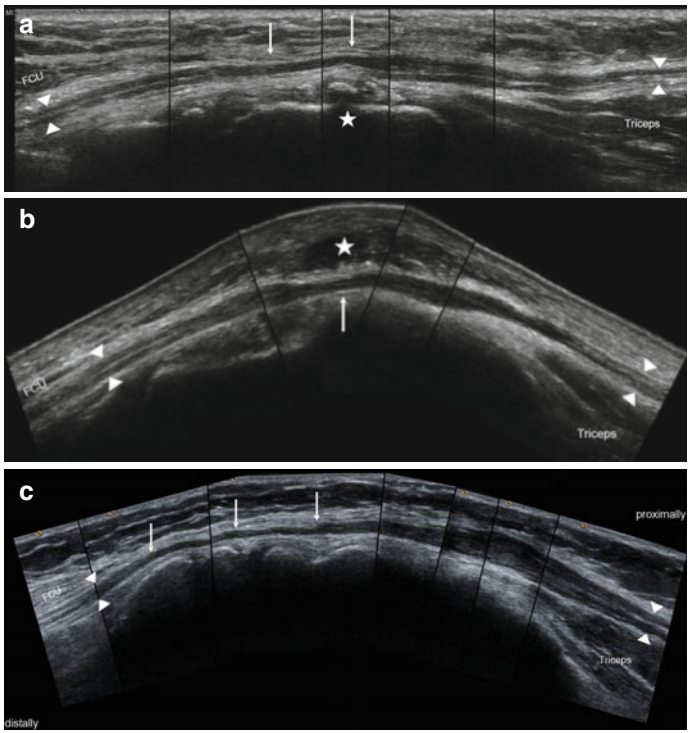


Fig. 8.25 Spectrum of ulnar neuropathy at the elbow in HRUS (II), longitudinal panoramic image reconstructions. FCU: Flexor carpi ulnaris muscle. Triceps: Medial head of the triceps muscle. Arrowheads: Ulnar nerve. (**a**) Ulnar nerve is compressed by flexor aponeurosis above (*arrow*) and osteophytes below (*asterisk*). (**b**) Ulnar nerve is compressed by an accessory muscle belly (*asterisk*, ancoaneus epitrochlearis muscle) in the retrocondylar groove (*arrow*). (**c**) Failed surgery after ulnar neuropathy at the elbow. Image shows a persistent long-segmental compression (*arrows*) of the ulnar nerve

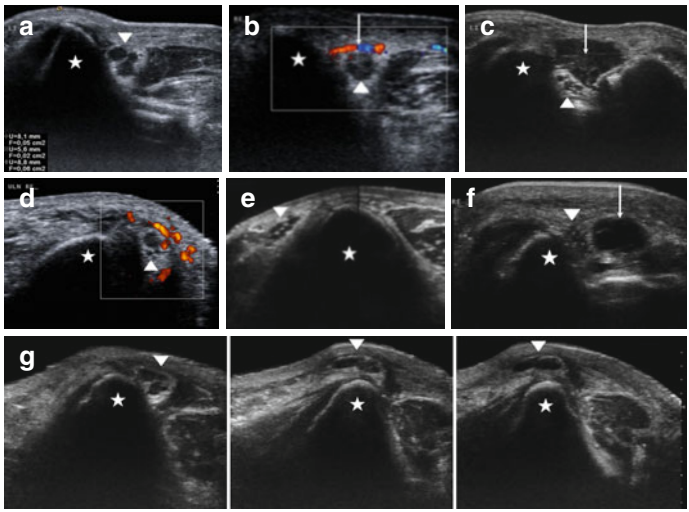


Fig. 8.26 Spectrum of ulnar neuropathy at the elbow in HRUS (III), cross sections at the retrocondylar groove. *Asterisk*: medial epicondyle. *Arrowhead*: ulnar nerve. (**a**) Anatomical variation with division in three major parts. (**b**) Anatomical variation: An atypical artery (*arrow*) crosses the ulnar nerve. (**c**) Accessory ancoaneus epitrochlearis muscle (*arrow*) compresses the ulnar nerve. (**d**) Angioma. (**e**) Appearance after subcutaneous transposition. (**f**) Ganglion-cyst (*arrow*) compresses the ulnar nerve. (**g**) Image sequence shows snapping of the ulnar nerve after surgery without transposition

- Simple in situ decompression is the treatment of choice
- The transposition of the ulnar nerve remains reserved to cases post-trauma or adjacent to joint deformities, with ulnar nerves embedded in scar tissue, and cases with nerve luxation, combined with pain

Unfortunately, the authors do not delineate exactly which clinical sign should be looked for, which degree of nerve lesion has should be assessed for making the decision, and which

available technical tool should be used (X-ray, HRUS, MRI, electrodiagnostic testing) to choose the appropriate procedure from the different surgical procedures available. They mention that nerve transposition can still be a secondary consideration; we will see below that secondary methods worsen the prognosis. In practice, two additional factors will impede a differential decision making:

- Surgery of ulnar neuropathies is usually done as an outpatient procedure and, because of this
- A large-scale, expensive and technical diagnostic is therefore commonly avoided

As result, for 13 years we were rather frequently, even from abroad, confronted with patients who had undergone simple in situ decompression, either using an open technique or endoscopically. Their ulnar nerve symptoms had slowly improved afterwards; their nerve conduction velocity had also improved a few months later, but radiating pain of extremely disturbing amount had been experienced. Either severe pain during elbow flexion is complained about or electric-current-like pain every time the elbow touches the body during arm movements. This type of pain is comparable to the characteristic pre-operative Tinel sign but is of more weight. How can we explain this phenomenon? Nerve re-explorations were always characterized by scar tissue around the epineurial sheath. This held the nerve trunk fixed within the retrocondyle groove. The gliding ability of surrounding soft tissue within the bony groove was completely lost. The question arose every time as to how surgery should have been carried out to avoid such findings; was it inadequate surgery, or have the special causative factors mentioned above been disregarded?

Surgical re-explorations of these cases were rather difficult and, of course, of worse prognosis and of more risk if compared with primary surgery. All these cases needed sufficient nerve

transposition, first, in order to get relief from adhesion in the bony groove, second, to break free from unfavourable traction forces during elbow flexion, and third, to set up chances to re-establish efficient intraneural blood supply from a soft tissue recipient bed.

By 1945, Sunderland had already outlined his opinion that the transposition of the ulnar nerve anterior to the humeral epicondyle does not impair its function and blood supply [201]; when we read this paper in detail it is easy to imagine how reduced intraneural blood supply remains within a scar tissue-embedded nerve in a bony groove. Because of these considerations and because of the reduced prognosis of secondary transpositions, we prefer subcutaneous nerve transposition as the primary procedure as long as hard pre-operative criteria to differentiate between small or extended surgery are not easily accessible. When out-patient surgery is intended, pre-operative examination methods need to be cheap, quick, easily available, and reliable. Until now, we have assumed ourselves to be in accordance with the experience of Dellon and Kline, who always advised transposition but, in contrast to us, sub-muscular in all cases [199, 200].

Subcutaneous transposition, which we go on to estimate as sufficient, needs three pre-requisites to be fulfilled [202]:

1. Proximally to the retrocondyle groove, the medial intermuscular septum always should be incised far enough
2. At the level of the bony groove, re-sliding of the nerve in the direction of the epicondyle should be prevented
3. Distally to the epicondyle, nerve kinking during elbow flexion should be prevented

We start the incision about three fingertips medial to the epicondyle in an inverse U-shaped manner and extend the incision about 3 cm over the upper arm and 4 cm over the lower arm, S shaped. The subcutaneous tissue is divided until the muscle fascia over the elbow joint comes into view. The disadvantage of this incision remains that branches of the medial ante-brachial

cutaneous nerve are frequently transected. The edge of the medial intermuscular septum at the medial upper arm can easily be palpated, and the ulnar nerve is exposed just under the septum. We follow the nerve from proximal to distal. Special care should be taken where the deep aponeurosis between the two muscle layers of the lower arm flexors starts. As described above, this point is the main causative factor of compression. The muscle fibers that lie above the aponeurosis have to be split over 3 cm, and then the aponeurosis should be incised longitudinally and parallel to the nerve course. No motor branch which arises from the ulnar nerve trunk needs to be sacrificed as formerly recommended in books. With your fingertip to proximal you can exclude the extremely rare Struthers ligament. The sharp edge of the medial intermuscular septum has to be exposed, and it has to be incised far enough that the transposed nerve does not touch parts of it. Opening the epineurial sheath a little, you are able to mobilize the first motor branches from the main trunk in a proximal direction. It allows the best nerve transposition and avoids nerve kinking when flexion re-starts. Some cases need the fascia to be incised over the flexor bellies, roughly parallel to the transposed nerve. You will then find a hard intermuscular septum between the muscle fibers. This septum will be incised only whereas the muscle fibers next to it should remain untouched. This surgical step in particular prevents the nerve from kinking distally to the elbow. Finish now with resorbable stitches which grasp subcutaneous tissue and ventral muscle fascia. Four or five of these stitches that you have to close manually are sufficient to hold the nerve, which remains able to slide behind them, preventing it from re-luxation. The wound will be closed with a suction drain, plaster immobilisation is unnecessary, and the skin stitches are removed 10–14 days later (Fig. 8.27).

During our experience over the last 15 years we have been happy to exclude recurrences and, particularly, the unfavourable postoperative pain cases that we have described above.

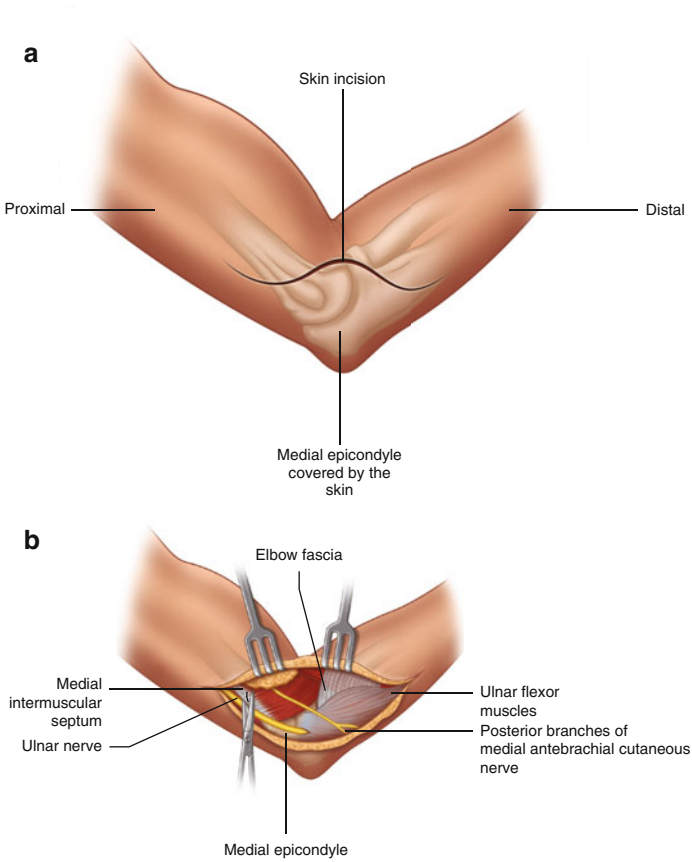


Fig. 8.27 Subcutaneous transposition of the ulnar nerve. (**a**) Skin incision. (**b**) Exposure of the ulnar nerve and the medial intermuscular septum. (**c**) Transection of the septum and splitting of ulnar flexor muscles. (**d**) Subcutaneous transposition; the nerve is fixed behind several stitches placed between the subcutaneous tissue and the ventral elbow fascia to prevent re-luxation

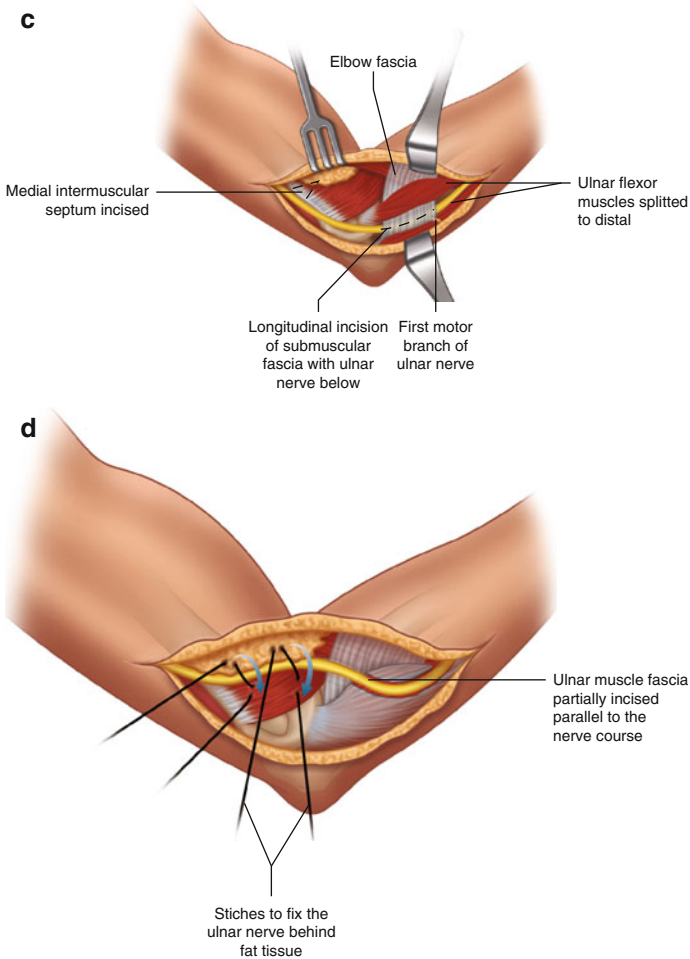


Fig. 8.27 (continued)

Three cases of rebleeding had to be explored on the same day as the primary exposure, and two of them turned out to be due to the lack of a coagulation factor.

8.16 GUYON's Canal Syndrome

Guyon's canal syndrome is an unsatisfactory term. Within the canal, the ulnar nerve trunk divides into several terminal branches, a terminal motor branch that innervates the hypothenar muscles, a superficial sensory branch that supplies the ventral area of the one and a half ulnar digits, and the important deep motor branch which innervates the intrinsic hand muscles except for the hypo-thenar, but including the first dorsal interosseus muscle. Due to this anatomy, several patients with Guyon's syndrome can present isolated impairments of only one of these branches; whereby most disturbing is the involvement of the deep palmar branch which supplies the intrinsic muscles.

8.16.1 Anatomy

Anatomical importance involves two facts: first, be aware of the above-mentioned ramifications and, second, be aware of the concomitant ulnar artery with its small branches. It is mandatory to avoid any harm to this artery. Guyon's canal is shaped by fibers of the ulnar ligament as roof, and at the lateral side by the pisiform bone. At the end of the canal, the deep motor branch to the intrinsic muscles arises from the main trunk at its lateral aspect, turns to the lateral side and then to the medial side where it courses around the hamate bone: There are several illustrations that don't respect this anatomical feature. The entrance point into the deep palm compartment is covered by a small tendon-like arch which serves as a muscle fiber origin. Exposure of Guyon's canal therefore needs to progress to this point in

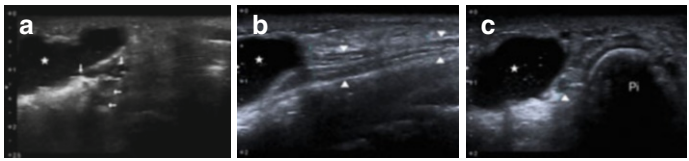


Fig. 8.28 Guyon's canal syndrome caused by a ganglion-cyst (*asterisk*) at the level of the pisiform bone (Pi) in HRUS. (**a**) *Arrows* show the connection of the cyst with a joint. (**b**) Ulnar nerve (*arrowheads*) is compressed by the cyst, longitudinal section. (**c**) Corresponding cross section

order to always exclude additional or isolated deep branch compression at this site.

8.16.2 Damaging Factors

Guyon's canal syndrome is rare when compared with ulnar neuropathies at the elbow. Nevertheless, we had to realize two or three cases per year. Spontaneous occurrence is uncommon whereas cases with repeated external pressure in the patient's history exerted over months are the best known cases. This fact seems in accordance with other observations in the literature [203]. Anomalous muscles within the Guyon's canal were described [203, 204], a giant cell tumor [205]; small ganglia are well known (Figs. 6.9 and 6.19); we ourselves accidentally found a small neurinoma (Fig. 8.28 and 8.29) originating from one nerve fascicle. Symptoms commonly start related to the nature of the patient's work, which mostly causes episodes of repeated pressure on the ulnar aspect of the hand.

8.16.3 Clinical Symptoms

Pain is uncommon, numbness if existent involves the one and a half ulnar digits at their flexion side, and sometimes it includes the skin

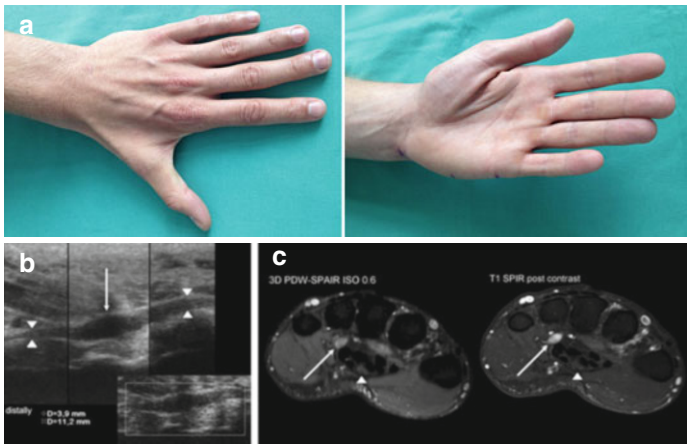


Fig. 8.29 Guyon's canal syndrome with deep palmar branch only affected caused by a small peripheral nerve sheath tumor. (**a**) Clinical presentation. The first interosseus dorsalis muscle shows severe signs of atrophy whereas the hypothenar-muscles are preserved. (**b**) HRUS, longitudinal ultrasound image revealed a non-vascularised, fusiform-swelling (*arrow*) of the deep palmar branch (*arrowheads*). (**c**) Corresponding 3 T MRN images with increased signal intensity on PDW-SPAIR images and a strong contrast uptake on T1 SPIR images (*arrows*) (MRN images courtesy of Karsten Stock, MD/Dessau-Rosslau Hospital)

over the hypo-thenar. The skin supplied by the dorsal branch which arises from the nerve trunk 4 cm proximal to wrist level is always excluded. Muscles of the hypo-thenar can demonstrate a visible atrophy, and the same holds true with the adductor pollicis muscle at the dorsal aspect of the hand between the first and second digit.

8.16.4 Electrodiagnostics

Electrodiagnostic workup in Guyon's channel syndrome includes fractionated motor and sensory nerve conduction studies and needle electromyography. On one hand, a fractionated motor nerve conduction study substantiates a demyelinizing

compression at wrist level by a prolonged distal motor latency, and on the other hand, it helps to exclude a rare simultaneous second demyelinating compression at the elbow. As mentioned above, the dorsal branch of the ulnar nerve is not affected in Guyon's channel syndrome in contrast to the ulnar neuropathy at the elbow. Consequently, sensory nerve conduction studies of the dorsal branch remain normal, whereas antidromic sensory nerve conduction studies recorded at the fifth finger are abnormal. In the case of an advanced axonal loss, needle electromyography is helpful. Signs of subacute or chronic axonal damage are limited to the intrinsic muscles of the hand with normal findings in the flexor carpi ulnaris muscle. If only the deep palmar branch is affected, a simultaneous recording of the distal motor latency over the first interosseus dorsalis muscle (deep branch) and the abductor digiti minimi muscle (superficial motor branch) can confirm the diagnosis. In the case of a demyelinating lesion of the deep branch only, the distal motor latency recorded over the first interosseus dorsalis muscle is prolonged whereas it remains normal over the abductor digiti minimi muscle. Axonal damage of the deep branch leads consistently to signs of a subacute or chronic axonal lesion only in the first interosseus dorsalis muscle [206, 207]. Recently it was reported that in patients with carpal tunnel syndrome, nerve conduction studies of the ulnar nerve can show abnormalities due to an involvement of ulnar nerve fibers at wrist level [208]. They usually disappear after carpal tunnel release [209].

8.16.5 Imaging

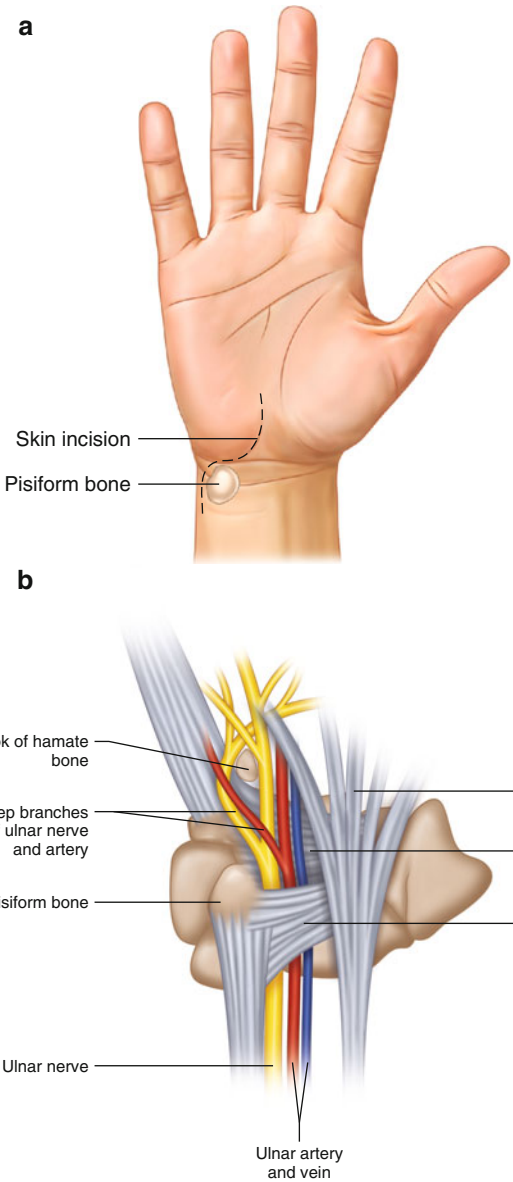
There are only a few case reports or small studies regarding the application of high resolution ultrasound and MRI to Guyon's channel syndrome. Both methods are very helpful in detecting space occupying lesions like ganglion cysts or tumors [210, 211]. Furthermore, they allow the examination of the entire nerve course up to the axilla. Perhaps rarely, different or pathologies located elsewhere can thus be detected. The normal cross

sectional area of the ulnar nerve at wrist level determined by high resolution ultrasound is 5.5 ± 2.5 mm^2 [212]. Due to the lack of systematic studies it should only be put to limited use as a cut-off value. As described above, the ulnar nerve and its terminal branches passing Guyon's channel change their course several times. This can lead to a "magic angle" artifact in MRI which looks like a T2 lesion (see Sect. 6.2.3). Two of these examples are illustrated in Figs. 8.28 and 8.29.

8.16.6 Treatment

If surgery is intended, we prefer an incision which starts over the pisiform bone, then turns to radial using the distal wrist skin crease, and then turns to distal between hyper- and hypothenar again using the main skin crease between these eminences. After dissection of a triangle flap over the hypo-thenar eminence, the fascia medial to the pisiform bone is carefully opened. The dissection thus starts proximal to the wrist skin crease in order to identify first the ulnar artery and then the ulnar nerve. The nerve is always covered by the artery which comes into view first. Now the roof of Guyon's canal is opened, always keeping in view the artery and its branches. Sometimes small arterial branches have to be coagulated and transected early enough to avoid rupture or arterial bleeding. At the end, one or two small retractors on the distal edge of your skin incision and on the retracted flap help to visualize the nerve branches and particularly the deep palmar branch whose additional compression has to be excluded. Wound closure needs a small drain, plaster immobilisation is unnecessary, and skin stitch removal is done 14 days postoperatively (Fig. 8.30).

Fig. 8.30 Surgical treatment of GUYON's canal syndrome. (**a**) Skin incision. (**b**) Principels of the exposure; nerve release first needs transection of the volar carpal ligament and, second, exposure of the deep motor branch by incising the fascial roof which covers the nerve entrance into the palm


a
Skin incision
Pisiform bone
b
Hook of hamate bone
Deep branches of ulnar nerve and artery
Pisiform bone
Ulnar nerve
Ulnar artery and vein
Palmaris longus tendon
Flexor retinaculum
Volar carpal ligament

8.17 Deep Palmar Branch Entrapment

Isolated focal lesions of the deep palmar branch of the ulnar nerve occur as frequently as whole nerve trunk entrapments within Guyon's canal. Nevertheless, these lesions are also rare.

8.17.1 Anatomy

The nerve branch supplying the intrinsic hand muscles leaves the ulnar nerve at its lateral aspect, turns a little to lateral and disappears into the deep compartment of the palm. When entering the deeper planes of the palm, the nerve passes under a typical and always consistent fibrotic arch which originates from the hamate bone and its process and from the flexor retinaculum [213, 214]. The fibers run transverse over the small nerve branch so that it is easy to see that repeated pressure episodes pinch the nerve.

8.17.2 Damaging Factors

It is well known that the syndrome occurs especially after a patient's report of a long bicycle ride, with a long time spent leaning on the extended wrist during several activities. Literature describes various additional damaging factors: a nerve branch lesion related to wrist fracture [215], concomitant harm caused by the hook of the hamate which was directed in a more horizontal direction than usual [216, 217], and of course ganglia or small neurinomas (Fig. 8.30). Early literature from 40–50 years ago is full of case reports.

8.17.3 Clinical Symptoms

The deep palmar (motor) branch of the ulnar nerve supplies the third and fourth lumbrical muscle, all the interossei muscles, and the two heads of the adductor pollicis muscle. It does not have any sensory function. Consequently, numbness never occurs whereas myokenetics of all five fingers deteriorate rather quickly. Atrophy of the intrinsic muscles is visible early and makes clinical examination necessary; pain is never complained of.

8.17.4 Electrodiagnostics

Please refer to Sect. 8.16.

8.17.5 Imaging

Please refer to Sect. 8.16.

8.17.6 Treatment

Experience in exploration of the deep palmar motor branch may differ between hand surgeons and neurosurgeons. My personal experience is to choose the same approach as the one into Guyon's canal because it secures the exposure of additional causative factors within Guyon's canal on one hand, and a reliable preservation of arterial and nerve branches in a very delicate area on the other. It should be noted that an anastomotic connection can exist between the ulnar nerve branch and the thenar muscles (Riche-Cannieu anastomosis; see Sect. 2.1), recognizing this fact is again the surgeon's responsibility.

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Chapter 9
Nerve Entrapment at Trunk and Leg

Josef Böhm, Götz Penkert, and Thomas Schelle

9.1 Ilio-hypogastric, Ilio-inguinal, Genito-femoral Nerves

The following chapter about the three inguinal nerves will be short because efforts to expose the small nerves to try neurolysis macro-surgically, or even with a microscope, are useless. We therefore resort to describing anatomical details which we find in visceral surgical descriptions and reports. Nevertheless, there are new therapeutic concepts which can simply be applied to neuropathies as we will later describe in detail.

9.1.1 Damaging Factors

Painful focal nerve lesions of one or more of the inguinal nerves result almost always from hernia surgery. When we read "Hernia Repair Sequelae" in 2010, visceral surgeons worldwide claim about 10–15 % of post-operative painful discomfort [1]. It is interesting that the different authors never distinguish between the different types of groin pain. Local pain syndromes may be due

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to post-hematoma, fluid effusion or tumor-like fibrotic conditions as a reaction on prosthetic implants. Nerve damage-related groin pain of a radiating character can instead demonstrate various pain qualities as already described in Sect. 4.2. Independent of the type of hernia surgery, herniorrhraphy or hernioplasty with mesh grafts, the rate of painful complications remains the same. Most crucial are post-hernia neuropathic pain cases. Major risk factors related to developing such a pain syndrome are: age below 40 years, pre-existing pre-operative pain, and the male gender. The investigation of mesh explants in order to determine tissue reaction led to the result that younger patients displayed a significantly enhanced infiltration of macrophages after mesh implantation. Older patients were at lower risk of reacting with inflammation on mesh material [1].

Of great importance are, first, an Italian prospective multicenter study done by Alfieri and co-workers in 2006 [2] and second, a previous report by Izard and co-workers in 1996 [3]. Intra-operative identification of all three inguinal nerves during open inguinal hernia repair with or without mesh is reported there to be able to reduce chronic groin pain to less than 1 %.

Although the results of these analyses were published years ago, incapacitating pain syndromes are continuing to occur constantly and frequently. We have therefore to suppose that ligatures or clips for mesh graft fixation or inflammatory tissue reactions are the main causative factors. Industry thus tries to influence surgeons to apply lightweight macro-porous mesh implants to reduce inflammatory reactions on the one hand, and to avoid the possibility of mechanical nerve irritation on the other.

9.1.2 Clinical Symptoms

The symptoms consist of pain, with numbness remaining in the background, weakness is of course absent. The inguinal nerves mainly carry sensory fibers so that, compared to other skin

nerves, pain is the leading complaint. It can mostly be manually triggered at the site where substantial damage exists in the abdominal wall. The nerve caliber is so small that intraneural destruction is of a high degree and, consequently, nerve axon regeneration can not be expected. The situation results in neuroma in continuity. The trigger point with radiating pain into the distribution area of the effected inguinal nerve indicates the site of the neuroma. Unfortunately, neuropathic pain types occur frequently. This pain has a burning character; slight touches are perceived as additionally painful (refer to Sect. 4.2). If causalgia occurs, the affected painful skin area is extended, a fact that no longer allows the conclusion which one of the inguinal nerves are really injured. Only the trigger point and the assessment of which skin area the pain then radiates into are helpful.

9.1.3 Electrodiagnostics

There is a significant lack of reports regarding electrodiagnostic testing of the ilio-hypogastric, ilio-inguinal, and genito-femoral nerves in the current literature. Some smaller recent studies or case reports have demonstrated abnormalities in nerve conduction velocities, needle electromyography of the abdominal muscles, reflexology (cremasteric) and somatosensory evoked potentials. All these electrophysiological studies require a technically skilled operator, but are not sufficiently sensitive and specific at the moment [4]. Whether these tests will become routine in general medical practice is uncertain.

Abnormalities in motor nerve conduction using conduction time to the cremasteric muscle, needle electromyography of the cremasteric muscle and cremasteric muscle reflex have been described in 47 % of patients which had undergone herniorrhaphy, whereas the 23 % of patients not treated surgically had problems with the genito-femoral nerve [5]. Similar results could be demonstrated under application of motor nerve conduction

studies of the ilio-inguinal nerve [6]. Stand-alone needle electromyography of the abdominal muscles supplied by the ilio-hypogastric and ilio-inguinal nerves revealed signs of subacute or chronic axonal damage in 60 % of patients with definite entrapment and in 37 % with probable entrapment [7]. In future, high resolution ultrasound guided needle electromyography or needle-electrode placement may play a key role in increasing the sensitivity and specificity of these methods. In addition, the registration of somatosensory evoked potentials may be helpful in confirming diagnosis. Therefore, in a single case report the lateral cutaneous branch of the ilio-hypogastric nerve was used for stimulation [8]. Generally, if a unilateral affection is present a side to side comparison of the electrodiagnostic results described above may help and must be closely correlated with clinical findings.

9.1.4 Imaging

High resolution ultrasound provides a simple and cost-effective way to visualize both the ilio-inguinal and ilio-hypogastric nerves [9]. A diagnostic block can be more safely established when guided by ultrasound. It is increasingly important to apply this technique simultaneously, because in general, the diagnosis is assessed on the basis of pain relief within 10 min of local infiltration with anaesthetics [10]. Therefore, the ultrasound probe should be placed with its lateral end just above the anterior superior iliac spine (ASIS) with a perpendicular orientation to the inguinal line. Now the hyperechoic surface of the ASIS and its post-acoustic shadowing is clearly visible laterally. Medially underneath the subcutaneous fat, three layers of abdominal muscles appear (from the outside to the inside): external oblique (EO), internal oblique (IO) and transverse abdominal (TA). Below the transverse abdominal muscle, the peritoneum and the movement of bowels can be observed. Usually the ilio-inguinal and ilio-hypogastric nerves

are visible between the layers of the TA and IO muscles, where a splitting of the fascia is normally present (Fig. 9.1b). Nevertheless, there also exist anatomical variants [11]. Sometimes, both nerves can pierce the IO and appear between the IO and EO muscles. They may run together or at a distance of approximately 10 mm. The technique for blocking the genital branch of the genito-femoral nerve under ultrasound guidance has not yet been published [10]. Furthermore, in some of our patients, we were able to demonstrate some pathological changes of the ilio-inguinal and ilio-hypogastric nerves. The most common reactions found included neuroma-formation and enlargement of the cross sectional area (Fig. 9.1a). Until now, major reports on 3 T-MRI of the three nerves are unavailable.

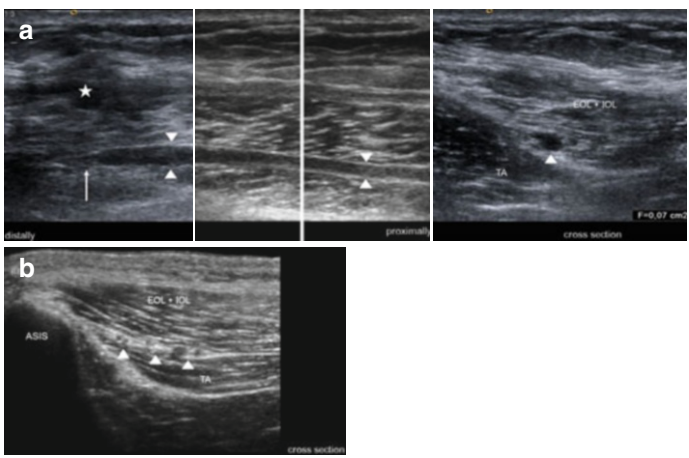


Fig. 9.1 Neuroma formation of the ilio-inguinal nerve after hernia surgery. (**a**) HRUS shows the stump neuroma (*arrow*) of the ilio-inguinal nerve (*arrowheads*) and scarring (*asterisk*) just above. (**b**) Normal anatomy. The *arrowheads* point on the ilio-inguinal and ilio-hypogastric nerves as well as on the deep circumflex iliac artery. *ASIS* anterior superior iliac spine, *EOL + IOL* external oblique and internal oblique muscle, *TA* transverse abdominal muscle

In a recent case report on nerve injury locations due to retropubic sling procedures, MRI could not evaluate any abnormalities despite severe chronic pain in symptomatic individuals [12]. Recently Chhabra and Andreisek described the assessment of all three nerves using MRN [13].

9.1.5 Treatment

Because neurolysis procedures have proved unsatisfactory and, of course, risk in resulting in hernia recurrence, circumscriptive neurotomy is performed worldwide, particularly as so-called "triple neurotomy" in the retroperitoneal space [1].

Neurosurgical pain surgeons have a completely new and comparably simple approach: the idea of pain relief by electrical stimulation of the involved peripheral nerve, a method that has been derived from the "Gate Control Theory" by Melzack and Wall in 1965 [14]. According to their concept, electrical stimulation of fast A-fibers is said to have the capacity to "gate out" pain stimuli conducted via slow C-fibers. It is well known that most pain input to the brain runs via the small unmyelinated and thinly myelinated C-fibers. This input and its transmission to the brain can be reduced by activating of fast running A-fibers. Currently, the concept of nerve stimulation is not only applied to sensory spinal cord fibers but just as successfully to injured peripheral nerves [15]. If the nerve caliber allows piercing of the epineurium, a small electrode is pushed slightly into the subepineurial space. We have demonstrated a case with superficial radial nerve branch pain in 2004 [16]. After an external test stimulation period of a few days, the external screener is changed into a pacemaker placed subcutaneously in chest or abdominal wall.

Due to their too small caliber, inguinal nerves do not allow one to advance electrodes into their sub-epineurial space.

Experience can now demonstrate that electrodes which were directly beside or in the neighbourhood of nerves did work. With reference to triple neurotomy of hernia surgeons, laparoscopic approaches into the intra-abdominal and retro-peritoneal space were made in order to place electrodes a few centimeters under the peritoneum and beside the affected nerve [17]. The idea was derived from experience with extra-uterine endometriosis-related nerve pain.

Meanwhile, a far simpler approach seems to be the gold standard: so-called subcutaneous peripheral nerve stimulation (sPNS). In local anaesthesia, the electrode is percutaneously advanced near to the location of the assumed nerve injury whereby the trigger point helps to find the site exactly. The intra-operative test stimulation can induce pleasing paresthesias in the distribution area of the affected nerve. After a short external test stimulation period, the pacemaker is – then under general anaesthesia – placed into the abdominal wall about 10 cm from the electrode. This method achieves astonishing responses by the formerly painful conditions as recently reported. In this prospective study of our center, 21 patients are included with a follow up of nearly 10 years. Of these, 76 % had a successful long-term outcome and 24 % had to be considered as long-term failures [18]. Here, after all, sPNS is simple and nearly free of risk, especially if compared with intra- or extra-abdominal approaches. Furthermore, a recent evaluation of 61 spinal cord stimulation patients resulted in the interesting fact that responses did not differ whether the stimulator was used continuously or intermittently [19]. Several of these patients were even proud to have successfully extended their stimulation-out-periods more and more. They stimulated still only facultatively, and after a few years they even requested removal of the complete system. It can be emphasized that responses on post hernia neuropathic pain syndromes are also encouraging but, of course, cases need to be further analyzed.

9.2 Lateral Femoral Cutaneous Nerve

9.2.1 Anatomy

The nerve arises from the L2-root and remains within the retroperitoneal tissue. It exits the pelvis just medial to the anterior superior spine of the iliac crest. The exit point is formed by a split in the lateral attachment of the inguinal ligament at the bone. It then angulates sharply downward, especially in extended hip joint position. The small nerve remains under the fascia and crosses over the upper portion of the sartorius muscle a little to lateral. A few centimeters below the inguinal ligament, the nerve pierces the thick fascia lata. Variations occur frequently [20]. First, the nerve course can be situated more lateral so that it crosses over the iliac bone, second, the level of division into branches varies. It can easily be imagined that a very high ramification makes the exposure more difficult.

9.2.2 Damaging Factors

Most of focal neuropathies of the lateral femoral cutaneous nerve occur spontaneously. The passage between sheaths of the inguinal ligament predisposes the nerve to become compressed or angulated during hip extension. It is often said that symptoms disappear with weight loss, but we also saw underweight patients with the same neuropathy. To summarize, in most cases reliable causative factors cannot be identified. Less common is scarring in the neighbourhood of the nerve responsible for neuralgic pain. It can result from abdominal surgery, open or laparoscopic, and from injury during removal of iliac bone parts as grafts.

9.2.3 Clinical Symptoms

Symptoms are frequently perceived as electric current-like, often suddenly radiating into the distribution area of the nerve of the anterior and lateral aspects of the thigh. The pain does not include the knee and patella region. It is worsened or induced by standing and walking, and relieved by flexion of the hip joint. If the pain history runs long enough, patients complain of additional numbness during painless periods. Numbness unfortunately indicates advanced intra-neural damage within the small nerve, and it worsens the prognosis when neurolysis is done. The electric current-like pain can be provoked by deep manual palpation immediately medial to the anterior superior spine. The same radiating pain disappears if a local anaesthetic is injected at this point. Both of these tests, palpation and injection, serve as clinical proof of the neuropathy of "meralgia paresthetica" as it is usually called. However, this term has to be restricted to those cases with spontaneous onset rather than those related to prior surgery. Those tests also act as an aid to assess the differential diagnosis between a peripheral neuropathy and an L2- or L3-root compression. Both roots have sensory distribution areas similar to that of the lateral femoral cutaneous nerve.

9.2.4 Electrodiagnostics

The lateral femoral cutaneous nerve (LFCN) remained difficult to test reliably by electrophysiological means in the past, likely due to anatomic nerve variability and lack of response in asymptomatic obese subjects [21, 22]. However, recent studies with up to date electrophysiological equipment demonstrate that responses could be obtained in at least 92 % of subjects,

even if they are obese [21]. Ultrasound-guided near-nerve needle placement and recording could provide a novel approach, especially when evaluation is critical and responses are difficult to obtain [23]. Orthodromic and antidromic sensory nerve conduction studies are frequently used in the clinical routine. The most appropriate stimulation site in an antidromic nerve conduction study is located 1 cm or more medial to the anterior superior iliac spine (ASIS) or 4 cm distal to the ASIS, whereas sensory nerve action potentials can be simultaneously recorded along an imaginary line between the ASIS and the lateral border of the patella and 2 cm medial to this line [24, 25]. However, normal values are variable and depend on the operator and recording technique. An alternative approach consists of orthodromic nerve conduction study of the lateral femoral cutaneous nerve distally to the ASIS. Of the 120 symptomatic subjects, 98 % had a side to side amplitude ratio greater than 2.3. Combined with the amplitude of the sensory nerve action potential that is lower than 3 μV, this yielded a specificity of 99 % [26]. Furthermore, recording of dermatomal somatosensory evoked potentials provides another option in the diagnosis of meralgia paresthetica. In a recent report, this technique was found to be superior to sensory nerve conduction studies: 81 % vs 65 % sensitivity [27].

9.2.5 Imaging

The lateral cutaneous femoral nerve can easily be identified in the intermuscular space between tensor fasciae latae and sartorius muscles by high resolution ultrasound [28]. Therefore, the sartorius muscle serves as an initial sonographic landmark (Figs. 9.2 and 9.3). Anatomical variations of the nerve course (e.g. piercing of the sartorius) can also be made visible. From this point, the nerve can be traced up to the inguinal ligament and to the anterior superior iliac spine. The cross sectional

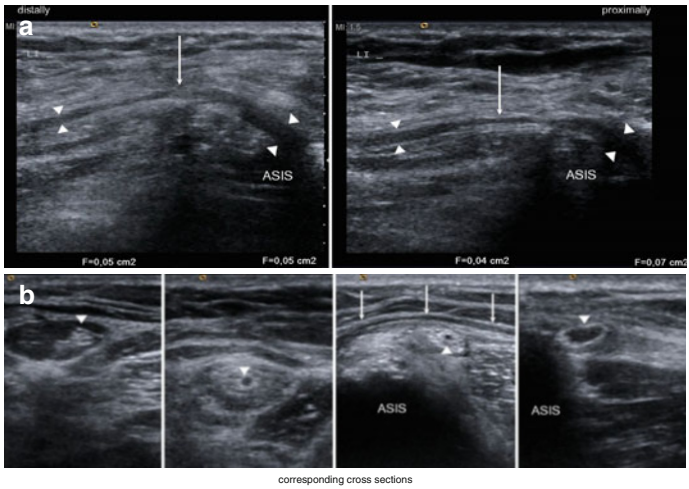


Fig. 9.2 (**a**) Longitudinal sections of entrapment of the lateral femoral cutaneous nerve (*arrowheads*) at the level of the anterior superior iliac spine (*ASIS*) below the inguinal ligament (*arrows*). Note the proximal nerve swelling. (**b**) Shows the corresponding cross sections with proximal nerve swelling and effaced fascicles, compression below the inguinal ligament (*arrows*) and the course of the nerve within tendon of the sartorius muscle and the fat-filled tunnel (from *right* to *left*). *Arrowheads*: lateral femoral cutaneous nerve

area of the lateral cutaneous femoral nerve move to behind 1.04 ± 0.44 mm^2 [28]. In symptomatic individuals suffering from meralgia paresthetica, high resolution ultrasound was able to show the enlargement of the nerve diameter and cross sectional area [29]. In a recent study the optimal cut-off value for the diagnosis of lateral femoral cutaneous nerve entrapment was 5 mm^2 [30]. In our own experience, even the demonstration of different sites of compression (e.g. below the inguinal ligament) with focal congestion of the nerve proximally to them is possible (Fig. 9.2). Moreover, high resolution sonography is not only useful in confirming the diagnosis of meralgia paresthetica but also in ultrasound guided treatment

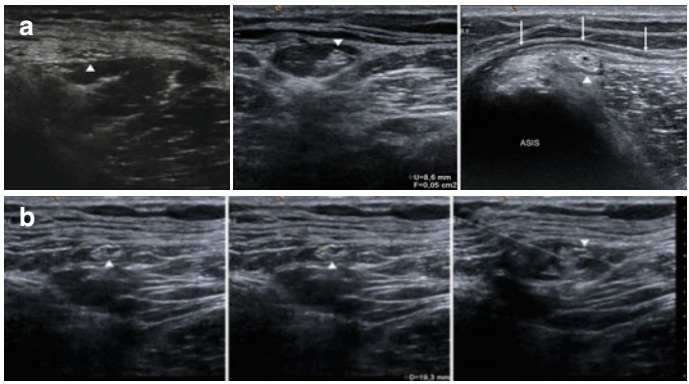


Fig. 9.3 (**a**) Several approaches of ultrasound guided injection of the lateral femoral cutaneous nerve (LFCN, *arrowheads*): *Left*: LFCN above the belly of the sartorius muscle. *Middle*: LCFN within a fat-filled tunnel. *Right*: LFCN below the inguinal ligament (*arrows*). *ASIS* anterior superior iliac spine. (**b**) Ultrasound guided injection of the LFCN using the "in-plane" scanning technique. *Arrowheads*: lateral femoral cutaneous nerve

modalities. Whereas conventional techniques (blind or nerve stimulator guided) yielded an incomplete block-success, the ultrasound guided approach was successful in all subjects [31, 32]. Pain relief following application of an anaesthetic confirms the diagnosis at first (Fig. 9.3). After that, one single or two therapeutic perineural injections of 1 mL of methyl-prednisolone acetate (40 mg/mL) and 8 mL of mepivacaine 2 % can significantly reduce symptoms over a 2-month period [32]. There exist additional case reports on patients treated successfully by means of pulsed radiofrequency ablation, and this may provide in future a relevant therapeutic alternative to surgical interventions described below [33].

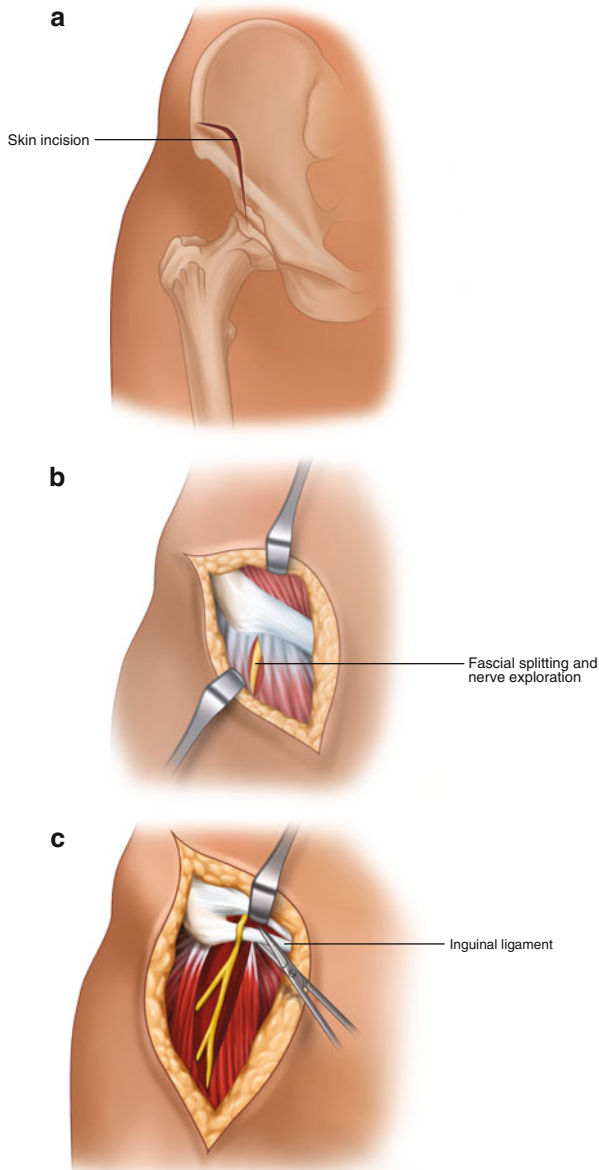
Signal intensity alterations of the lateral femoral cutaneous nerve are difficult to visualize by MR imaging owing to the small size of the nerve [34]. Accordingly, MRI is not the first choice for the diagnosis of lateral femoral cutaneous neuropathy.

However, a secondary etiology which is sometimes discussed from e.g. avulsion fractures of the ASIS, sartorius tendon injury, pelvic osteotomy, and acetabular fracture may be demonstrated by MRI [34].

9.2.6 Treatment

If conservative treatment fails (weight reduction, avoidance of tight clothing, drugs acting on neuropathic pain, ultrasound guided injections – see section above) [32], surgery is to be recommended. It consists of nerve release by transection of some parts of the lateral attachment of the inguinal ligament at the iliac bone. It sounds easy, but difficulties may occur with the small caliber of the nerve and its branches and with the above-mentioned anatomical anomalies. Therefore, repeated injections with corticosteroids or ultrasound-guided radiofrequency ablation are used and are also reported to be successful [32, 33]. Unfortunately, however, pitfalls of injection therapy trials can have a worsened prognosis if surgical neurolysis is in the end intended. First, the numbness is progressed, and, second, direct nerve substance damage can be caused by needles or injected substances. The prognosis is especially deteriorated when the character of the pain changes into a neuropathic one. We therefore prefer early surgery and perform it as neurolysis. We avoid a neurotomy so as not to risk a painful neuroma as a result (Fig. 9.4).

We personally prefer the approach which starts below the inguinal ligament. The skin incision is done anterior and a little superior of the spine and it then runs straight downward over a distance of about 4–5 cm. Next the fascia of the thigh is incised longitudinally. Beside the medial border of the sartorius muscle, the cutaneous nerve has to be identified as embedded in fat tissue; parallel to the nerve or its branches, a small venous vessel helps to find the thin nerve. After careful identification of the nerve or two or even three of its branches, we advance to


a
Skin incision
b
Fascial splitting and
nerve exploration
c
Inguinal ligament

proximal until the above-mentioned split between two parts of the inguinal ligament becomes visible. It is more important to transect the part which crosses under the angulated nerve than the part which covers the nerve course. During ligament fiber transection, the small nerve should be gently kept away a little. Particularly, if small bleedings appear at this moment, bipolar coagulation requires the nerve to be kept away. Surgical results of this approach are formally published [35].

As an alternative, some authors use a supra-inguinal approach which can facilitate nerve identification proximal to its ramification [36]. It needs the help of an assistant who keeps a retractor to medial manually whereas the infra-inguinal approach can be accomplished with self-holding retractors.

As we have experienced early recurrences following the surgery as outpatient management, we always advise hospitalization for a few days, and a suction drain to remain for no less than 3 days to avoid any re-bleeding or lymph fluid pad which likely occur in particular when the patient does not remain immobilized for long enough.

Pitfalls and pain recurrence depend on different factors, of course; on the efficiency of the inguinal ligament fiber transection, but also on the amount of intraneural damage that happened preoperatively, and, not to be neglected, on the preoperative pain type. Cases with hyperesthesia or even allodynia have a worse prognosis, whereas cases with a pain that can be triggered, and at the same time is without numbness, have the best prognosis. Injury to one of the small nerve branches can be followed by remaining neuropathic pain involving its small skin distribution, as experienced in rare cases.

Fig. 9.4 Surgical treatment of "Meralgia paresthetica". (**a**) Skin incision. (**b**) First step: nerve exposure by splitting of the ventral limb fascia medial to the sartorius muscle. (**c**) Second step: nerve exposure to proximal aiming at transection of transverse inguinal ligament parts particularly inferior to the nerve

9.3 Obturator Nerve

Focal neuropathies of the obturator nerve may commonly result from intra-pelvic trauma or surgery. As an intra-pelvic nerve exposure with neurolysis or proximal neurotomy seems to become unsatisfying, as described in comparable cases of inguinal nerve lesions in Sect. 9.1, we will neglect these almost incurable lesions in the following. However, several interesting experiences with spontaneous neuropathy exist, and they shall be covered in this chapter. Literature is rather lacking concerning these lesions.

9.3.1 Anatomy

The obturator nerve originates from the lumbo-sacral plexus (roots L2–L4). These are the same roots which contain all the fibers to the femoral nerve. It runs downward at the medial border of the psoas muscle, and then takes a vertical course to pierce the obturator membrane. At extra-pelvic level, the nerve at once divides into two branches, an anterior or superficial branch which supplies the adductor longus and gracilis muscles, and a posterior or deep branch, which supplies the adductor magnus and obturator externus muscles. Some sensory fibers represent sensation in a small skin area on the medial aspect of the thigh.

9.3.2 Damaging Factors

The possible relation of nerve injury to pelvic fractures or hip surgery has already been mentioned. Intra-pelvic malignancies may invade the nerve. Obturator hernias can involve the nerve. Newborns can suffer from obturator palsy; but they usually recover quickly [37]. Reliable factors which cause a compression syndrome with spontaneous onset are quite unknown. We

could gain experience with the relatively easy exploration of the nerve's exit site where the extra-pelvic course starts. We would like to emphasize the point that the split between obturator membrane and margin of the superior pubis bone branch is the location of a focal "spontaneously occuring" entrapment.

9.3.3 Clinical Symptoms

A small skin branch distributes a circumscriptive area at the medial and proximal aspect of the thigh. Pain is located there in the case of nerve entrapment. Numbness indicates a severe lesion and is therefore restricted to intra-pelvic nerve damage. Pathological potentials in the adductor muscles may be found in electromyography testing (see below). The diagnosis mainly consists of an infiltration with local anaesthesia at the point where we can elicit the pain by deep palpation, namely through the pectineus muscle. This test also helps to differentiate the obturator neuralgia from genital branch neuralgia.

9.3.4 Electrodiagnostics

Literature is rather sparse concerning nerve conduction studies of the obturator nerve and therefore limited in collaborative electrophysiological data. None of the few reported techniques on the subject are used routinely. A distal motor latency can be obtained by stimulating the obturator nerve at the inguinal ligament, while M-responses were recorded with needle electrodes from the gracilis muscle. An additional stimulation point – using needle electrodes at the level of the upper lumbar roots between L1–L2 vertebral laminae – generates a proximal conduction time [38]. In healthy individuals, the following normal values were reported: distal motor conduction latency 3.9 ± 0.7 ms, proximal conduction time 10.4 ± 0.3 m, and conduction velocity of the

proximal segment 62 m/s [38]. An alternative method consists of magnetic stimulation of the paralumbar area, where the M-responses are recorded via needle electrodes placed in the adductor longus and adductor brevis muscles [39]. However, needle electromyography still remains the key method of evaluating obturator nerve function preexisting axonal damage. Due to the assessment of muscles only supplied by the obturator nerve, of muscles innervated by adjacent nerves, and of paraspinal muscles, a differentiation between an isolated obturator nerve neuropathy, lumbar plexopathy, or L3-root affection becomes feasible [40].

9.3.5 Imaging

High resolution ultrasound is of subordinate importance in the diagnosis of obturator nerve neuropathy, because only the extra-pelvic nerve course can be assessed. The femoral vein and artery below the inguinal ligament serve as a landmark; by moving the probe medially and caudally to the pectineus muscle and then to the adductor muscles (consisting of the adductor longus, brevis and magnus from cranial to caudal) the nerve can easily be identified. The anterior branch of the obturator nerve with the adjacent vessels can be found in the fascia split between the pectineus, adductor longus and adductor brevis muscles, whereas the posterior branch with the accompanying vessels appears in the fascia split between the adductor brevis and magnus muscles. The following normal values (anterior-posterior diameter) have been reported so far: Anterior branch 1.4 ± 0.6 mm, posterior branch 1.7 ± 0.6 mm, main trunk 2.7 ± 1.2 mm [41, 42]. Tilting the probe cranially, the main trunk may be visible in slim individuals (Fig. 9.5). This technique is in use for ultrasound guided obturator nerve blocks [43]. As mentioned in the previous chapter, pain relief after infiltration with local anaesthetics again supports the diagnosis [40]. However, sonographical reports about extra-pelvic affections of the obturator nerve are missing at the

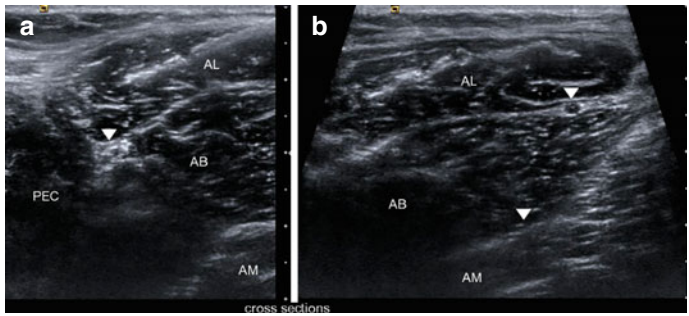


Fig. 9.5 (**a**) Normal cross-sectional anatomy of the extra-pelvic course of the obturator nerve in HRUS. The main trunk (*arrowhead*) appears caudally between pectineus (*PEC*), adductor longus (*AL*) and adductor brevis (*AB*) muscles. (**b**) More distally the anterior branch (*arrowhead*) appears in the fascia split between adductor longus (*AL*) and the adductor brevis (*AB*) muscles whereas posterior branch (*arrowhead*) can be located in the fascia split between adductor brevis (*AB*) and adductor magnus (*AM*) muscles

moment. In contrast to that, 3 T-MRI allows the assessment of the entire course of the obturator nerve: the intra- and extra-pelvic parts. Particularly, by means of MRI, space-occupying lesions such as hematoma, bone fragments, bursa, nerve sheath tumor, malignancies, or osteo-synthesis materials as well as fat infiltration just proximal to and within the obturator canal can be assessed. On the other hand, muscle substance alternation related to acute or chronic denervation may be documented [34].

9.3.6 Treatment

In the following, we restrict ourselves to describing the approach to the easily accessible extra-pelvic nerve part, the nerve segment, which may be involved in the sense of focal entrapment. We start with a skin incision parallel to the medial segment of the inguinal ligament and extend the incision vertically downward over 4–5 cm. The incision allows an approach between the

long adductor muscle and the pectineus muscle. Of course, fibers of the pectineus muscle can also be split by blunt dissection. Underneath the layer of these two muscles, we enter a flat layer of fat tissue in which the two mentioned branches of the obturator nerve are embedded. These nerve branches and the concomitant obturator artery and its branches are gently isolated, and next we follow to proximal in order to find the exit point through the obturator membrane. By means of a small punch, the lower margin of the ramus superior ossis pubis is opened in an inverse U-shape always with the exiting nerve and artery in view. The only literature concerning this approach we believe to be available is from Millesi from 1992 [44]. However, interestingly his contribution does not consider spontaneous entrapment cases at all (Fig. 9.6a, b).

9.4 Sciatic Nerve

A focal entrapment of the sciatic nerve can rarely occur within the infra-piriforme foramen. Birch specifies the historical literature which we can find dealing with the so-called "piriformis syndrome" [45]. The term is controversial in that a lot of pain syndromes in neighbourhood of the hip joint are included by different authors. In the following, we restrict ourselves to applying this term to cases which are severely associated with pain radiating under the foot, indicating a sciatic nerve irritation.

9.4.1 Anatomy

The sciatic nerve leaves the pelvis together with the inferior gluteal nerve and vessels through the lower part of the great sciatic foramen. The foramen is divided into higher and lower openings by the transverse running piriformis muscle. Its tendon

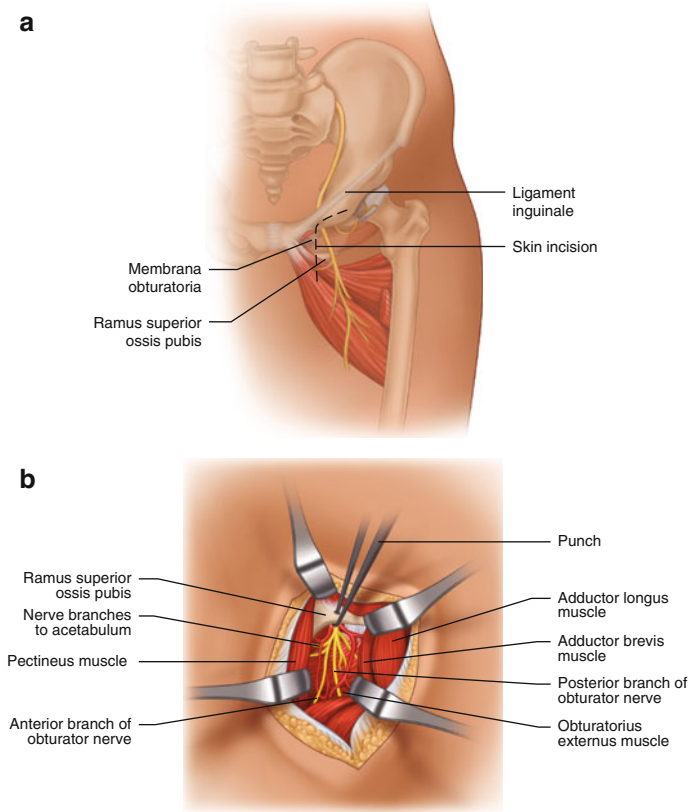


Fig. 9.6 Surgical treatment of obturator nerve entrapment. (**a**) Skin incision. (**b**) Nerve exposure via split pectineus muscle fibers; the bony roof of the nerve's exit is opened by a punch

attaches at the great trochanter of the femur. Sometimes, the tendon is extended more than normal to medial so that a fibrotic structure crosses over the sciatic nerve. As anatomical variation, the nerve may be divided into two trunks with the piriformis muscle running between both. In these cases, the superior trunk

leads fibers running into the peroneal nerve distribution whereas the lower trunk contains fibers of the tibial nerve. A surgeon should be aware of this variation which can frequently occur.

9.4.2 Damaging Factors

Completely comparable to the thoracic outlet syndrome, where fibrotic structures within the scalene muscles cause plexus irritation, here a fibrotic and too medially extended tendon within the piriformis muscle leads to discomfort. It is not the muscle belly itself, but its tendon. Small persons with little buttock substance are predisposed. Three times we have observed patients who always used to keep their purse in the same back pocket while sitting in a car or on a bicycle. They instinctively left it out sometimes and the pain progressively disappeared. Another time, we were confronted with a patient who presented with a subtotal foot drop and additional weakness of the plantar flexion on both sides after he had been confined to bed in a supine position for 2 weeks in an intensive care unit. On the other hand, cases with spontaneous onset or without any known cause have required us to find a solution.

Magnetic resonance imaging (MRI) has, of course, to exclude space-occupying lesions within the pelvic area. The main differential diagnosis is commonly a lumbar disk protrusion; the sign according to Lasegue is always pathognomic in root compression whereas likely to be negative in the case of a “piriformis syndrome”, but, unfortunately, also negative in the case of a narrowed root recess.

9.4.3 Clinical Symptoms

The patient suffers from electric current-like pain which radiates into the tibial and peroneal nerve distribution either spontaneously,

or when we palpate the gluteal area deep enough and just medial to the hip joint. A palsy of foot extension and/or flexion appears rarely, but has to be taken into account as mentioned above.

Computerized tomography- or ultrasound-guided test injections can be used to differentiate between sciatica due to either spinal root compression or piriformis irritation: A nerve root infiltration influences the afferent input from both levels, from spinal and infrapiriforme ones, whereas an infiltration into the infrapiriforme foramen cannot influence afferent fibers which are compressed superiorly; thus, a positive effect in the periphery excludes a spinal root compression. Of course, magnetic resonance imaging (MRI) of the thoraco-lumbar spine and the pelvis is mandatory to use all possible means to exclude sciatica of alternative origin.

9.4.4 Electrodiagnostics

Electrodiagnostic testing may be of some assistance in determining nerve irritation. The sciatic nerve is stimulated proximally at the gluteal fold using a 75-mm monopolar needle, and distally at popliteal level, the peroneal and tibial nerves being stimulated separately. The recording electrodes are positioned over the extensor digitorum brevis and abductor hallucis muscles for examining the peroneal and tibial portions of the sciatic nerve. Motor nerve conduction velocities (MNCV) are, for the tibial nerve division 52.8 ± ms and, for the peroneal nerve division 54.3 ± 4.4 ms [46]. MNCV of the sciatic nerve can be measured at the gluteal segment by magnetic stimulation, proximally at L5 and S1 roots and distally at the sciatic nerve at the gluteal fold followed by recording at the corresponding muscles. The mean MNCV of the gluteal nerve segment in the L5 component was 55.4+/−7.8 m/s in patients with piriformis syndrome, which was slower than the mean value of 68.1+/−10.3 m/s obtained in healthy controls. The MNCV of

the sciatic nerve in the S1 component showed no significant difference between patients and controls. A negative relation was found between the disease duration and the MNCV values in piriformis syndrome. It was concluded that magnetic nerve stimulation provides a painless, non-invasive and objective method to evaluate sciatic nerve function in patients with piriformis syndrome [47].

In clinical practice, however, commonly the electrodiagnostic studies include the sensory study of sural and further superficial peroneal nerves, and, of course, the motor studies of the peroneal and tibial nerves (see below). Abnormal SNAPs of sensory nerves imply an infraganglional lesion. H-reflexes and F-waves are unable to localize the lesion focus. Needle electromyography is the most informative in sciatic nerve lesions. Gluteal sciatic nerve irritation – such as in piriformis syndromes – can demonstrate slightly pathologic findings in semitendinosus and semimembranosus muscles, in the long head of biceps femoris, and in lower leg muscles [48].

9.4.5 Imaging

As previously mentioned in Sect. 6.1.3, the sciatic nerve course can be visualized well by means of high-resolution ultrasound distally of the gluteal fold, whereas the intrapelvic and gluteal regions hardly permit to obtain reliable information in ultrasound (Figs. 9.7, 6.8e and 6.22a).

The method of choice is MR neurography because the sciatic nerve is easily evaluated in all imaging planes due to its large size and abundant perineural fat (Figs. 6.21 and 6.22b). The nerve has intermediate signal intensity on T1-weighted images and mildly high signal intensity on fluid-sensitive images. It can be followed as it descends in the greater sciatic foramen and exits

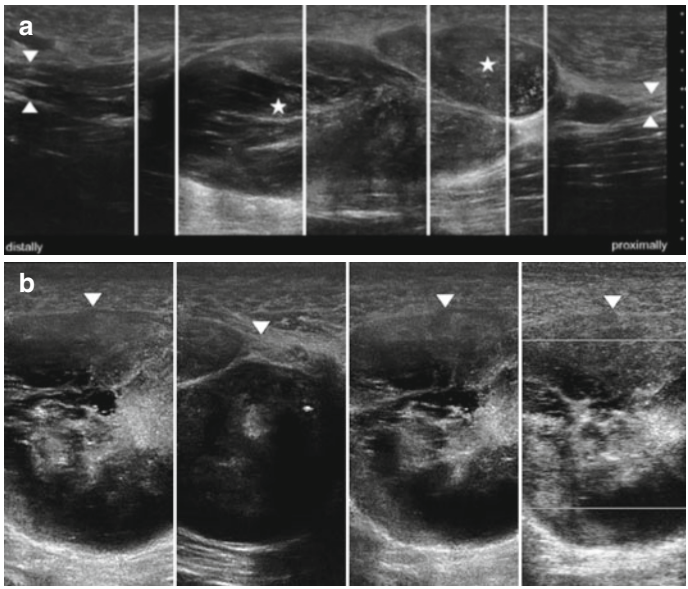


Fig. 9.7 (**a**) HRUS, longitudinal section of the sciatic nerve (*arrowhead*) demonstrating multiple large neurofibromas (*asterisk*) in a patient with neurofibromatosis type I. (**b**) Cross sections. The tumor matrix (*arrowheads*) shows no vascularization as well as regressive changes

the pelvis under the inferior margin of the piriformis muscle. Direct MR imaging evidence of sciatic neuropathy includes course deviation, increased size and increased signal intensity. The latter is more easily detected than in lower extremity nerves owing to the large size of the sciatic nerve. Obliteration of the fat planes around the nerve trunk may also be noted [34]. However, imaging diagnosis of piriformis syndrome is problematic. Although the diagnosis can be commonly inferred from changes in the appearance of the affected nerve, evaluation of

piriformis muscle size and its comparison with the contralateral side are less than satisfying, as muscle anomalies and significant variations in size are noted in symptomatic and asymptomatic individuals [34]. In a review of 100 patients with no history or clinical suspicion of piriformis syndrome, muscle size asymmetry of 2 mm was present in 81 % of patients. None of the patients with asymmetry of 4 mm or more had symptoms suggestive of piriformis syndrome [49].

Using MR neurography it was found that, of patients who responded well to piriformis surgery, 38.5 % had ipsilateral muscle hypertrophy and 15 % had muscular atrophy. Muscle asymmetry alone turned out to have a specificity of 66 % and sensitivity of 46 %, identifying patients with muscle-based piriformis syndrome. Conversely, ipsilateral nerve edema was 88 % associated with reproducible symptoms of piriformis syndrome. Use of both asymmetry of the piriformis muscle and increased nerve signal intensity improved the diagnostic ability of MR neurography, with 93 % specificity and 64 % sensitivity [50]. In a retrospectively reviewed MRN study of the sciatic nerve in the pelvis and thighs of 34 subjects, of which 17 had a final diagnosis of sciatic neuropathy according to electrodiagnostic or surgical confirmation, it was found that sciatic nerves of the affected sides exhibited higher nerve-to-vessel SI (signal intensity) ratios and, furthermore, higher incidences of T2 hyperintensity, enlargement, and abnormal fascicular shape compared to sciatic nerves of the normal side. A cut-off value of nerve-to-vessel SI ratio of 0.89 exhibited high sensitivity and specificity in predicting sciatic neuropathy. In particular, calculation of the nerve-to-vessel SI ratio demonstrated excellent reliability. The authors conclude that both qualitative and quantitative criteria should be used to suggest the MRN diagnosis of sciatic neuropathy [51]. Nevertheless, most authorities agree that delineation of the neuromuscular relationship will turn out to become the most important goal of imaging. Until reproducible and reliable criteria have been established, piriformis syndrome will remain a diagnosis by exclusion [52].

9.4.6 Treatment

CT- or ultrasound-guided botulinum toxin injections into the piriforme muscle belly are performed repeatedly with success. However, the first surgical approach was described by Robinson in 1947 [53]. He sectioned the piriformis muscle a few centimetres beside its attachment at the great trochanter completely. He chose a lateral transection in order to assure the preservation of all major vessels which lie concomitant to the sciatic nerve and its branches. The same author describes several anatomical variations as existent in 10 % of specimens, e.g. the division of the sciatic trunk.

We prefer surgery and choose a skin incision parallel to the expected fibers of the gluteus maximus muscle; then the muscle fibers are split and separated to both sides and manually retracted. Fibers of the gluteus medius muscle which are situated underneath can be lifted up and also retracted. The sciatic nerve trunk can be felt medial to the hip joint as a longitudinally downward running structure of index finger diameter. It is embedded in fat tissue and accompanied by the inferior gluteal artery and a venous plexus. The dissection to proximal requires care to preserve as much of this plexus as possible. Transverse vessels have to be visualized before rupture, closed by bipolar coagulation far enough from nerve fibers, and finally transected. Vessel rupture immediately colours the operating field black. The sensory posterior femoral cutaneous nerve branch which runs downward and medial to the sciatic nerve has to be preserved; its injury can result in numbness or even neuropathic pain at the back of the thigh and in the perineum via a small perhaps not unimportant nerve branch. By proceeding to proximal, the transverse running fibrotic lower edge of the piriformis should come into view, and it then requires transection piece by piece above the exiting nerve. However, before doing so, the inferior gluteal nerve should be visualized; it curves around the lower edge of the piriformis and then upwards. The muscle belly itself can of course be preserved,

comparably to the situation in the scalene notch, as muscle fibers do not irritate nerve structures at all.

It may rarely happen that a large venous convolute occurs within the infrapiriforme foramen on the left side. This rare phenomenon is related to the possible venous "pelvic congestion syndrome" which can occur by a so-called "nutcracker-phenomenon" – a compression of the left iliac vein between lumbar spine and left iliac artery or of the left renal vein between aorta and superior mesenteric artery [54, 55].

As it is almost impossible to preserve the whole venous plexus during surgery, a deeply advanced suction drain of large caliber is mandatory to avoid hematoma-complications, especially when the patient gets up. As a final remark concerning our indication, surgical treatment is only being considered when typical electric current-like radiating sciatica is present. Our goal cannot be anything else than decompression of a focal nerve entrapment. Pain syndromes which instead remain in the buttock – e.g. inflammatory tendopathies – do not offer any surgical option at the present time.

9.5 Pudendal Nerve

9.5.1 Anatomy

The pudendal nerve arises from the second, third and fourth sacral nerve roots, leaves the pelvis through the great sciatic foramen, and re-enters the pelvis through the lesser sciatic foramen. The fibrous canal between both foramens is referred to as the Alcock canal; therefore, neuropathies of the pudendal nerve were described as Alcock's canal syndrome [56]. The nerve supplies half of the perineum and genital region with sensory fibers. Its focal entrapment evokes intractable pain attacks in

this area. The clinically important fact that the perineum is also supplied by a small branch of the posterior femoral cutaneous nerve should never be neglected (Fig. 9.8a–d).

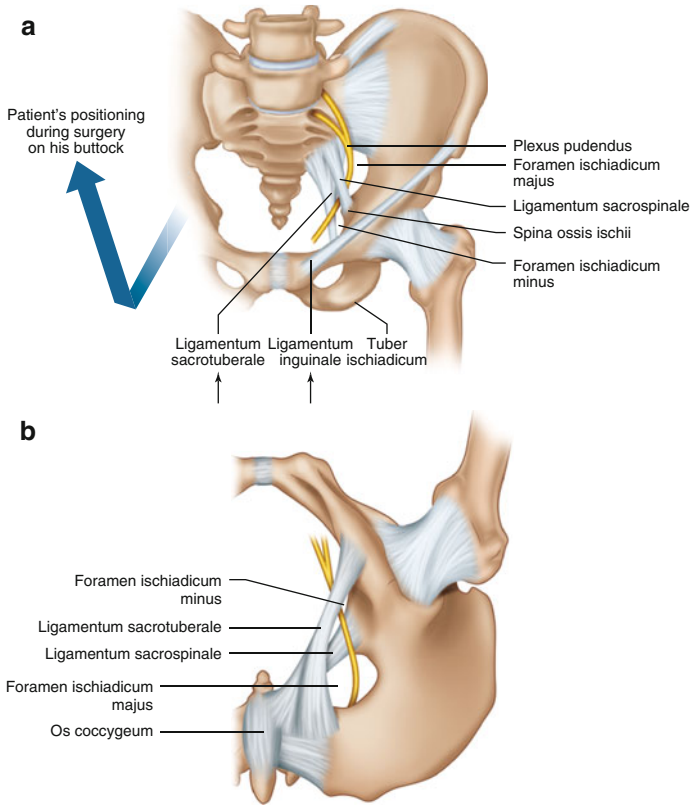


Fig. 9.8 (**a**) Anatomy of the pudendal nerve course. (**b**) Anatomy after patient's positioning on his back with lifted legs; nerve's relation to sacrospinal and sacrotuberal ligaments. (**c**) Skin incision. (**d**) Nerve exposure and transection of the sacrotuberal ligament under the gluteus maximus muscle fibers; the ligament is, if existent, to be identified by your fingertip

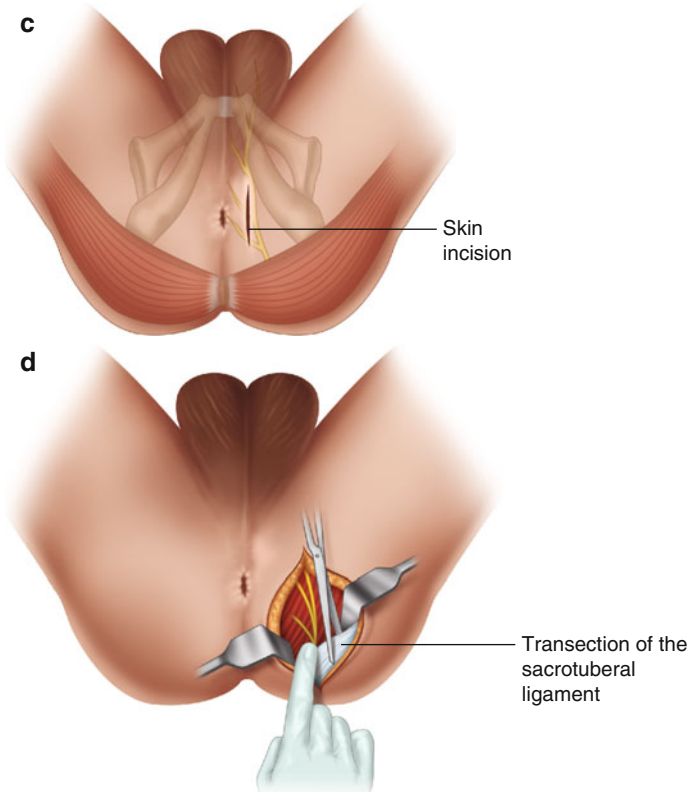


Fig. 9.8 (continued)

9.5.2 Damaging Factors

Of course, trauma relation or iatrogenic damage and space-occupying lesions can affect the nerve. However, on the other hand, prolonged bicycle rides have been already repeatedly described [57]; similar histories have also been observed by us. This mechanism should be assessed as the main causative factor

when a neuropathy seems to appear spontaneously. The nerve is then trapped between the ischial spine and the sacro-tuberous ligament. In particular, the falciforme lower edge of the ligament is able to pinch the nerve if pressure arises between body and bicycle saddle. A figure in Steward's book gives us a reasonable impression of this entrapment site [58].

9.5.3 Clinical Symptoms

Patients describe unilateral, electric current-like, pain within the genital region and perineum with sudden onset comparable to the "tic doloureux" in a trigeminal neuralgia. Only one of our patients –a small person with little buttocks – suffered from alternating pain attacks on both sides.

A more or less continuous pain which remains restricted to the perineum might be of another origin: This restricted pain area can likely belong to the distribution of the perineal (cluneal) branch of the posterior cutaneous nerve, as mentioned in Sect. 9.4.

When the patient lies supine with his hip joint 90° flexed so that the leg stands vertically, the typical Tinel sign can be elicited about three fingertips beside the anus. It indicates the lower edge of the sacro-tuberous ligament. A careful test infiltration – perhaps ultrasound-guided – with local anaesthesia at this point removes the pain attacks for 2 h and creates a genital numbness instead. In a recent paper, these nerve blocks even resulted in therapeutic effects in 44 % of cases [59]. Only patients who satisfactorily responded to our test infiltration at the described site were selected for a small surgical decompression.

9.5.4 Electrodiagnostics

According to the anatomical course of the pudendal nerve, several sites of compression can lead to various clinical

presentations of pudendal neuropathy [60]. As mentioned in Chap. 5, the unmyelinated C-fibers, carrying pain sensation are not assessable with conventional nerve conduction studies, whereas fecal and urinary incontinence, impotence and numbness of the skin area supplied by the nerve thereby may be relevant. In the past two decades, different electrodiagnostic techniques have been introduced, but most of them are not in general use. Needle electromyography of the anal sphincter, registration of terminal motor latency of the pudendal nerve after transrectal electrical stimulation, as well as somatosensory evoked potentials of the pudendal nerve may be helpful for making a diagnosis. In particular, a prolonged terminal motor latency and signs of anal sphincter denervation in needle electromyography suggest an involvement of the anal motor nerve, whereas changes in somatosensory evoked potentials of the pudendal nerve may verify a sensory neuropathy [61]. However, in 2008, a group of experts published five essential (exclusively clinical) diagnostic criteria for pudendal neuralgia by focal nerve entrapment ("Nantes criteria"): "(1) pain in the anatomical territory of the pudendal nerve; (2) worsened by sitting; (3) the patient is not woken at night by the pain; (4) no objective sensory loss on clinical examination; (5) positive effect by anaesthetic pudendal nerve block" [62]. In this set of criteria, electrodiagnostic testing plays only a subordinate role. We have unfortunately to realize that our electrodiagnostic techniques do not differentiate between entrapment and alternative nerve lesion (especially obstetrical damage in uni- or multiparous women). Therefore they do not give any direct information about pain etiology. Consequently, they have a rather limited sensitivity and specificity. In fact, they might perhaps be helpful before surgery in order to assess the quality of motor innervation of the nerve, and they might predict the outcome [63].

9.5.5 Imaging

In a recent report it has been shown that high resolution ultrasound can identify the pudendal nerve at the ischial spine when using a medial approach, and its terminal branches when using an anterior approach [64]. Unfortunately, its value in the diagnosis of different pathological processes involving the nerve, especially if entrapment is supposed, remains unclear at the moment. Until now, pain relief after blocking the pudendal nerve with anaesthetics is the superior diagnostic criterion in pudendal neuralgia [62]. Sonography may also be applied for ultrasound guided nerve blocks. Using colour Doppler to localize the internal pudendal artery at the ischium, in a second step, the sacro-spinous and sacro-tuberous ligaments have to be targeted. The pudendal nerve may be found in the plane between these two ligaments [10]. Regarding pudendal nerve entrapment and MR imaging, it was recently reported that the neurovascular bundle can be visualized by MR neurography between the ischial spine and the Alcock canal. In the case of entrapment, it is reported as typical to find asymmetric nerve swelling and hyperintensity on T2-weighted and fat-saturated images [65].

9.5.6 Treatment

Therapeutic concepts extend from repeated steroid injections via trans-vaginal approach, CT-guided trans-perineal injections [66], surgical decompression via trans-perineal approach [67], trans-gluteal approach with exposure of the entire Alcock's canal [56], until neuro-modulation via electrodes on sacral nerve roots [68] or even on the spinal cord [69]. Of course, we have first to consider that a disorder of the pudendal nerve distribution can present

a wide field of individual symptoms, and, second, that it is commonly combined with progressed social disintegration. Therefore, dedicated therapeutic approaches and options can only be offered where centers deal with all types of pudendal neuralgia [70]. Particular difficulties arise when the pain has achieved neuropathic character. Our own experience is exclusively restricted to those cases which could be manually triggered at the lower edge of the sacro-tuberous ligament, and which could be tested there by local anaesthetics.

The surgical approach to this focal entrapment site is very simple. It can be easily derived from anatomical figures [72]. The patient's positioning is comparable to that needed in proctologic approaches. The skin incision is done approximately 3 cm beside the anus and a little bit dorsal to respectively below it. The lower edge of the gluteus muscle fibers is separated from fat tissue. At this point, the trapping ligament underneath the muscle should be palpated with a fingertip. In a few operated cases, the ligament as a solitary structure was absent, but instead, several transverse running fibrous bands could be palpated. The pudendal nerve exits upon the ligament or the fibrous bands when we bring to mind the patient's position in the operating theatre. Identification of the pudendal nerve is not as easy as expected because it is embedded in abundant fat tissue and therefore difficult to survey. However, consideration of the preoperatively discovered trigger point in relation to the position of the anus finally helps to isolate the nerve. The last surgical step is partial or complete transection of the sacro-tuberous ligament where it covers the nerve's exit (Fig. 9.8c–d).

As mentioned, several specialized centers exist worldwide where experts of different disciplines work together in the field of pudendal neuropathy. However, around 60 % of these cases are suitable for trying the simple approach described above [70]. Our own experience is rather small, but at least it proved successful provided we remained restricted to neuralgias which

originated from the nerve's exit zone. This chapter shall not in any case cover the whole entity of pudendal neuropathies.

9.6 Femoral Nerve

The femoral nerve is never affected by a spontaneously occurring irritation or entrapment only. Pre-formed anatomical structures like tendons, ligaments or a narrowed channel which could irritate the nerve in a sense of focal neuropathy do not exist. Therefore, our description remains short.

9.6.1 Anatomy

The femoral nerve arises from the L3 and L4 spinal roots, and it then starts downward at the lateral aspect of the psoas muscle. Its course is covered by the iliacus fascia. Under the inguinal ligament it exits the pelvis lateral to the femoral artery and vein. Then it divides at once into several motor branches which supply the quadriceps muscles, and into sensory branches which innervate the skin of the ventral thigh medial to the area of the lateral femoral cutaneous nerve. One of its branches – the saphenous nerve – will be the subject of the next main section.

9.6.2 Damaging Factors

We have already mentioned that entrapments of focal character with spontaneous occurrence do not exist. Nontheless we once tried a single neurolyisis under the inguinal ligament in a patient with progressive loss of quadriceps muscle strength without success. The young man probably

suffered from one of the inflammatory disorders we will describe in Chap. 12.

Reasonable femoral nerve impairments are instead the following: penetrating injuries in the groin, or below, at the thigh, lesions in association with pelvic fracture, inguinal node resection, femoral artery puncture, all kinds of intra-pelvic surgery, hip arthroplasty, hematoma or abscess beneath the iliacus fascia, false aneurysms following rupture of an aortic aneurysm, malignancies of the iliac bone or other space-occupying lesions like harmless cysts or ganglia. In 1934, anatomists described those rare cysts as originating from occasional communications with the hip joint [72]. In addition, in 1982, a possible way to evaluate these rare lesions by ultrasound was published; the paper again indicates that this type of investigation is easily applicable [73]. Therefore, in general, if a femoral neuropathy occurs, imaging seems mandatory to discover the reason before surgery is considered.

9.6.3 Clinical Symptoms

Patients always suffer from loss of muscle strength of the quadriceps. The disability more or less keeps the knee joint extended and impairs patients' standing and walking. Numbness and paresthesias are not the main complaints, but rather the weakness. The knee reflex can be absent.

9.6.4 Electrodiagnostics

Motor evoked responses can be detected with surface electrodes located over the vastus medialis muscle using reference electrodes over the patella. The deep location of the nerve often requires high current intensities at the three recommended

stimulation sites above and below the inguinal ligament and at Hunter's canal along the medial aspect of the thigh. The distance between the proximal stimulation sites should be at least 5–6 cm. The latencies from above and below the inguinal ligament are 7.1 ± 0.7 ms and 6.0 ± 0.7 ms [46], respectively. Evaluating the CMAP from the two sites allows quantification of the degree of axonal loss. Femoral neuropathies which allowed estimation of its axonal loss of ≤50 % turned out to have a fine prognosis with improvement within 1 year; fewer than half the patients with axonal loss >50 % can expect to improve to some extent. Irrespective of the origin, spontaneous functional improvement was seen in two out of three patients; and assessment of amount of axonal loss was the only prognosis predicting factor [74]. In pure femoral neuropathy, the quadriceps and sartorius muscles show pathological results in needle electromyography, while the adductor muscles and the lower limb muscles remain normal. Additional involvement of the iliopsoas muscle reveals a lesion located more proximally. The differentiation from lumbar plexus lesions may be quite challenging and is needed to investigate paraspinal muscles, the adductor muscles, which are supplied by the obturator nerve, and also sciatic nerve innervated muscles [75].

9.6.5 Imaging

The femoral nerve can be visualized by high-resolution ultrasound (HRUS) in the inguinal region where it lies superficially. In a case report even a retroperitoneal femoral nerve injury after perforating injury at the lower abdomen could be demonstrated. A mass was observed in the retro-peritoneum, indicating a traumatic neuroma composed of disordered fascicles [76]. In our own experience, nerve lesion after angiography can easily be visualized (Fig. 9.9).

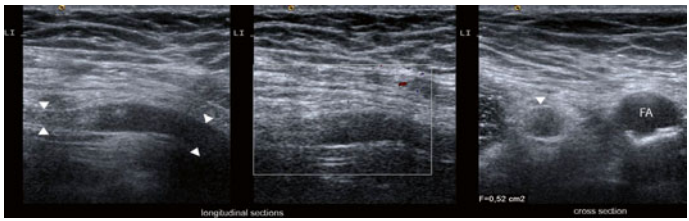


Fig. 9.9 Neuroma in continuity of the femoral nerve (*arrowheads*) at the level of the inguinal ligament in HRUS. *FA* femoral artery. *Left*: longitudinal section, *middle*: longitudinal section with color-flow-doppler, *right*: cross-section

However, 3-T MRN as imaging seems to be the method of choice, because it can assess the nerve appearance in its pelvic course, where the sonography is uncertain (Fig. 6.17). A comprehensive review describes different types of abnormalities involving the femoral nerve and illustrates their 3-T MRN features with relevant case examples. Nerve compression may result from space-occupying lesions, such as hematoma, iliopsoas bursitis, neoplasm or psoas muscle abscess. Mild to moderate nerve abnormalities on MRN should be distinguished from nerve discontinuity resulting from severe injury which requires immediate surgery; fascicular morphologic abnormalities become very evident. Multiplanar reconstruction using 3D SPACE images is particularly helpful to differentiate between neuroma in continuity and physical nerve discontinuity. Extensor compartment muscle denervation changes serve as useful secondary signs of nerve abnormality [77].

9.6.6 Treatment

As the location of a focal femoral neuropathy is common within the pelvis and retroperitoneal, the approach nearly always requires opening of the retroperitoneal space. If unfamiliar with

the exposure, a general surgeon should be requested to help. The approach also needs final proper wound closure to preclude an incisional hernia. For further details of this access refer to the work of Kline and Hudson from 1995 [78].

The management of trauma-related femoral nerve lesions needs extended nerve trunk exposure over its whole distance so that a two-portal approach above and below the inguinal ligament becomes unavoidable. However, it is not purpose of this book to describe the treatment of these extensive lesions related to the therapy of nerve traction during hip joint surgery. Iliacus hematomas, however, need prompt evacuation via the retroperitoneal approach, as recovery of nerve function then seems better than under more conservative treatment [79].

9.7 Saphenous Nerve with Infrapatellar Branch

9.7.1 Anatomy

The saphenous nerve arises from the femoral nerve a few centimeters below the inguinal ligament and runs downward alongside the femoral artery. In the lower third of the thigh, nerve and artery enter a channel of more or less 10 cm extension (Hunter's channel). The nerve has an exclusively sensory function at the medial aspect of the lower leg. When exiting the channel the infrapatellar nerve leaves the saphenous nerve as a branch supplying the skin beneath the patella. Sometimes this branch pierces the facial roof of Hunter's channel.

9.7.2 Damaging Factors

In our experience, older patients are commonly involved. Frequently, an arteriography of the femoral artery has previously been done to find or exclude peripheral arteriosclerotic

occlusion, and sometimes interventional procedures of thrombus removal have been added. In any case, a severe femoral vessel sclerosis with loss of tissue elasticity has always been found during exposure. Nerve irritation then started between the sclerotic vessel and the channel's roof. On the other hand, vein removal for use as arterial graft has sometimes resulted in saphenous pain syndrome [80].

Focal neuropathies of the infrapatellar branch mostly result from arthroscopic knee joint surgery. Unfortunately, substantial nerve tissue damage has then occurred with the frequent consequence that neuropathic pain appears under the patella.

9.7.3 Clinical Symptoms

Severe pain in the nerve distribution of the lower leg is the main complaint, perhaps aggravated by walking [81]. It can occur in the form of attacks, or it exists permanently, and it can be triggered with your fingertips at the level of the Hunter's channel. Sometimes the triggered pain radiates retrograde to proximal. Test infiltrations into the channel to confirm the diagnosis involve risk of bleeding from the accompanied artery. In contrast to involvement of the lateral femoral cutaneous nerve, for instance, we avoid infiltrations.

The infrapatellar nerve branch commonly suffers from a severe intraneural lesion when it has been injured. The result is very frequently that either a neuroma pain type or, unfortunately, a neuropathic pain type appears (see Sect. 4.2). Simple entrapments are unknown in our experience.

9.7.4 Electrodiagnostics

In order to assess antidromic saphenous nerve conduction a proximal and a distal recording method has been developed.

In the case of proximal recording, the nerve is activated along the medial aspect of the knee between the sartorius and gracilis tendons 1 cm proximal to the inferior border of the patella while the recording electrode is positioned 15 cm distal from the stimulation point at the medial border of the tibia. The values of peak-to-peak amplitudes are 10.2 ± 0.2 μV and of conduction velocity 58.8 ± 2.3 m/s [46]. In the case of distal recording, the nerve is activated between the medial border of the tibia and the gastrocnemius muscle – 14 cm proximally to the recording electrode of the previously described proximal method – while the recording electrode is positioned 3 cm proximal to the medial malleolus and posterior to the tibialis anterior tendon. The reference values of the peak-to-peak amplitudes are 9.0 ± 3.4 μV ms and of conduction velocity 41.7 ± 3.4 m/s [82].

9.7.5 Imaging

Ultrasound examination of the saphenous nerve is difficult because of its small size and anatomic variability. The sartorius muscle is identified first as an anatomic landmark. This long and flat muscle can be identified at once at the medial aspect of the knee: it is the only muscle located anterior to the gracilis and semitendinosus tendons. The muscle is then followed cranially. Attention is then focused on the deep surface of the muscle, where the saphenous nerve can be found running close to the deep fascia. In a more distal location the nerve can be visualized running between the sartorius and gracilis tendons to reach the subcutaneous tissue where they join the great saphenous vein. Detection of its thin infrapatellar branch can be very difficult [83]. In a cadaveric anatomical study, HRUS was performed on five cadavers and ten healthy volunteers. Useful landmarks to detect the nerve branch could be defined. Some anatomical variations were noted. It was concluded that the infrapatellar branch can be depicted by ultrasonographical means [84].

9.7.6 Treatment

Surgery on saphenous nerve entrapment requires opening of Hunter's channel. A longitudinal skin incision is done over the lower third of the medial aspect of the thigh. The sartorius muscle covers Hunter's channel and, when it is kept away, the surgeon's fingertip feels pulsation of the femoral artery. Try first to isolate the artery, and afterwards the concomitant nerve where they enter the channel. With nerve and artery in view, the fascial roof of the channel is incised and finally sectioned transverse to its fibers as for the transverse carpal ligament fibers. The underlying sclerotic femoral artery has to be kept permanently in mind during nerve relief. Additional care should be taken to the infrapatellar nerve branch whose injury would induce the next pain syndrome. Wound closure needs to leave behind all fascial structures unclosed, and it needs a suction drain for 2 days (Fig. 9.10a, b).

The treatment of an injured infrapatellar branch remains controversial to date. Nerve repair can duplicate the pain intensity; the same holds true if a neuroma is exposed and removed. Shortening of the nerve branch is restricted up to the level where it arises from the saphenous nerve, a region where soft tissue is sufficiently lacking so that the new neuroma remains between skin and femur epicondyle. Nerve stimulation placed along the saphenous nerve below the inguinal ligament proved is more successful in such a case.

9.8 Common Peroneal Nerve at the Knee

9.8.1 Anatomy

The common peroneal nerve arises from the sciatic nerve at its mid-thigh level, turns a little to lateral and further superficially into the popliteal fossa, passes around the fibula bone about 1–2 cm below the tibio-fibula joint, and finally enters the compartment

of the peroneal muscles. There it divides into two main branches, the superficial and deep ones. Both branches together supply muscles which extend the foot and toes. Further anatomical details have only importance when nerve repair is needed. Focal entrapments remain instead restricted to the site where the main nerve turns around the fibula and makes an almost 60° angled turn to ventral into the peroneal compartment.

9.8.2 Damaging Factors

The literature is full of causes or events that impair the common peroneal nerve at the above-mentioned exposed site [80]: external compression in operating theatres, due to plaster casts or

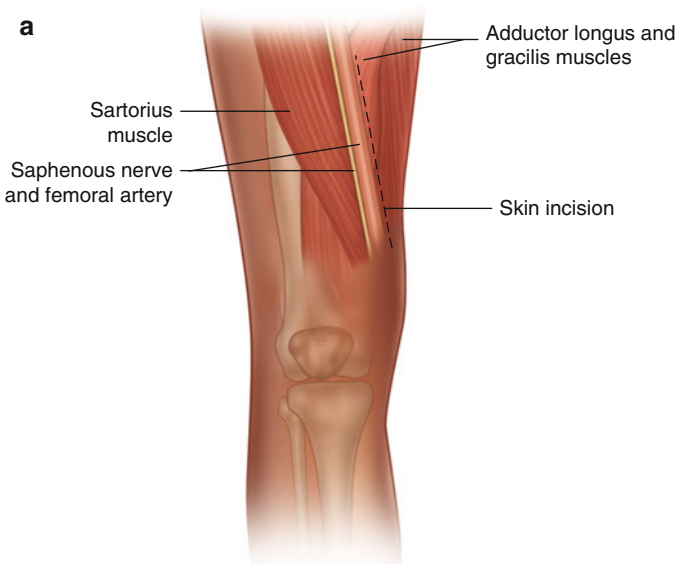


Fig. 9.10 Treatment of saphenus nerve neuropathy. (**a**) Anatomical situation and skin incision. (**b**) Nerve release by transection of the adductor magnus tendon beginning at its tendinous hiatus

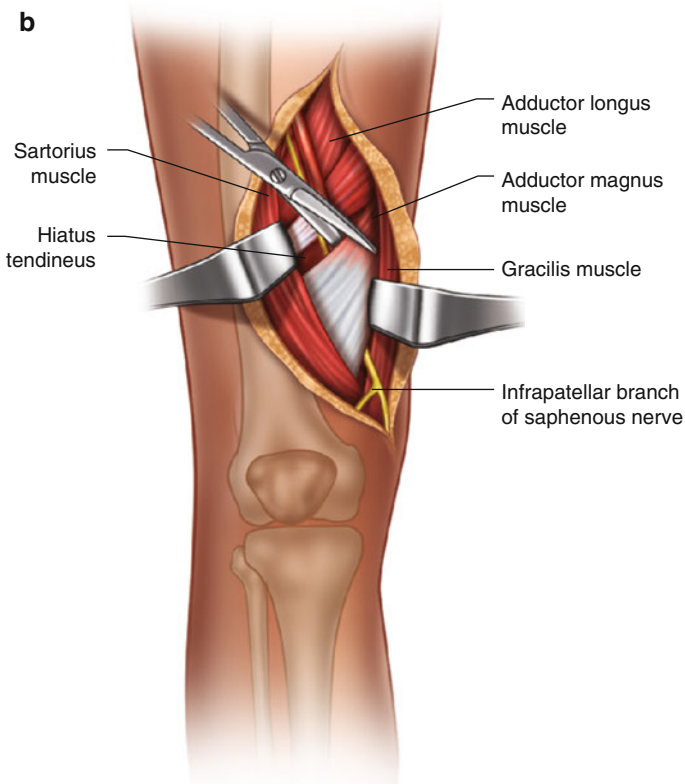


Fig. 9.10 (continued)

braces, following habitual leg-crossing or sitting cross-legged, after prolonged kneeling, frequently in relation to special professions like floor-tilers. Trauma-related nerve lesions are mostly not focally restricted but far extended like typical traction lesions usually are.

The nerve may occasionally be compressed by a variety of space-occupying lesions like intraneural or extraneural ganglia,

Baker's cysts, hematomas or hypertrophic tibio-fibula joint arthrosis. With or without such lesions, patients often notice the symptom of a peroneal palsy after awakening from a normal sleep. Such palsy is probably the result of sleeping on one's side. We remember a patient who used to press his knee against the left door continuously during professional car driving. However, most patients usually do not remember any possible causative factor. The physician therefore needs the patience to enquire about their habits, because a peroneal palsy almost never occurs really spontaneously.

9.8.3 Clinical Symptoms

One day during walking, the patient notices foot drop and finds himself in danger of stumbling. Slight paresthesias or numbness are additionally and often observed much later. The paresthesias can always be triggered at the typical compression site 2 cm below the prominent tibio-fibular joint. The typical occurrence of this Tinel sign is completely comparable to that just so often found in ulnar neuropathies at the elbow.

Ganglia cannot be detected by palpation because they are often hidden behind the fibular bone. Therefore, preoperative ultrasound imaging should be used in future, although, on the other hand, surgical exploration often enables to detect these lesions early. However, when we surprisingly find intraneural ganglia whose existence should always be considered, we will suddenly be confronted with the requirement for time consuming microsurgery (see Sect. 13.3.8).

Symptoms of "idiopathic" palsies and those related to space-occupying lesions do not differ from each other at all. Therefore, if preoperative differential diagnosis between both is required, and if uncomfortable findings during surgery are to be avoided, imaging becomes mandatory.

9.8.4 Electrodiagnostics

Demyelinating lesions of the peroneal nerve (e.g. due to compression neuropathy) across the fibular head are assessed with fractionated motor nerve conduction studies, whereby the recording electrode is placed over the extensor digitorum brevis (EDB) muscle, while nerve stimulation starts at the ankle and is then followed by stimulation below and above the fibular head [85]. Normal values have been published in 1999: distal motor latency 4.8 ms ±0.8, CMAP amplitude <40 years 6.8 mV ± 2.5, CMAP amplitude >40 years 5.1 mV ± 2.5, motor nerve conduction velocity 44–49 m/s ±4–5, normal amplitude difference from side to side 61 % [86]. A segmental delay of motor nerve conduction velocity by more than 10 m/s and/or a segmental drop in amplitude by more than 25 % (conduction block) are common findings in case of demyelinating lesions [85, 86]. However, in some previous studies, recording from the EDB was found to be less sensitive because only 33 % of affected cases were detected [87]. Thus, an additional motor nerve conduction study recording over the tibialis anterior muscle (TA) and stimulating below and above the fibular head has been suggested due to the fact that motor fibers supplying the EDB and TA may be damaged differently [85, 88, 89]. In addition, some anatomical variations can potentially disturb the motor nerve conduction study to EDB only. In fact, in 12.2 % of healthy individuals, an accessory deep peroneal nerve innervates the EDB. In this variability, no compound muscle action potential can be obtained while stimulating the nerve at the ankle, whereas stimulation below and above the fibular head generates a compound muscle action potential. Stimulation of the accessory deep peroneal nerve behind the lateral malleolus may solve this problem. Second, in 3.5 % of the healthy subjects, an agenesis of the EDB exists, and no compound muscle action potential can be obtained. Neither by stimulating the nerve at

the ankle nor below and above the fibular head [90]. Finally, overlying diseases such as chronic compression by footwear or a polyneuropathy may affect the EDB.

Therefore, normal values for the tibialis anterior muscle have also been published in 2003: upper limit of distal motor latency to TA 4.9 ms, lower limit of CMAP amplitude 1.7 mV, lower limit of motor nerve conduction velocity 43 m/s, upper limit of side to side CMAP amplitude difference 50 %, upper limit of normal drop of amplitude from below to above fibular head stimulation 36 % [91]. A further alternative method, which is considered to be more specific, is the short segment stimulation after successive supramaximal stimuli of the nerve at 2 cm intervals, starting 4 cm distal and ending 6 cm proximal to the fibular head prominence [92].

As already explained in Chap. 5, a pure axonal peroneal nerve lesion will result as a drop or absence of compound muscle action potentials at all stimulation sites. Thus, no exact localization of the site of the nerve damage will be feasible with fractionated motor conduction studies. However, in this situation, needle electromyography is mandatory: the distribution of signs of denervation and chronic neurogenic changes in different muscles supplied either by the deep peroneal nerve (e.g. tibialis anterior, extensor halluces longus) or by the superficial peroneal nerve (e.g. peronaeus longus). Or perhaps also by the peroneal part of the sciatic nerve (e.g. caput breve m. biceps femoris), by the lumbar plexus (e.g. gluteus medius), and, finally, by the L5-root innervated autochthonous para-vertebral muscles may localize the lesion. Needle myography will thus close the diagnostic gap [85].

The sensory function of the superficial peroneal nerve is easily assessable with an antidromic sensory nerve conduction study [85]. If the study is pathologically changed it indicates an infraganglionic lesion, whereas in the case of L5 radiculopathy, the SNAP should be preserved in the majority of cases. Furthermore, it has been recently demonstrated that even SNAPs of main

branches of the superficial peroneal nerve, of both the medial and intermediate dorsal cutaneous sensory nerves, can easily be obtained by antidromic motor nerve conduction studies [93]. Technically more challenging and time-consuming are orthodromic nerve conduction studies with near nerve needle placement. This method is applicable on the superficial and deep peroneal nerves in certain patients, where the standard tools described above may not solve a complete differential diagnosis in common peroneal nerve neuropathy [87, 94].

In summary, if a peroneal nerve entrapment at the fibular head is assumed, initially, a fractionated motor nerve conduction study to the EDB may be helpful. If this does not localize the lesion, the peroneal motor nerve conduction study should be repeated recording the TA [85]. Finally, a fractionated orthodromic sensory nerve conduction study with near nerve needle placement could provide an alternative method, but it is more challenging and time-consuming [87, 94]. Axonal lesions are assessable with needle electromyography. By analysing the pattern of possible axonal damage in different muscles a mono-neuropathy of the peroneal nerve can be differentiated from other disorders, e.g. sciatic nerve neuropathy, lumbar plexopathy and L5 radiculopathy. Antidromic sensory nerve conduction studies are helpful in examining the superficial branch of the peroneal nerve and excluding a supra-ganglionic lesion. Furthermore, if a generalized and overlying disorder is supposed, extension of the examination including the opposite side and other peripheral nerves is mandatory [85].

9.8.5 Imaging

Both MR-neurography and sonography are applicable in focal peroneal neuropathy. With high resolution ultrasound, the main landmark is the fibular head of course. At this level, the common peroneal nerve can easily be obtained [95]. Tracing the

nerve from this point proximally, it is first located below the short head of the biceps femoris muscle, and in front of it the lateral sural cutaneous nerve appears. More proximal, the common peroneal nerve runs medially and reaches the bifurcation of the sciatic nerve at variable depth and height. However, tracing the common peroneal nerve distally from the level at the fibular head, in our experience, the fascicles of the deep branch are located more distal, whereas the fascicles of the superficial branch can be found more proximal. The deep branch runs further deep below the peroneus longus muscle, and there it approaches the anterior tibial artery. Its terminal branches are difficult to detect within the surrounding muscles. The superficial branch first runs deep between the peroneus longus and extensor digitorum muscles. At the distal third of the lower leg (high variability!) the superficial peroneal nerve returns to the surface, pierces the crural fascia and now runs between the fascia of the two above-mentioned muscles and the subcutaneous fat-layer. Finally it reaches the back of the foot, where the nerve divides into its two terminal branches (high variability) [95]. The normal cross sectional area of the common peroneal nerve at the level of the fibular head is 0.10 ± 0.04 cm^2 [95]. Recently a cut-off value for peroneal neuropathy >8 mm^2 has been described [97].

In an epidemiological study, which involved our own patients ($n = 2{,}071$), the amount of subjects with a focal peroneal neuropathy was 2.8 % involving either the common peroneal nerve or its deep branch, and 0.33 % involving the superficial branch, respectively. Most frequently (67 %) the common peroneal nerve and its deep branch were affected by a traumatic nerve injury. High resolution ultrasound showed the typical signs already described in Chap. 6. Non-traumatic etiology included peripheral tumors of the peripheral nerve sheath, intra-neural ganglion cysts of the peripheral nerve sheath, compression by extraneural ganglion cysts, and compression below the peroneus longus muscle/fascia. Each of

these lesions can be detected reliably using high resolution ultrasound (Figs. 6.12, 9.11, 9.12 and 9.13). Similar results are to be found in the literature [98–101]. Unfortunately, skin incisions resulting from different surgical interventions (varicose vein extraction, osteosynthesis, emergency compartment syndrome treatment, etc.) frequently painfully affect superficial peroneal nerve branches at the distal third of the lower leg. High resolution ultrasound enables to detect the compression by scar tissue or the stump neuroma/neuroma in continuity, and furthermore, an ultrasound guided diagnostic/therapeutic block can again be done [102]. Some examples are shown in Figs. 9.14, 9.15, and 9.16.

However, the 3 T-MR-neurography can also assess the common peroneal nerve and its branches as well as the muscles supplied by them. This kind of imaging can be of advantage in obese individuals, where sonography is inferior to MRI. Furthermore, cases where differential diagnosis between

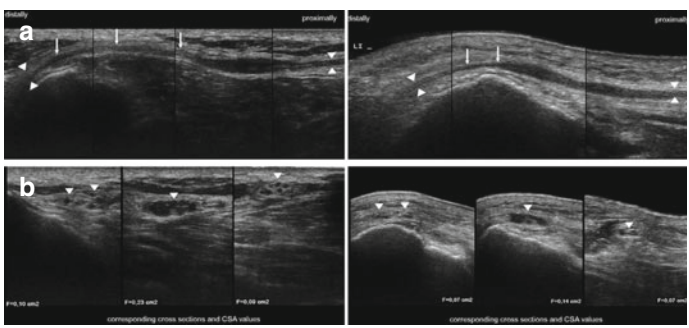


Fig. 9.11 (**a**, **b**) HRUS, two cases of peroneal nerve (*arrowheads*) compression at the fibular head (*arrows*) in a patient working as a tiler (**a**) and after leg crossing (**b**). Note the hypoechoic swelling of the fascicles just above the site of compression

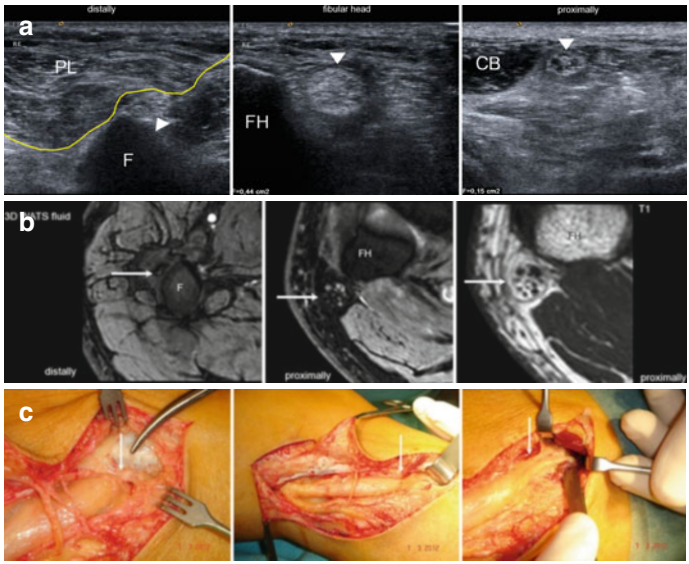


Fig. 9.12 Chronic entrapment of the peroneal nerve below the peronaeus longus muscle in a patient with food drop lasting more than 9 months. (**a**) Ultrasound cross sections show a normal nerve (*arrowhead*) at level of the short head of the biceps femoris muscle (*CB*). At the fibular head (*FH*), a massive nerve swelling (*arrowhead*) appears. The *left* image demonstrates the compression of the peroneal nerve (*arrowhead*) between fibula (*F*) and peronaeus longus muscle (*PL*). (**b**) Corresponding MRN images showing increased nerve size on T1 weighted images at the fibular head (*FH*) as well as hyperintensity of single fascicles on 3D WATS (fluid) images (*arrow*). More distally, the site of compression is demonstrated between the tendon of peronaeus longus muscle (*arrow*) and fibula (*F*) (MRN courtesy of Karsten Stock, MD (department of radiology and neuroradiology, Dessau-Rosslau Hospital/Germany)). (**c**) Corresponding intraoperative images before (*left*) and after (*right*) decompression (*arrows*). After surgery symptoms disappeared within 3 months (Images courtesy of Werner Schneider, MD (department of hand and plastic surgery, Dessau Rosslau Hospital/Germany))

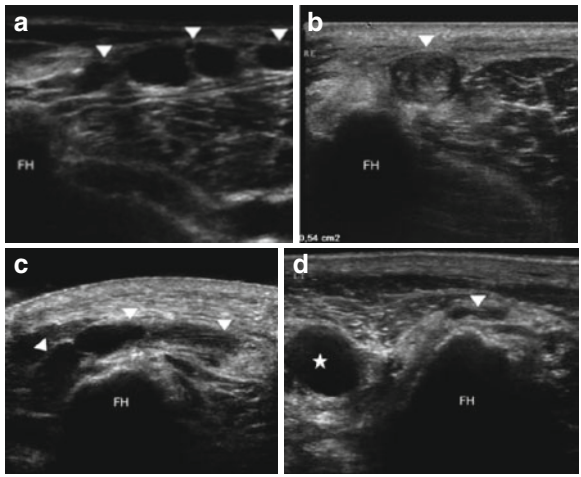


Fig. 9.13 HRUS, different peroneal nerve (*arrowheads*) pathologies at the fibular head (*FH*). (**a**) Multiple neurofibroma. (**b**) Neuroma in continuity. (**c**) Intraneural ganglion of the nerve sheath. (**d**) Extraneural ganglion (*asterisk*)

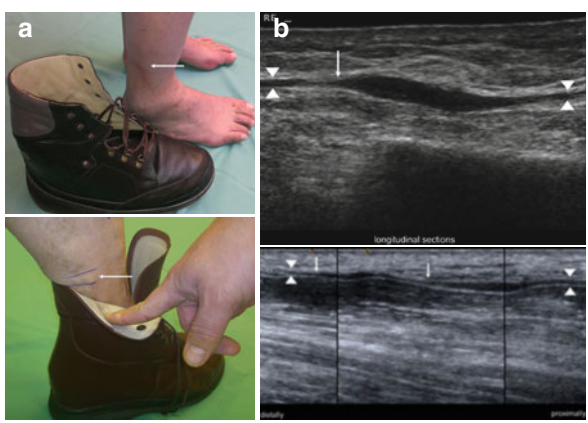


Fig. 9.14 Two cases of superficial peroneal nerve compression due to footwear. (**a**) Clinical findings. *Arrow* points on the site of compression. (**b**) HRUS, hypo-echoic swelling of the superficial peroneal nerve (*arrowheads*) just above the site of compression (*arrows*)

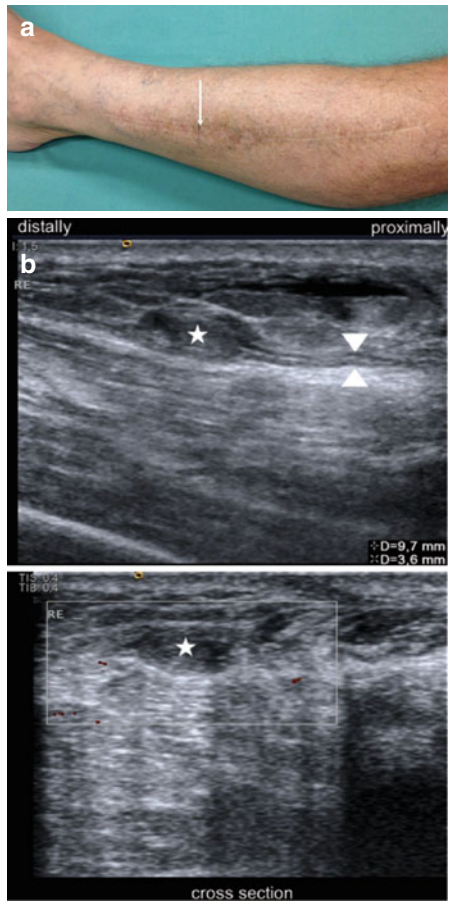


Fig. 9.15 Patient with stump neuroma of the superficial peroneal nerve after compartment syndrome surgery. (**a**) *Arrow* points on the neuroma in the course of the scar. (**b**) Neuroma (*asterisk*) at the end of the proximally located nerve stump of the superficial peroneal nerve (*arrowheads*) in HRUS

different neoplasia manifestations or intraneural ganglion cysts (refer to Sects. 6.1.7 and 6.2.7 and Chap. 13) is required, and where the administration of contrast agent is needed, profit from MRI more than from ultrasound imaging [34, 103, 104].

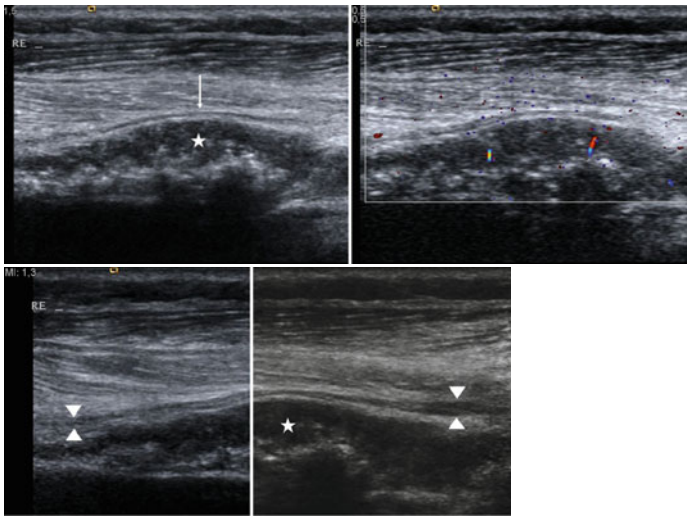


Fig. 9.16 Patient with history of prostatic cancer presenting an osteoplastic metastasis of the fibula (*asterisk*), in HRUS, tumor compression of the superficial peroneal nerve (*arrowheads*). *Arrow*: inguinal ligament

Please also refer to Figs. 6.19 and 9.12b. Not to be neglected are the diagnostic difficulties due to the "magic angle artefact" that can mimic a mild T2-lesion in MRI, a phenomenon that has already been described in Sect. 6.2.3).

9.8.6 Treatment

Surgery of trauma-related peroneal nerve lesions is done in the prone position as microsurgery in case of a tumor mass. Surgery of focal entrapment only can easily be fulfilled in supine position, with the body turned 30° to contralateral, knee joint about 90° angulated.

We prefer an incision using the skin crease of the knee joint, leading to the lateral aspect of the popliteal fossa, and finally turning around the prominent tibio-fibular joint into a distal direction. By doing so, a triangular skin flap is lifted up from popliteal fossa fascia and fascia over the peroneal compartment. The flap allows a final skin closure which keeps the nerve far away from overlying scar tissue. At first, the fascia of the popliteal fossa is gently opened parallel to the expected nerve course, whereby we have to be aware that the nerve here is located quite superficially and very closely connected to the fascia. With the identified nerve in view, further dissection leads around the fibular bone and finally to the most delicate point where superficial and deep branches enter the peroneal compartment. The common nerve trunk has to be lifted a little; both branches are separated from each other. Then, as a final step, the muscle fascia over the peroneal compartment has to be incised far enough so that both nerve branches enter the muscle compartment free of any more trapping structure. Sometimes a vertically downward running septum between muscle bellies can also be incised. Of course, a nerve transposition which would be desirable to prevent ongoing compression between fibula and skin is impossible. Finally, all incised fascias remain unclosed, the triangle skin flap is turned back, and skin stitches have to be good enough to tolerate flexion and extension movements of the knee joint when the patient gets up. A suction drain is mandatory because venous bleeding can occur when the patient gets up into vertical position. We have observed a few times re-bleeding 24 h postoperatively after mobilization leading to deterioration of the nerve function (Fig. 9.17a, b).

Surgery of space-occupying lesions is better done in the prone position because this facilitates the surgeon's situation, especially if magnification of the operating field by microscope is required. With the patient supine one would create a situation comparable to acoustic-neurinoma surgery with semi-sitting patients.

Ganglia beside the tibio-fibular joint as the most common tumor-like mass can occur relatively frequently as mentioned above. It is entirely thanks to the work of Spinner that we understand that these ganglia always have a connection to the

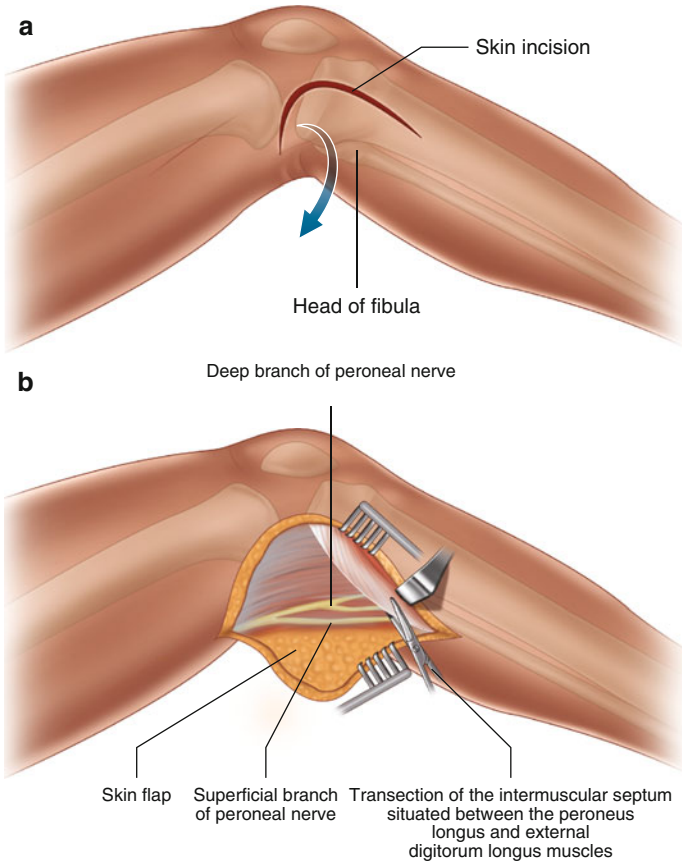


Fig. 9.17 Treatment of peroneal nerve entrapment at the head of fibula. (**a**) Limb positioning and skin incision. (**b**) Release of the deep nerve branch by transection of several intermuscular septa between the peroneus longus and extensor digitorum longus muscles

joint from which they originate [105]. The surgeon should therefore address these hidden connections and ligate them [106]. Nevertheless, nobody can guarantee that these cases will not recur. Everybody can imagine the importance and amount of pre-operative information that patients now need in order to understand the prognoses of their following treatments. Just as important seems preoperative imaging to reveal such comparably difficult lesions, although MRI findings not always succeed in describing how and where to find the small joint connection. It is very difficult for a radiologist to recognize them [107]. However, careful surgery that first skeletonises a nerve and then dissects around the ganglion cyst creates a situation which should allow the revelation of such a joint connection. Intraneural multi-cystic ganglion-like lesions which can also occur require magnification and real microsurgery which we will describe in Chap. 13 in detail. It is then much more difficult or even impossible to identify and ligate a joint communication. The same necessity of microsurgery holds true when imaging reveals a nerve sheath tumor in or below the popliteal fossa.

The question of how to treat injured and sometimes extremely painful terminal branches of the superficial peroneal nerve surgically remains unanswered. Perhaps, in future, experience and high resolution of ultrasound will improve in such a way that differentiation between extraneural compression by scar tissue and intraneural neuroma-formation becomes reliably possible. We must be enabled to rely on preoperative ultrasound findings, because surgical neurolysis when initiated can only succeed in the first kind of slight nerve injuries, and mostly not in one of the latter kind of nerve damage as described in Sects. 3.2 and 4.2. We need to remember the small caliber of superficial nerve branches which does not allow microsurgery, and if tried nevertheless, easily ends as duplicated neuropathic syndrome. However, sonographic imaging of superficial nerve branch lesions will gain greatly in value in making pain authentic, e.g. in the eyes of insurance companies. The prediction of eventual

surgery outcome will furthermore depend on capabilities to visualize intraneural details.

9.9 Deep Peroneal Branch at the Ankle

9.9.1 Anatomy

The final course of the deep peroneal nerve branch runs alongside the anterior tibial artery between tendons of the extensor hallucis longus and tibialis anterior muscles. At ankle level, nerve and artery are covered by the retinaculum extensorum which can be divided into a superior and an inferior one. They serve as hypomochleon of the tendons. A terminal motor branch arises under the inferior retinaculum, turns to lateral and supplies the extensor hallucis brevis and extensor digitorum brevis muscles. A terminal sensory branch represents a small inter-digital area between the first and second toes, a situation that is comparable to the sensory distribution of the posterior interosseus nerve at the dorsal surface of the hand (see Sect. 8.10.1). A focal and painful entrapment rarely occurs under the retinaculum, and since 1963 it has been termed “anterior tarsal tunnel syndrome” [108].

9.9.2 Damaging Factors

The nerve crosses the anterior ankle beneath the extensor retinaculum, a preformed situation comparable to the carpal tunnel. Tight shoes, sprains, and bone dislocations after ankle fractures may narrow the canal and thus cause a focal nerve compression [109].

9.9.3 Clinical Symptoms

Impairment of the terminal motor branch usually remains unnoticed, but it can eventually be helpful to find the diagnosis by electromyographic testing. In contrast to the motor branch, involvement of the sensory branch often causes rather painful paresthesias which radiate into the first interdigital space. An electric current-like pain component can then be triggered either typically under the retinaculum extensorum, or, quite rarely, at the level where the branch passes under the tendon of the extensor hallucis muscle. The trigger point leads to the diagnosis.

9.9.4 Treatment

Before anaesthesia, the trigger point is marked in the operating theatre. A small skin incision is done longitudinally and lateral to the tibialis anterior tendon which can easily be palpated through the skin. Special care has to be taken immediately with branches of the superficial peroneal nerve running to the medial aspect of the dorsal foot surface. They are situated subcutaneously so that injury to these branches can again cause a pain syndrome comparable to injury of the skin branches of the superficial radial nerve at the wrist (see Sect. 9.8.6). Palpation of the anterior tibial artery between the long extensor tendons leads to the small anterior tarsal tunnel. The entrance of this tunnel is covered by a fascia which has to be opened very carefully, carefully because injury of the artery will produce a questionable result. The dissection then leads to distal and finally aims at transection of one or both extensor retinacula. Palpation with your the tip of your fifth finger can exclude additional compression sites either proximal or distal. Previous literature has even reported ganglia lying between the tibialis anterior and extensor digitorum longus tendons as a possible compression factor [110].

9.10 Tibial Nerve Below the Popliteal Fossa ("Soleal Sling")

9.10.1 Anatomy

While the tibial nerve is most commonly entrapped in the tarsal tunnel (see Sect. 9.11), more proximal entrapment sites are increasingly appreciated as a potential cause of focal tibial neuropathy. Mastalgia et al. first described a tibial nerve entrapment by the tendinous arch of the soleus muscle origin in 1981, referred to as the soleal sling [111]. The fascial arch spanning its bilateral origin from the posterior aspect of the tibia and fibula forms a sling constituting a roof over the tibial neurovascular bundle which continues into a tunnel between the superficial and deep posterior compartments containing the tibialis posterior, flexor hallucis, and flexor digitorum longus muscles [112].

9.10.2 Clinical Symptoms

Symptoms of tibial nerve entrapment under the soleal sling include popliteal fossa and proximal calf pain, weakness of toe flexion, and sensory deficits in the foot sole that are aggravated by walking [113].

9.10.3 Imaging

High-resolution ultrasound experiences in depicting this rare focal neuropathy have not existed until now but will surely arise soon. However, it is capable of demonstrating other lesions of the tibial nerve (Figs. 9.18 and 6.25a). Incidentally,

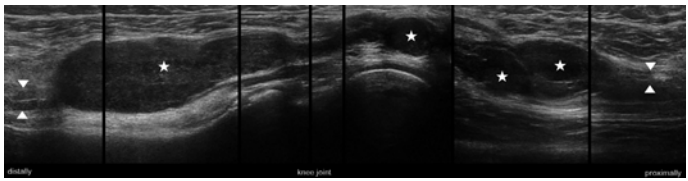


Fig. 9.18 Tibial nerve at the knee joint (*arrowheads*) affected by segmental neurofibromatosis (*asterisk*) in HRUS

the last figure is remarkable insofar as we have been once more confronted with an unexpected manifestation of a Parsonage-Turner syndrome which we describe in Sects. 6.2.8 and 10.3 in general. While radiology literature regarding the MRN appearance of this particular neuropathy is also sparse, our own experience after all consists of the fact that direct visualization of an abnormally T2 hyperintense and flattened tibial nerve at the soleal sling with proximal nerve enlargement is well possible by means of 3 T MRI and dedicated MRN sequences (Fig. 6.25b). Secondary signs of gastrocnemius and soleus muscle denervation oedema can serve as additional important findings.

9.10.4 Treatment

Patients with soleal sling entrapment neuropathy can of course benefit from soleal sling release [114]. We do not ignore our own surgical experiences in nerve decompression but the surgical approach below the popliteal fossa should be quite easy. We would try to avoid surgery as outpatient management for the same reasons discussed in Sect. 9.8, although insurance companies tend to refuse hospitalization of cases needing a simple decompression.

9.11 Tibial Nerve at the Ankle

9.11.1 Anatomy

A second preformed narrow channel at ankle level is located behind the medial malleolus. It is therefore named the "posterior or medial tarsal tunnel" [115]; its roof is formed by one or two flexor retinacula. A few centimeters above the medial malleolus, the tibial nerve starts to divide into three branches, the medial and lateral plantar nerves and a small branch to the medial aspect of the heel. The latter can penetrate the flexor retinaculum [116]. Not to be neglected, and surgically important, is the fact that the posterior tibial artery runs along the nerve branches through the tunnel. When entering the sole of the foot, the artery divides into two main branches which supply all soft tissue structures of the sole. In the case of any surgery, preservation of both arterial branches is mandatory, comparable with the necessity of preserving the ulnar artery within the Guyon's canal. The region under the flexor retinaculum contains not only the terminal tibial nerve branches and arteries just mentioned, but also, and within a separated compartment, both tendons of the flexor digitorum longus and tibialis posterior muscles.

If we try to compare the anatomy of arteries and nerves between hand and foot, we have to bear in mind that the posterior tarsal tunnel contains both structures of the carpal tunnel and Guyon's canal. The medial plantar nerve branch is functionally comparable to the median nerve, the lateral plantar nerve branch to the ulnar nerve, and the posterior tibial artery to the ulnar artery. It is well known that preservation of all these structures is a mandatory precondition when the tarsal tunnel is entered surgically.

9.11.2 Damaging Factors

In 1978, Steward listed a large number of factors that cause functional impairment of the tibial nerve and its branches, such as posttraumatic fibrosis, tendon sheath cysts, tenosynovitis, rheumatoid arthritis, and acromegalia, and extraneural and intraneural ganglia [117]. Multiple intraneural ganglion cysts of the tibial nerve are a particular challenge to microsurgical ability [118]. Soft tissue alterations related to hypothyroidism have been reported in literature to be associated with multiple entrapments [119]. We observed two such patients with bilateral tarsal tunnel syndromes. Keck described findings like a varicocele in the tunnel in 1962 [120]. However, on the other hand, so-called idiopathic cases of tarsal tunnel syndrome can occur as unilateral and even bilateral disorders comparable to most carpal tunnel syndromes [121]. They have the best prognosis whereas ankle distortion-related cases likely develop intraneural fibrosis so that they often can no longer improve despite a careful external nerve decompression. Because of the large variability of damaging factors at this location, preoperative imaging has already been recommended in the early periods of imaging development [122].

9.11.3 Clinical Symptoms

It cannot be mentioned often enough that the tarsal tunnel comparably contains structures of carpal tunnel and Guyon's canal together. For this reason, the tarsal tunnel syndrome looks similar to a combination of entrapments of the median and ulnar nerves at the wrist. Therefore, the associated symptoms are not so clearly distinguishable. All muscles of the sole

are involved as well as the whole skin of the plantar surface. Patients commonly suffer from severe pain over the whole plantar skin, whereas weakness or muscle atrophy within the sole remains unremarkable in the patient's eyes. We have no reliable explanation as to why a tibial nerve entrapment is almost always associated with severe pain, whereas, for instance, the common peroneal nerve entrapment at the knee is always associated with weakness and without pain, although both nerves are mixed ones [123]. In summary, the main disturbing symptoms are paresthesias, tingling, and pain restricted to the sole, and rarely pain that radiates back into the dorsal lower leg. Some patients observed deterioration after long walking tours. The physician should ask for a Hashimoto thyreoiditis in the patient's history because it can promote multiple entrapments as just mentioned [119]. Although spontaneous onsets and slow deteriorations are reported in the majority of cases, severe entrapments can also be related to distortions involving the ankle joint. Pain then starts early subsequent to the distortion and increases more rapidly. The posttraumatic soft tissue reaction results in extraneural and intraneural fibrosis so that even one of the neuropathic pain types can occur. These trauma-associated cases need early surgical decompression if ever possible to prevent the patient from lifelong neuropathic pain. Unfortunately, trauma itself and the necessity for tight casts over a period of time cause situations where it seems impossible to differentiate pain type and location. Consequently, the final diagnosis of a posttraumatic tarsal tunnel syndrome is often considered much later.

Very rarely, cases can occur which present with either lateral plantar branch compression or medial plantar nerve compression only. The plantar nerves may normally be damaged within the tarsal tunnel as just described, but much more rarely, more distally in their course, where they enter the sole and pierce the fascia of the abductor hallucis muscle. This fascia forms an

arcade comparable to that of the supinator muscle of the forearm. At this site, isolated involvements of one of both plantar branches can be observed with clinical symptoms like a carpal tunnel or Guyon' canal syndrome. Another variant of symptoms is an entrapment of the first sensory branch of the lateral plantar nerve only, named Baxter's neuritis. Pain is here restricted to the medial heel.

9.11.4 Electrodiagnostics

Concerning electrodiagnostic testing of the tarsal tunnel syndrome (TTS), one would expect a prolongation of the distal latency and a delayed or absent sensory response of the plantar branches (see Sects. 5.3.1 and 5.3.2). Reliable electrodiagnostic testing is nevertheless very difficult due to the high percentage of asymptomatic subjects who yet have abnormal sensory and motor results. In a study from 1980, 33 % of asymptomatic subjects older than 55 years presented with absent medial plantar sensory conduction, and 50 % of individuals had electromyography evidence of denervation in intrinsic muscles [124]. In a systematic evidence-based review of electrodiagnostic evaluation of tarsal tunnel syndrome cases, only four papers met criteria required to meet class III level of evidence. It was concluded that nerve conduction studies may somehow be useful confirming the diagnosis. Sensory NCSs turned out as more likely to be abnormal than motor NCSs, but the actual sensitivity and specificity could not be determined. The sensitivity of needle EMG abnormalities could not finally be determined [125]. In a recent review of the literature, the continuing controversial role of electrodiagnostic techniques in TTS was again assessed. As a conclusion, the role of NCS remains controversial insofar as the inability to predict which cases will respond to decompression [126].

9.11.5 Imaging

Ultrasound is a useful examination tool because it can be performed rapidly, and the tibial nerve and its main branches (medial and lateral plantar) can be visualized on transverse and longitudinal images. Ultrasound is reliable in measuring the cross-sectional area of the tibial nerve. Despite possible small measurement errors, a side-to-side difference of approximately 1.8 mm can be interpreted as meaningful in an individual patient [127]. In a recent report on 17 cases with tarsal tunnel syndrome, the preoperative sonography diagnosis was said to be intraoperatively confirmed in all cases. The neuropathy origins were ganglia ($n=10$), talocalcaneal coalition ($n=1$), talocalcaneal coalition associated with ganglia ($n=3$) and varicose veins ($n=3$). Among the cases involving ganglia, three ganglia were not clearly palpable before surgery, and they were revealed as small. Among the cases involving talocalcaneal coalition, ultrasonography indicated a beak-shaped bony process on the short axis images [128]. Ultrasound can provide direct evidence of nerve compression, because it then demonstrates focal nerve enlargement and changes in nerve echogenicity. A synovial cyst attached to the flexor hallucis tendon can play a role [129]. Hypertrophy of the abductor hallucis muscle is one of the reported further potential compression causes affecting either one or both tibial nerve branches. In a cadaveric study, nerve compression due to abductor hallucis muscle hypertrophy was evaluated by sonography. If ultrasonographic estimation of dorso-plantar thickness is >12.8 mm and medio-lateral width >30.66 mm, an abductor hallucis muscle hypertrophy associated compression neuropathy is said to be possibly suspected [130]. Some examples are shown in Fig. 9.19.

Magnetic resonance (MR) imaging, including high-resolution MR neurography, allows detailed evaluation of the course and

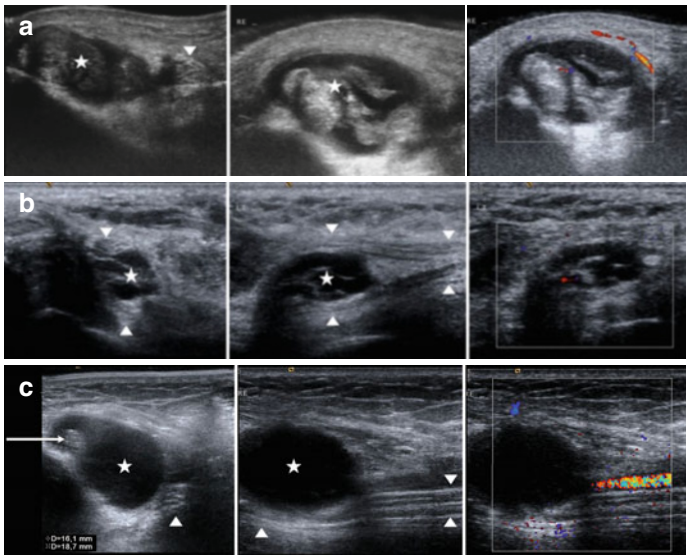


Fig. 9.19 Tarsal tunnel syndrome caused by different pathologies in HRUS imaging. (**a**) Tendinopathy both of the tibialis posterior and flexor halluces tendons (*asterisk*) displacing the tibial nerve (*arrowhead*). (**b**) Multi-cystic ganglion (*asterisk*) between the two plantar branches of the tibial nerve (*arrowheads*). (**c**) Ganglion (*asterisk*) of the tendon sheath of the flexor hallucis tendon (*arrow*) compressing the tibial nerve (*arrowheads*)

morphology of peripheral nerves, as well as accurate delineation of surrounding soft tissue and osseous structures that may contribute to tarsal tunnel narrowing (Fig. 9.20). Even the medial calcaneal nerve can be seen on axial images and, less commonly, on sagittal images as it separates from the tibial nerve proximal to the flexor retinaculum and continues toward its subcutaneous position along the medial aspect of the hindfoot. The medial and lateral plantar nerve branches are optimally to be visualized on axial images. Additionally, the inferior

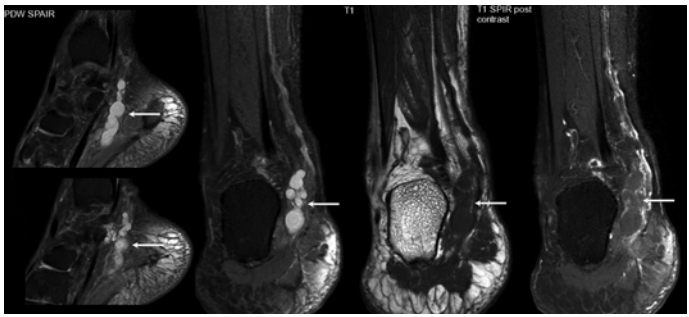


Fig. 9.20 MRN of a multi-cystic ganglion (*arrow*) within the tarsal tunnel. Note the slight contrast uptake at the borders of the cyst. (MRN courtesy of Karsten Stock, MD (department of radiology and neuroradiology, DessauRosslau Hospital/Germany))

calcaneal nerve can often be followed as it takes off from the lateral plantar nerve and dives toward the abductor digiti minimi muscle [130]. The tibial nerve and its branches may be compressed in the tarsal tunnel owing to trauma (fracture, surgery and scarring), space-occupying lesions (tumor, ganglia, varicosities and anomalous muscles) and foot deformities (hind-foot valgus and – less typically – hind-foot varus, with fore-foot pronation, pes planus and tarsal coalition). However, in up to 40 % of patients, an underlying cause for the tarsal tunnel syndrome is not yet identified [130]. Nevertheless, accessory muscles within the tunnel (e.g. a supplementary flexor digitorum longus muscle) can be found.

Direct evidence of trauma-related tibial nerve damage may be difficult to detect due to the small nerve caliber, but increased signal intensity, increased size and nerve course deviation may occasionally be depicted. As tarsal tunnel entrapment usually manifests rather than motor deficits, muscle denervation is still rather later to

be sensed, but when present, muscle atrophy is visible more distally at the mid- or fore-foot [130].

By means of MR-neurography (MRN), isolated mild T2 hyperintensity is frequently observed during routine evaluation, likely reflecting subclinical traction/friction neuritis. Therefore, in the literature, further signs of neuropathy such as enlarged nerve fascicles, encasing perineural fibrosis and potential denervation muscle changes should also be assessed before making diagnosis of tarsal tunnel syndrome [131].

MRN may be used in a post-operative setting to evaluate the etiology of failed surgical decompression due to incomplete decompression or encasing perineural fibrosis, or even neuroma-in-continuity formation if trauma associated, and possible persistent muscle denervation changes when compared with pre-operative settings [131]

9.11.6 Treatment

Surgical treatment has to aim at:

1. Pain relief by adequate nerve decompression – widely known facts since 1962 [120, 121]
2. Permanent pain relief by preservation of sliding soft tissue structures in a very delicate area [132]

The latter goal is particularly difficult to achieve in a region which is extremely moved and loaded by each step. After previous negative experiences, we refuse outpatient surgical procedures, hospitalize the patient for 4–5 days, and instruct them not to unroll the operated foot for 2–3 days. These precautions prevent postoperative hematoma and reactive fibrosis which destroy sliding abilities within the tunnels.

Surgery to the posterior tarsal tunnel syndrome should only be used if pathological electrodiagnostic testing can prove the indication. The differential diagnosis for similar diseases such as plantar fasciitis only succeeds by such means. In our experience, spontaneous occurring symptoms with pathological tests have the best prognosis, whereas traumatic traction damage to the tibial nerve has a rather controversial or even bad prognosis. Cases with a neuropathic pain type in the sole, particularly after trauma, need to be discussed thoroughly preoperatively if surgical decompression or even microsurgical neurolysis is considered.

The patient may lie supine or prone; when supine, he should be turned 30° so that the region behind the malleolus comes within easy reach. The skin incision is slightly curved behind the medial malleolus; it starts about 3 cm above the malleolus in order to enable to identify artery and nerve before entering the tarsal tunnel. Palpation of the arterial pulse is helpful and indicates where the lower leg fascia has to be opened longitudinally. We remain parallel to the course of artery and nerve. When afterwards proceeding downward, the surgeon is always prevented from entering into the wrong compartment; the hypomochleon of the posterior tibial and flexor digitorum longus tendons has to be preserved. The transection of the flexor retinaculum still follows, always with the nerve or its branches, artery and its branches, and sometimes tortuous venous vessels in view. An enlarged venous plexus should be considered as the source of re-bleeding when the patient gets up. The tibial artery often shows a serpentine course, and its branches intertwine with the plantar nerve branches. Small branching vessels should be tied with bipolar forceps, keeping far from the nerves, and then transected before being disrupted. The transection of the flexor retinaculum has, of course, to be complete. Medial and lateral plantar

nerve branches now pierce a fascia which covers the abductor hallucis muscle. The entrance into the sole compartment should be dissected for both nerve branches and opened enough so that ongoing compression at these sites is excluded. This area is particularly delicate insofar as the tibial artery divides there into the medial and lateral plantar arteries to form the plantar vessel arch, comparable to the hand anatomy. Rare isolated compression syndromes of either the medial or the lateral plantar nerve branch have their origin at this far distal localization, and not under the flexor retinaculum. The surgeon should therefore bear in mind that the relief does not always succeed after retinaculum transection only. This important part of the decompression just described takes place far distal and needs to extend the skin incision slightly curved under the medial malleolus ahead and never into the plantar surface. We should avoid any skin incision which is later loaded by each step.

The surgical exposure needs time and therefore requires general or spinal anaesthesia. The surgeon has to be alert to expose unusual findings such as dilated veins, unusual tenosynovitis of tendons which pass through the tarsal tunnel and hypertrophy of the abductor hallucis muscle [116]. A particular challenge arises from intraneural or extraneural ganglia which have occult communications to neighboured joints [118]. The surgeon has to manage these lesions, especially the intraneural ones, if he is unexpectedly confronted with these not quite rare lesions. In our experience, it is seldom that the surgeon succeeds in identifying a joint communication in order to avoid recurrence. Therefore, a recurrence of extra or intraneural ganglia cysts have to be borne in mind (see Sect. 13.3.8) [118]. On the other hand, preoperative high resolution ultrasound imaging of the tarsal tunnel region will gain more and more importance as described above. It is well known that

local anaesthesia is completely inadequate to manage all the previously mentioned possible findings. At the end of the operation we avoid subcutaneous stitches because they can strangle the tibial nerve. A drain, with or without suction, is inserted for 2 days, and the skin stitches remain for 14 days unconditionally. The need to avoid loading the foot by the patient's body weight for a few days has already been mentioned above (Fig. 9.21).

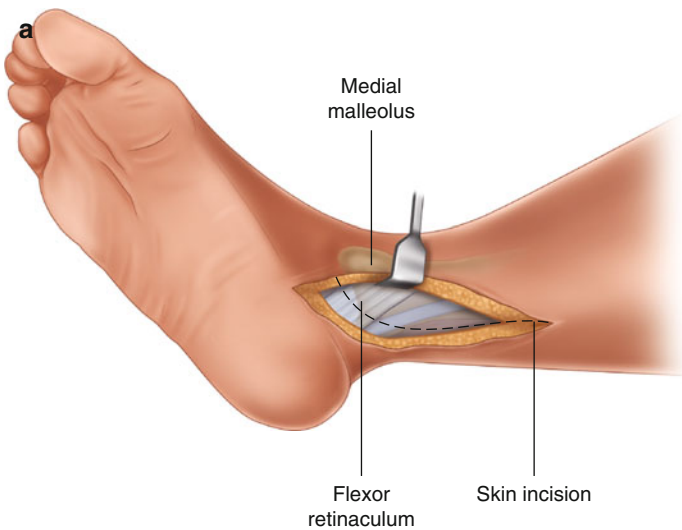


Fig. 9.21 Treatment of the tarsal tunnel syndrome. (**a**) Positioning of the lower leg and skin incision. (**b**) Exposure; concomitant vessels and the calcaneal nerve branch have to be preserved; transection of the flexor retinaculum as the first step. (**c**) Release of both nerve branches at the entrance into the sole of the foot as the second step

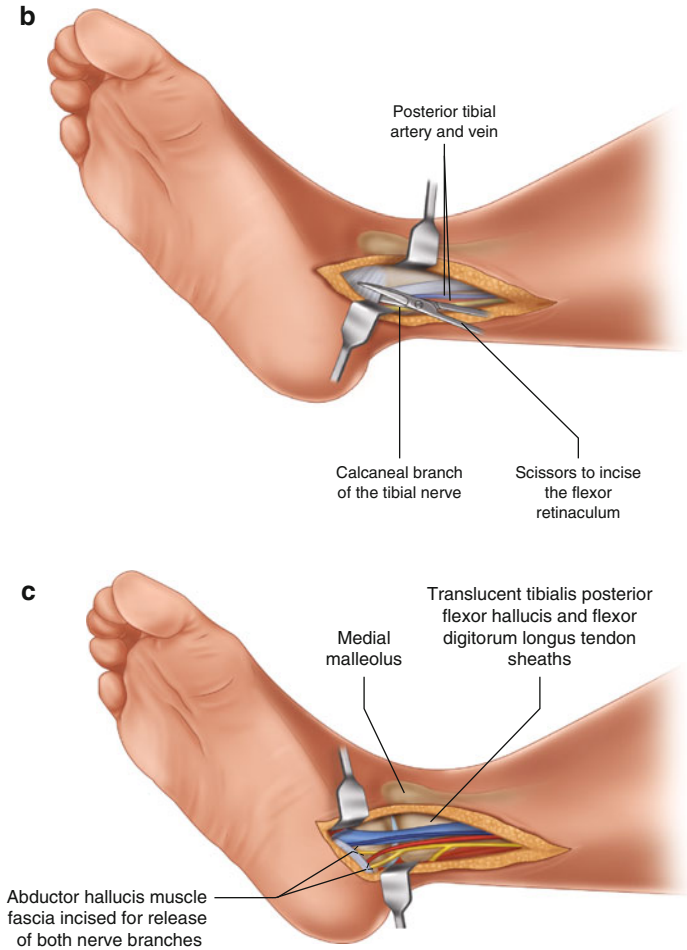


Fig. 9.21 (continued)

9.12 Plantar Digital Nerve Entrapment

9.12.1 Anatomy

Four common plantar digital nerves run between the five metatarsal bones and divide into two individual branches about 0.5 cm before the metatarso-phalangeal joints. These joints are connected with each other by inter-metatarsal ligaments. Joints or tendon sheaths can develop bursa cysts. The space between joints, transverse ligaments and sometimes additional cysts is extremely small so that a very acutely noticed pain syndrome can occur as already described in 1876 by Morton [133].

9.12.2 Damaging Factors

It is well known that anatomy variations can cause slight repeated sharp compression of the small nerves. The role of an additional bursa cyst is controversial, but exposures revealed that nerve fibers and cyst membrane could not be separated from each other, a situation reminiscent of typical intraneural ganglia (see Sect. 13.3.8).

9.12.3 Clinical Symptoms

The compression causes electric current-like extensive pain normally radiating into the distribution of the affected common plantar digital nerve, the space between two contiguous toes. Mostly, the interdigital space between the third and fourth toes is involved but, in our experience, all the other interdigital spaces can alternatively or additionally be affected. The pain is characteristically provoked by walking or manually pressing the toes

against each other. This phenomenon serves as a clinical test to discover the diagnosis whereby the physician can add digital pressure from plantar [134]. Apparative diagnosis has again to be assumed controversial, because routine MR imaging mostly reveals diffuse signal alterations which probably indicate inflammation of a bursa or a tenosynovitis. Common imaging cannot reliably reveal a nerve neuroma in continuity as we likely would expect. We remember a case where a neurotomy was carried out through an interdigital space with such MRI findings, whereas the patient had affirmed pain within the neighbouring space; the neurotomy resulted in a neuropathic pain syndrome in the operated and in a remaining neuralgia in the neighbouring interdigital space. Our message is therefore at present to rely on the clinical presentation and on imaging findings not before they seem to provide security. The diagnosis consists, first, of a patient's report that the pain repeatedly radiates into a certain interdigital space, second, of positive effects by provoking maneuvers, and third, of pain disappearing after installation of local anaesthesia into the interdigital space for a period of 1–2 h. Recent reports have, however shown that high resolution ultrasound is able to detect Morton neuroma. Of typical appearance would be a hypoechoic mass within the intertarsal region that does not abut against adjacent bony structures and has either well-defined borders or is poorly marginated. A biconcave appearance of the mass ("ginkgo leaf sign") from compression by adjacent structures may be specific [135]. An example is shown in Fig. 9.22. Again, the future will answer questions as to how we can value HRUS findings in the interdigital foot space.

9.12.4 Treatment

We completely avoid a plantar approach to protect the patient from any plantar scar, and we additionally avoid a transection of

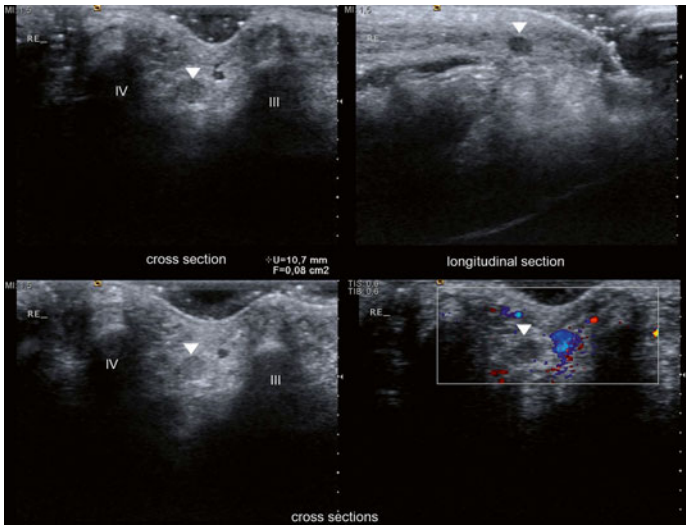


Fig. 9.22 Morton neuroma (*arrowhead*) between IV and III metatarsal bone in HRUS

the inter-metatarsal ligament to protect the patient from static alterations of the foot. Therefore, an approach from dorsal through the inter-metatarsal bones remains mandatory. It needs a microscope, a dissector and a blunt hook, small scissors, and a small suction device. We do not apply a neurolysis as sometimes recommended [136]. Instead we prefer a neurotomy because the surgeon cannot alter the narrow anatomical situation between two bony structures or perhaps enlarge the interosseous space. On the other hand, the neurotomy bears the risk of developing a new painful neuroma at the proximal nerve end. For this reason, the neuroma has to be located far proximal, within soft tissue with less risk of being newly irritated during walking.

The approach from the dorsal surface needs a 2- to 3-cm longitudinal incision. A small self holding retractor is helpful to

enter between the two metatarsal bones. A microscope for magnification should be added in any case. The approach between bulging muscle fibers into the deep plantar compartment mostly proved more difficult than expected. The assistant cannot help, the suction in your left hand helps to dissect, and the instruments in your right hand are scissors, dissector or hook. Try to preserve the inter-metatarsal artery because the management of arterial bleeding from its stumps between bulging muscle fibers is difficult. At the end, the blunt hook is needed to isolate the nerve, lift it up and transect it. Sometimes during this maneuver, small tendons of plantar muscles can mimic the nerve. Their colour is quite similar to that of the nerve because of the commen soft tissue fibrosis in the sole. If you have such a tendon under your hook, it comes out as less elastic than a nerve structure. Try to remove a small piece from one nerve end so that the pathologist can tell the patient that a neurotomy was really done.

The surgery can be done as an outpatient procedure, but then questions are raised every time on how to manage bandaging with a foot that should not be loaded for several days. A small drain which ends in the compressing bandage, without suction, prevents a postoperative hematoma.

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Chapter 10
Generalized or Multifocal Disorders Occasionally Mimicking Focal Neuropathies

Thomas Schelle

10.1 Introduction

The following chapter will focus on a couple of generalized or multifocal disorders affecting peripheral nerves, plexus and motor-neurons, which occasionally mimic focal neuropathies. Most of them are rather uncommon or rare. However, specialized knowledge will help to prevent patients undergoing surgical interventions or other therapies without benefit or even that they are harmed in a worst case scenario. Therefore, it is crucially important to always bear in mind the disorders described below and to perform – especially after failed surgery or unsuccessful treatment of a suspected focal neuropathy – an extensive differential-diagnostic approach. It would go beyond the scope of this book to go into every single detail. Instead, important facts are highlighted only so that physicians are helped to differentiate generalized respectively multifocal from focal neuropathies clinically. Furthermore the essential diagnostic pathways such as different imaging techniques and nerve conduction studies are explained.

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10.2 Motor Neuron Disease

Behind the term "motor neuron disease" different degenerative disorders affecting the upper and/or lower motor neuron can be assumed. We will focus on amyotrophic lateral sclerosis (ALS) which was originally described by the French physician Jean-Martin Charcot [1]. The highest incidence of this rare disease is reported in Finland (2.4/100.000 subjects/year), [2]. Due to increased life spans of the population as well as improved diagnosis, the incidence is said to be increasing with a peak between the sixth and seventh decades [3, 4]. Most cases are sporadic, and approximately 10 % of them are said to be hereditary due to mutation of the SOD1 gene [5]. ALS affects both upper and lower motor neurons. Degeneration of upper motor neurons leads to a spastic paralysis, whereas degeneration of lower motor neurons results in muscle weakness and wasting [6]. Early manifestation of symptoms usually begins focally. Approximately two-thirds of cases start in the lower limbs [7]. This fact explains why some patients are initially misdiagnosed as cervical myelopathy (spasticity and hyperreflexia in their lower limbs in conjunction with muscle atrophy and weakness in the upper limbs), or as ulnar neuropathy or carpal tunnel syndrome (weakness and atrophy of intrinsic hand-muscles) as to be seen for instance in Fig. 10.1. In fact, 5 % of patients suffering from a motor neuron disease had cervical or lumbar decompression surgeries early in their course of complaints [8]. In the majority of cases, ALS rapidly progresses within 2–5 years after diagnosis, and death results from ventilatory weakness [9]. Currently, there exists no further curative treatment. The following three clinical signs may help to differentiate from a focal neuropathy in an early stage of ALS:

- No sensory symptoms (numbness, neuropathic complaints etc.) since primarily only the motor-neurons are affected
- Widespread muscle twitching (fasciculations), even in clinically unaffected areas

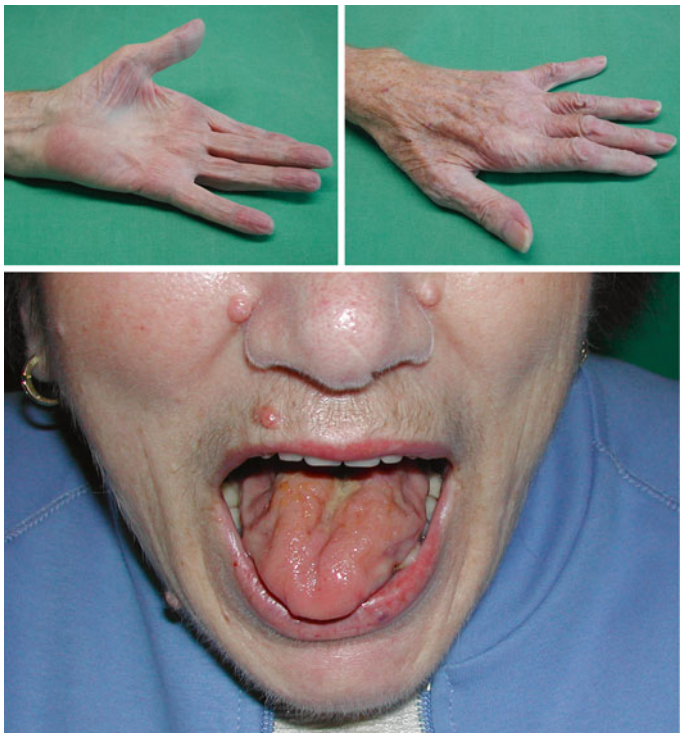

Fig. 10.1 Patient with amyotrophic lateral sclerosis. Note the focal beginning of the disease mimicking an ulnar neuropathy (*upper part*). Later on, most of the patients show signs of bulbar involvement (tongue atrophy)

- Combination of muscle wasting with hyperreflexia (combination of signs of lower and upper motor-neuron)

These symptom-combinations should raise doubts regarding the eventual diagnosis of a focal peripheral neuropathy, and they should create the need for further examinations [10]. Typical signs of ALS are then moderately elevated levels of serum creatine kinase on one hand, and evidence of widespread denervation and reinnervation (in thoracic paraspinal

muscles, muscles supplied by cranial nerves, etc.) in needle electromyography on the other. Sensory nerve conduction studies remain normal as well as motor nerve conduction velocity since the abnormality consists of axonal loss of motor fibers only [11, 12]. Signs of demyelination, especially abnormal temporal dispersion or conduction block, should instead raise serious doubts concerning diagnosis (see also multifocal motor neuropathy) [13]. Moreover, transcranial magnetic stimulation can show the involvement of the upper motor neuron [11].

Widespread fasciculations as well as muscle atrophies, additionally characterized by remodelling of muscle tissue with connective and fatty tissue, will lead to hyperechoic muscle appearance together with some decrease of muscle diameter in non-invasive high resolution ultrasound [14].

10.3 Diseases of Brachial and Lumbosacral Plexus

Among the different disorders of the brachial plexus there is one known as "neuralgic amyotrophy" that was first described in detail by Parsonage and Turner in 1957 [15]. We already mentioned this increasingly diagnosed disease in earlier sections, for example in Sect. 6.2.8. In addition to common idiopathic forms, a hereditary form is characterized by an autosomal-dominant trait. Recently, it has been reported that a mutation in the SEPT9 gene is detectable in several individuals suffering from such a hereditary neuralgic amyotrophy. The result is probably a myelin defect [16]. Etiology of the idiopathic form is unknown so far. Several mechanisms, especially of autoimmune origin, have been discussed [17]. Compared to motor neuron diseases, the incidence of 2–3/100.000/ individuals/year regarding the idiopathic form is rather low [18]. The typical clinical course, sometimes with attacks, consists of three stages [19]:

- Stage one is characterized by a sudden onset of severe aching or throbbing pain reaching a score between 8 and 9 on a

pain-scale ranging from 1 to 10. Typically the pain arises from the cervical spine, shoulder or scapular region and radiating down to the arm. This stage usually lasts for 4 weeks on average.

- In stage two mostly a motor deficit with a "patchy" distribution pattern in the territory of one or different peripheral nerves or nerve roots occurs, characterized by weakness, paralysis and muscle atrophy, outlasting the pain for a long time. In the majority of cases (71 %), the upper and middle brachial plexus with or without long thoracic (winged scapula) or suprascapular nerve are involved but nearly every nerve in the brachial plexus can be affected. Sensory symptoms are less clinical prominent.
- In the third stage – lasting 3 years or more – one-third of the patients experience a complete recovery, whereas in two-thirds the overall recovery is less favourable with persisting pain and residual paresis [19].

Attacks are more relapsing in patients with hereditary neuralgic amyotrophy (74.5 %) than with the idiopathic form (26.1 %). Antecedent events were monitored in approximately half of the cases, primarily infection, surgery and exercise [19].

In a smaller subgroup of patients neuralgic amyotrophy may predominantly involve the anterior interosseus nerve (see Sect. 8.13.1 and Fig. 6.24) or the radial nerve (see Sects. 8.9 and 8.10.1) as well as the lower brachial plexus (Fig. 6.25). This applies for women especially [19]. Here, the typical clinical course of attack-like onset helps to differentiate neuralgic amyotrophy from a true focal compressive neuropathy.

An important differential diagnosis of a painful brachial plexus lesion is infiltration by a malignant tumor/metastasis. This fact should always be borne in mind, particularly concerning patients with history of lung or breast cancer.

Laboratory tests and lumbar puncture often reveal things as normal and may help to exclude other conditions with similar symptoms (e.g. neuroborreliosis) [19]. Approximately 3 weeks after onset needle electromyography can demonstrate the extent and distribution of the axonal damage in 96 % of cases. In particular, subtle denervation in muscles not supplied by a single nerve

as well as in muscles of the contralateral side may help to differentiate from a focal neuropathy [17]. In 6 % of cases, by means of MR imaging, a T2 lesion or hypertrophy of the brachial plexus was described [19]. There are no actual reports about ultrasound imaging regarding the neuralgic amyotrophy. In our own experience, hypoechoic and long-segmental CSA enlargement of the affected roots, of parts of the brachial plexus, of peripheral nerves, or of even single fascicles can be detected weeks after onset of the disease but not at the beginning (Figs. 6.11, 6.24, 6.25, and 8.19). Pham et al. [20] found 20 cases of neuralgic amyotrophy mimicking an interosseus anterior syndrome a T2-lesion of the corresponding fascicle group within the median nerve at upper arm. It remains unclear at the moment whether these changes will be reversible. Perhaps there is also an affiliation with spontaneous torsional nerve neuropathy in some cases, as demonstrated in Fig. 8.11 [21]. Although neuralgic amyotrophy is a self-limiting condition, some supportive therapies may be helpful. Most important is adequate analgesia during the initial phase of the disease as well as physical therapy. Some very preliminary data on patients treated with corticosteroids suggest that the course of the symptoms might thus be favourably influenced [19].

10.4 Hereditary Neuropathies (CMT I, HNPP)

10.4.1 CMT – Charcot-Marie-Tooth Disease

CMT – also known under the term hereditary motor-sensory neuropathy (HMSN) – was first described by Charcot, Marie and Tooth in 1886. It affects approximately 1 in 2,500 individuals [22, 23]. It is represented by a heterogeneous group of different hereditary disorders whereby CMT Types Ia and IIa are to be found most frequently [24]. CMT Type Ia is associated with a duplication on chromosome 17 which contains the gene encoding peripheral myelin protein 22kD (PMP22) leading to a demyelinating

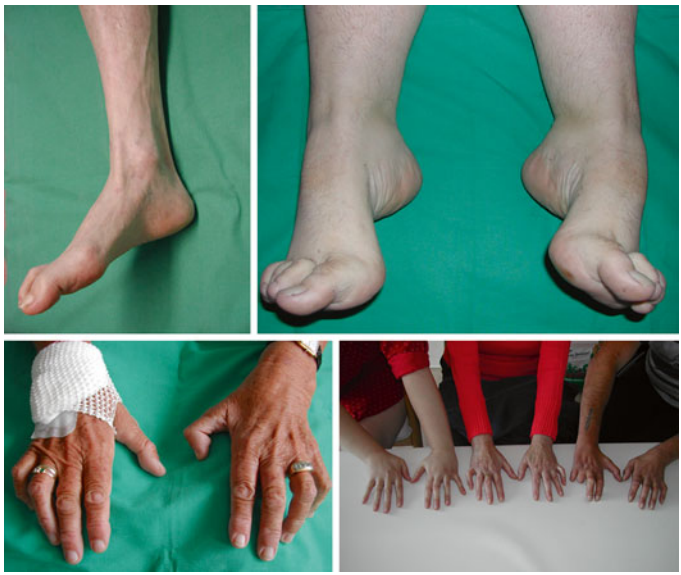

Fig. 10.2 Clinical presentation of hereditary motor-sensory neuropathy (HMSN). *Upper part*: Pes cavus and hammer toes. *Lower part*: HMSN mimicking an ulnar neuropathy. *Right image* shows an HMSN family (daughter, mother and brother of the mother from *left to right*)

neuropathy [25]. A mutation in the gene encoding kinesin, a protein that is involved in microtubule-mediated axonal transport, leads to CMT Type IIb, clinically represented by an axonal neuropathy [26]. Both CMT I and II are usually inherited as an autosomal dominant trait. Regarding further, mostly rare, types and subtypes please refer to specialist literature [27]. The classical phenotype of CMT affects individuals younger than 20 years of age and progresses slowly (Fig. 10.2). Muscle weakness/atrophy and sensory dysfunction usually starts distal-symmetric at the lower extremities, characterized by a foot drop/peroneal atrophy with steppage gait and a sock-shaped sensory impairment [28, 29]. In an advanced stage of the disease, distal and symmetric muscle weakness and atrophy of the intrinsic hand muscles (Fig. 10.3) as well as the not always glove-shaped sensory impairment may be

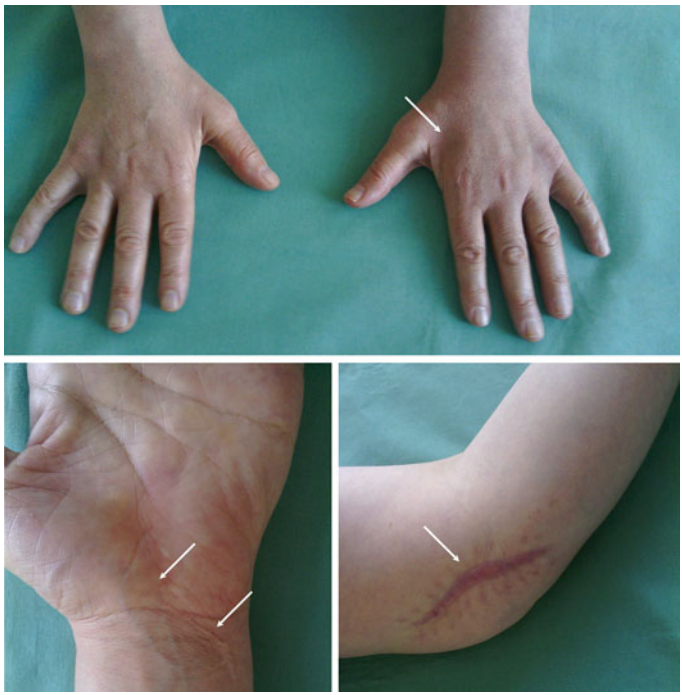

Fig. 10.3 HMSN mimicking multiple entrapment syndromes (carpal tunnel syndrome, Guyon's canal syndrome as well as ulnar neuropathy at the elbow). After surgical decompression at the left site (*arrows*) a worsening of the symptoms appeared

misdiagnosed as carpal tunnel syndrome or ulnar neuropathy at the elbow. Moreover, skeletal abnormalities such as pes cavus, hammer toes and scoliosis are very characteristic [28, 29]. Beyond this classical phenotype, some variants without risk of confusion with a focal neuropathy have been described: infantile and early onset forms are characterized by a mostly severe muscle weakness ("floppy infant"), severe sensory impairment, and they may end fatally [27].

Suspected diagnosis is established by the typical clinical signs mentioned above, a positive family history, electrodiagnostic

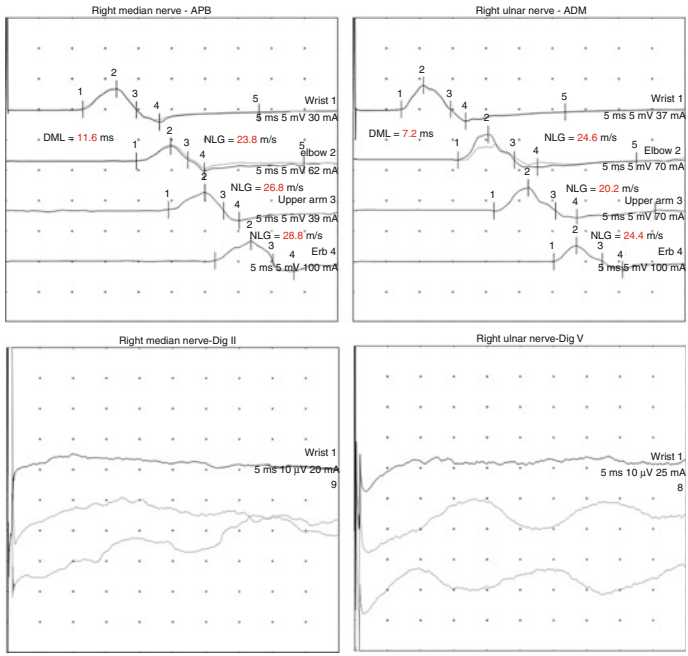

Fig. 10.4 Corresponding EDX. Note the uniform slowing of motor nerve conduction velocities as well as the prolonged distal motor latencies in the ulnar and median nerve suggesting a generalized demyelinating process. Sensory nerve action potentials could not be obtained. The same results revealed nerve conduction studies at the opposite site (not shown). At the lower extremities (not shown) both compound muscle action potentials and sensory nerve action potentials were absent

testing and high resolution ultrasound. As stated in Chap. 5, sensory and motor nerve conduction studies are the primary tests to distinguish between axonal and demyelinating neuropathies. CMT Type I (demyelinating) is characterized by a slowing of motor nerve conduction velocities ~20 m/s and prolonged distal motor latencies [25] (Fig. 10.4). Except for in CMT I, such slow conduction velocities regularly only occur in some neuropathies associated with a gammopathy. Moreover, in contrast to acquired

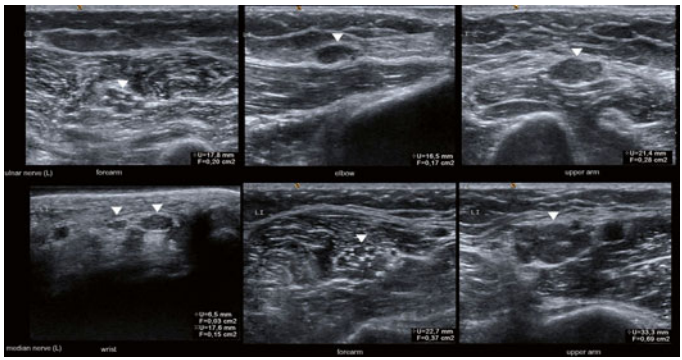

Fig. 10.5 Corresponding ultrasound image of the left ulnar and median nerve (*arrowheads*) demonstrates a severe hypoechoic swelling of fascicles and the entire nerve far away from the typical sites of entrapment

neuropathies, the slowing of nerve conduction velocities in an inherited neuropathy occurs uniformly, whereas acquired neuropathies usually exhibit asymmetric or non-uniform slowing [30]. However, the axonal loss in CMT Type II results in a reduction of the amplitude of CMAP or SNAP.

The most recent diagnostic tool is the high resolution ultrasound. Individuals with CMT have a profound (CMT I) or modest (CMT II) enlargement of peripheral nerve cross-sectional area [31] (Fig. 10.5). If the evidence points to CMT disease, one of the most important issues consist of genetic testing and counselling to confirm the diagnosis [32]. There exists no curative treatment at the moment but supportive therapies may gain a partly symptom control [27]. In summary, important clues for a CMT are:

- Clinical signs of a symmetric "focal" neuropathy such as both-sided carpal tunnel syndrome and/or ulnar neuropathy combined with signs of a distal-symmetric sensory-motor neuropathy of the legs in individuals of relatively young age
- Pes cavus, hammer toes

- Uniformly slowed motor nerve conduction velocities ~20 m/s, prolonged distal motor latencies (CMT I), signs of axonal loss (CMT II)
- Profound (CMT I) or modest (CMT II) enlargement of peripheral nerve cross-sectional area on ultrasound
- Positive family history

10.4.2 HNPP – Hereditary Neuropathy with Liability to Pressure Palsies

Another important hereditary neuropathy mimicking a focal neuropathy is the HNPP, a disorder that was probably first described by De Long in 1947 [33]. The disease is also inherited as an autosomal dominant trait. In contrast to CMT I it is caused by a deletion on chromosome 17, containing the gene encoding the peripheral myelin protein 22kD (PMP22) [34]. The abnormal myelin may be seen in histo-pathological studies in the form of a focal, sausage-shaped thickening of myelin sheaths, also called tomacula [35]. Typically, the disease has an early onset, and it likely occurs in adolescents and in young adults [35]. Due to the abnormal myelin protein, affected individuals suffer from multiple remitting and relapsing nerve entrapments. Just slight pressure, which nerves easily tolerate normally, may induce a nerve impairment in subjects with HNPP (Fig. 10.6). Nerve entrapment often occurs in the peroneal, ulnar and radial nerves, but it has also been described in the brachial plexus [36, 37]. The clinical manifestations include both sensory impairment and muscle weakness.

Electrophysiological studies typically demonstrate features of demyelination such as prolonged distal motor latencies, ubiquitous reduced sensory and motor nerve conduction velocities and a segmental slowing of nerve conduction across the typical entrapment sites. However, nerve conduction is not as low as in CMT I [38]. These changes may generally be observed in asymptomatic individuals at asymptomatic entrapment sites [39].

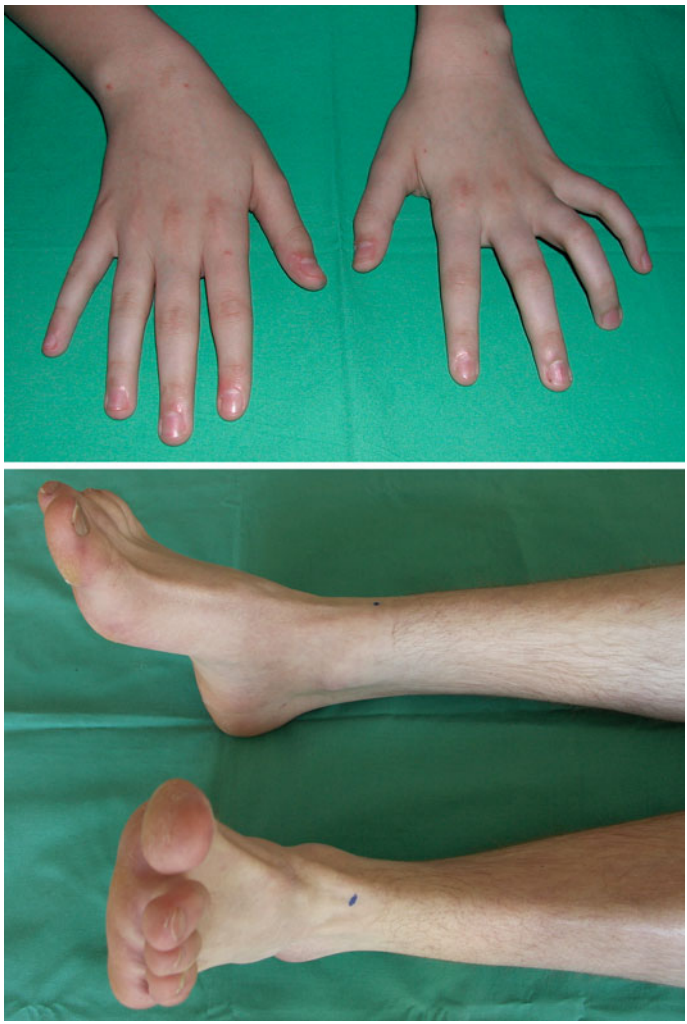

Fig. 10.6 Clinical presentation of HNPP in a 13-year-old boy. Remitting and relapsing nerve entrapments of the left ulnar nerve after excessive computer gaming (*above*). After 10 weeks the patient developed a palsy of the right peroneal nerve after leg crossing (*below*)

As in CMT, high resolution ultrasound of the peripheral nerves provides a novel tool in the diagnosis of HNPP. The average nerve CSA was greater in CMT1 than in HNPP and the most common site of nerve hypertrophy was at sites of entrapment. It is again associated with profound enlargement of peripheral nerve cross-sectional area [31]. HNPP has a favourable prognosis because of possibly spontaneous recovery within weeks or months. In some case reports it has been stated that patients also have received a benefit from surgery of the entrapped nerves [40]. However, it remains unclear whether it was due to surgery or to the natural course of the disease.

Today, it is easy to confirm the diagnosis by a genetic test, whereas meanwhile a biopsy of the sural nerve is unnecessary [41]. In summary, important clues for HNPP are:

- Remitting and relapsing nerve entrapment in adolescents and young adults after just slight pressure, mostly seen in the peroneal, ulnar and radial nerves with a sensory-motor impairment and spontaneous recovery
- Ubiquitous slight slowing of motor and sensory nerve conduction velocities, prolonged distal motor latencies, segmental slowing of nerve conduction across the typical entrapment sites, and changes in asymptomatic individuals
- Profound enlargement of peripheral nerve cross-sectional area on ultrasound most frequently on entrapment sites
- Positive family history

10.5 Autoimmune Neuropathies

10.5.1 Chronic Inflammatory Demyelinating Neuropathy and Variants

Among the autoimmune-induced neuropathies, we have to take into account some conditions which again may be confused

with a focal neuropathy. Most of them are mainly characterized by a chronic course. The classical prototype is a disease also referred to as chronic inflammatory demyelinating neuropathy (CIDP). Its etiology is completely unknown to date. However, similarity to the Guillain-Barre syndrome, as well as response to an immune-modulating therapy, suggest an autoimmune origin [42]. The primary hallmark of CIDP is demyelination followed by secondary axonal loss [43]. The prevalence has been reported to be 1–8 affected individuals within a population of 100,000 inhabitants [44]. These are likely underestimates, since the condition is not always easy to diagnose. Therefore it is supposed that a considerable percentage of the affected subjects are overlooked or misdiagnosed [45]. CIDP is characterized by predominant motor symptoms persisting for more than 2 months together with reduced or absent tendon reflexes. Either progressive or relapsing-remitting weakness may start asymmetrically at the onset of the disease, but it typically affects symmetrically the distal and/or proximal muscles of the extremities, whereas involvement of the cranial nerves is uncommon. In addition, sensory loss such as numbness may also be present. Some of the affected individuals show a self-limiting course [42]. In contrast to hereditary neuropathies, family history is negative. Besides the classical CIDP, there are conditions with an atypical clinical presentation (see below), some of which may be considered variants and others distinct disorders. Due to this heterogeneity as well as the lack of a specific diagnostic test, several diagnostic criteria have been suggested. They all have a high specificity but are less sensitive [46–48]. For instance, the frequently used criteria of the American Academy of Neurology (evidence of demyelination in nerve conduction studies, elevated CSF protein level and signs of demyelination on nerve biopsy) miss more than 50 % of the patients suffering from CIDP, but they approach a specificity of 100 % [49]. Therefore, it has been suggested to start with an immune-modulating therapy if the typical clinical

symptoms mentioned above are present and if additional criteria support the diagnosis [42]. Nerve conduction studies play a key role since they since they demonstrate the underlying process of demyelination in various nerve segments by detection of slowed nerve conduction velocities, prolonged distal motor and F-wave latencies as well as conduction block or abnormal temporal dispersion (please also refer to Chap. 5). Electrophysiologic criteria of the American Academy of Neurology require at least three demyelinating range abnormalities in two nerves; however, they are less sensitive as stated above [46]. Moreover, in 90 % of the affected individuals, an elevated liquor protein is present, and in 29 % of patients antibodies to P0 protein were identified [50]. Furthermore, sural nerve biopsy may yield evidence of demyelination, but it is considered to be less sensitive and less specific [42].

Recently, imaging modalities have been introduced in the diagnosis of CIDP. Hypertrophy of the lumbar or cervical nerve roots as well as of the brachial or lumbosacral plexus and/or gadolinium uptake in MRI may support the diagnosis (Fig. 6.23) [51, 52]. Additionally, high resolution ultrasound provides a cost-effective tool for the detection of focal, multifocal or generalized alterations of the peripheral nerves and plexus. Ultrasonographic findings that have been reported in CIDP cases are characterized by an modest enlargement of cervical nerve roots and non, focal or multifocal peripheral nerve enlargement (see Sect. 6.1.8 concerning HRUS and Sect. 6.2.8 concerning MRI) [53]. However, in some cases focal enlargement corresponding to sites of conduction block was demonstrated. Similar findings are reported to occur in multifocal motor neuropathy (see Sect. 10.5.2), but further investigations are required to determine the potential diagnostic value of these methods [31, 54]. Treatment is immune-modulation either with corticosteroids or alternatively with intravenous immune globulin [55, 56]. As in CMT I, chronic inflammatory demyelinating neuropathy may be confused with multiple entrapment syndromes of the

upper extremities such as both-sided carpal tunnel syndrome due to prolonged distal motor latencies. Therefore, doubts concerning the diagnosis of a focal neuropathy should arise if:

- There are clinical signs, or there is electrodiagnostic evidence, of a demyelinating lesion concerning multiple nerves, especially in the proximal parts of the upper extremity (proximal conduction block)
- There are additional clinical signs, or there is electrodiagnostic evidence of a distal/proximal symmetric/asymmetric demyelinating sensory-motor neuropathy
- There is evidence of focal/multifocal nerve thickening outside the typical sites of entrapment on ultrasound or MRI

Besides the classical phenotype of CIDP, some clinical variants have been described. The most important of them – with respect to the content of this book – is a syndrome first described by Lewis and Sumner in 1982; it is also referred to as multifocal acquired demyelinating sensory and motor neuropathy (MADSAM). The condition is characterized by a chronic multifocal-demyelinating sensory-motor neuropathy of several nerves, reminiscent of a mononeuritis multiplex (see vasculitic neuropathy). Besides signs of demyelination, electrophysiologically, a persistent multifocal conduction block may be evaluated [57, 58]. Apart from that, there are no other distinctions compared to the CIDP. Patients have responded to treatment with corticosteroids and intravenous immune globulin [58]. If only one nerve is clinically affected, the condition may easily be confused with a focal neuropathy, and sometimes it becomes impossible to avoid a misdiagnosis. Therefore, it is very important to take into account this rare disease in the case of failed surgery of a suspected focal neuropathy. We treated a patient who underwent carpal tunnel surgery twice with the typical clinical symptoms. Indeed, electrodiagnostic testing suggested a carpal tunnel syndrome. After failed surgery, high

resolution ultrasound demonstrated multifocal nerve enlargements of the median nerve at the upper arm. Fractionated motor nerve conduction studies have been able to show an abnormal temporal dispersion along the involved nerve segments (Figs. 5.1, 5.2, 10.7, 10.8, and 10.9). Another regional variant of CIDP is also referred to as distal acquired demyelinating symmetrical neuropathy (DADS, anti-MAG positive); it is characterized by a slowly progressive demyelinating and sensory ataxic neuropathy with positive IgM monoclonal antibodies against myelin associated glycoprotein (anti-MAG). It typically presents a distal sensory ataxic neuropathy [59]. In most of the affected subjects, monoclonal gammopathy is of unknown significance, whereas some of the patients show comorbidity with Waldenstrom's lymphoma or chronic lymphocytic leukaemia [60, 61]. In most patients, especially prolonged distal motor latencies may be confused with a focal neuropathy. However, this condition shows neither conduction block nor temporal dispersion [62]. Regarding the other variants (other forms of DADS, sensory CIDP, and cranial perineuritis) please refer to specialist literature. Finally, CIDP may be associated with other disorders (monoclonal gammopathy, diabetes mellitus, CMT disease, or central nervous system involvement).

10.5.2 Multifocal Motor Neuropathy (MMN)

In 1985 Parry and Clarke reported a condition, characterized by weakness, muscle wasting and fasciculations without any sensory impairment [63]. The onset of the disease may be abrupt or slowly progressive, sometimes over more than 20 years, with inactive periods. A generalized as well as a focal or asymmetric affection of one or several nerves has been described, mainly involving the arms. MMN is more common in men than in women, and it can begin at any age.

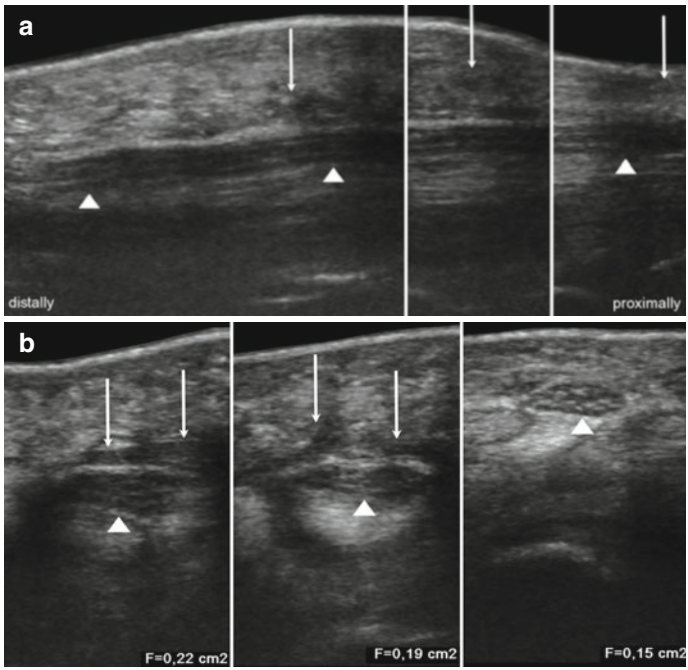

Fig. 10.7 Condition after carpal tunnel surgery of the right median nerve (*arrowheads*) in a patient with typical clinical symptoms of CTS. (**a**): Longitudinal section and (**b**): corresponding cross sections. Ultrasound revealed no residual stenosis despite worsening of the symptoms. *Arrows* point to scarring on the roof of the carpal tunnel

For those reasons, MMN may resemble motor neuron disease [64, 65]. Unlike a motor neuron disease, involvement of cranial nerves is rare, and there are no upper motor neuron signs [66]. Fractionated motor nerve conduction studies show the typical sign of MMN: multifocal conduction blocks, whereas sensory nerve conduction is not impaired. Late secondary axonal loss as well as fasciculations may simulate a motor neuron disease [67].

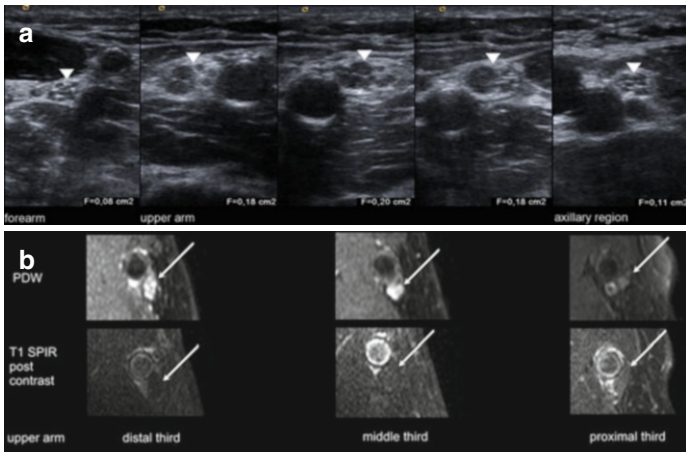

Fig. 10.8 (**a**) Corresponding ultrasound images of the entire course of the right median nerve (*arrowheads*) from Fig. 10.7 up to the axilla. A focal hypo-echoic swelling of the fascicles and enlargement of nerve crosssectional area can be clearly seen at the upper arm on ultrasound images. (**b**) Corresponding MRN shows hyper-intense fascicles on PD-weighted images but no contrast uptake on T1-SPIR images (*arrows*) (MRN courtesy of Karsten Stock, MD (department of radiology and neuroradiology, DessauRosslau Hospital/Germany))

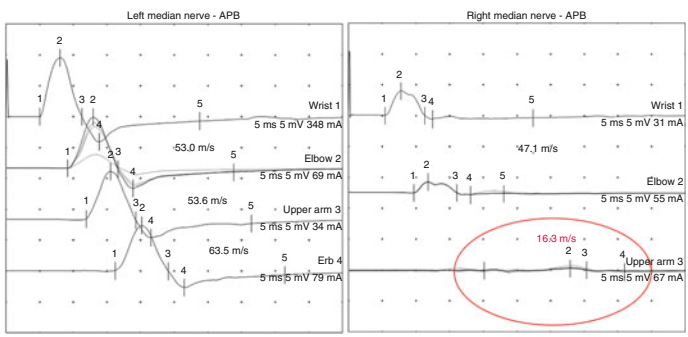

Fig. 10.9 Corresponding motor nerve conduction study of the right median nerve from Figs. 10.7 and 10.8 compared with the left side. A focal slowing of the motor nerve conduction velocity as well as a pathological temporal dispersion in the right median nerve at the upper arm could be obtained. Furthermore, the sensory conduction velocity was slowed (not shown) suggesting a focal CIPD variant

As mentioned above and in Sect. 6.2.8, high resolution ultrasound in individuals suffering from MMN may detect changes similar to those in patients with CIDP, characterized by an enlargement of cervical nerve roots and focal or multifocal nerve enlargement, sometimes corresponding to the site of conduction block (Figs. 6.13 and 6.23) [68]. Therefore, ultrasound may play a key role in differentiating between MMN and a motor neuron disease at an early stage [31, 69]. Only a proportion of patients have high titres of anti-GM1 or GD1a ganglioside autoantibodies [67]. In the majority of cases, MMN responds to a treatment with intravenous immune globulin, whereas corticosteroids are not effective. Moreover, studies with rituximab have been promising [70, 71]. As well as in MADSAM, differentiation between a focal manifestation of MMN and a focal compressive neuropathy may be challenging.

In particular, a multifocal or asymmetric involvement of several nerves without any signs of sensory dysfunction, nerve enlargement outside the typical sites of entrapment on ultrasound, and a multifocal conduction block/abnormal temporal dispersion should suggest the diagnosis of MMN.

10.6 Vasculitic Neuropathies

Vasculitic disorders predominantly affect the small epineural vessels (vasa nervorum) due to a destruction of the arterial wall by inflammatory cell infiltration. This fact causes a stepwise infarction of a single or multiple peripheral nerves or nerve segments also referred to as mononeuritis multiplex (Fig. 10.10) by acute or subacute ischaemia. Some of the patients may also show a distal symmetric nerve involvement. As a result pure axonal loss occurs [72, 73].

Systemic vasculitis may be subdivided into primary and secondary forms. Primary necrotizing vasculitis affecting the

medium-sized and small vessels is the key manifestation of different disorders such as polyarteritis nodosa (PAN), Wegener's granulomatosis (WG), Churg-Strauss syndrome (CSS) and others [74]. Moreover, vasculitis is observed secondary to connective tissue disorders (rheumatoid arthritis, systemic lupus erythematosus, Sjögren syndrome, systemic sclerosis, sarcoidosis and others), or it represents a complication from infection (HIV, hepatitis B and C and others), drugs or malignancy [72].

Vasculitis is a systemic disease. That means that the affected individual shows clinical features of impairment of other organs beside the mononeuritis multiplex [75]. The typical clinical signs of affection of the peripheral nerves are:

- Stepwise involvement of one or more sensory, motor or "mixed" peripheral nerves (mononeuritis multiplex) over weeks or months. With progressing disease these mononeuropathies may overlap leading to pattern of asymmetric neuropathy accounting for three quarters of the patients [76]. As mentioned above a distal symmetric form is uncommon.
- Severe aching or throbbing pain in the affected nerve territories is said to be strongly associated with a vasculitic neuropathy [73]. However, there is a similarity with Parsonage Turner Syndrome (see above) what should not be neglected.

The facts mentioned above explain why vasculitic neuropathies may be sometimes confused with a focal (compressive) neuropathy.

Most commonly, the peroneal, tibial, ulnar and median nerves are involved, either unilaterally or bilaterally [72]. Besides the above mentioned neurological complications vasculitic disorders show general symptoms like weight loss, malaise, subfebrile temperature, and fatigue. Moreover, laboratory tests often indicate features of systemic inflammation, such as an elevated blood sedimentation rate and C-reactive protein and positive anti-neutrophil cytoplasmic antibodies.

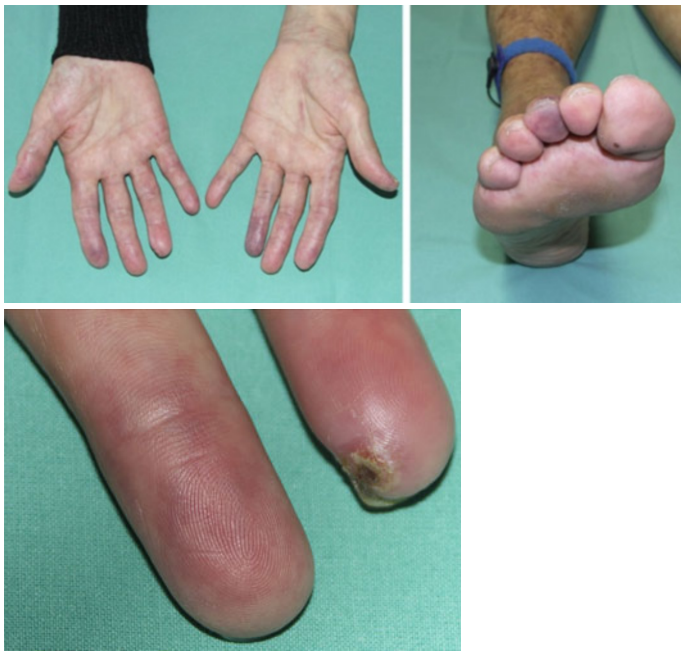

Fig. 10.10 Clinical presentation of polyarteritis nodosa with multiple acute and subacute ischaemic lesions of the fingers and toes

Depending on the underlying basic disease, clinical dysfunction of different organs including kidney, skin, lung, bowel, etc. may be evident [75]. However, it would go beyond the scope of this book to give a detailed overview of all primary and secondary vasculitic disorders, including their special organ manifestations, associated clinical features, and laboratory findings. For further information please refer to the specialist literature [75, 77].

Nerve conduction studies show the typical signs of primary axonal loss, characterized by a reduction of the amplitude of the

compound muscle action potentials (CMAP) and/or the sensory nerve action potentials (SNAP) at all recording sites [78]. In an early stage within a few days after an ischemic nerve infarction, a pseudo-conduction block may be observed, resulting from the fact that Wallerian degeneration has not completely finished [79].

In some small series, nerve hypertrophy was observed in the tibial, ulnar, and median nerves using high resolution ultrasound in subjects with vasculitic neuropathy. After treatment with corticosteroids the cross-sectional area of the affected nerves again decreased [80, 81]. However, the diagnostic value of this method remains unclear at the moment. In the majority of cases, diagnosis of vasculitic neuropathy is confirmed by nerve biopsy or biopsy of another affected organ, as well as by the typical constellation of auto-antibodies in laboratory tests. Treatment includes corticosteroids, immunosuppressants, and intravenous immune globulin [72].

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Chapter 11
Focal Nerve Trauma

Götz Penkert

11.1 General Considerations

Focal nerve lesions due to trauma are relatively rare diseases we have to deal with. The following chapter is therefore quite short. Most nerve trauma is associated with bone fractures or distortions of joints (Fig. 6.8). These injuries are characterized by enormous traction forces which cause long-distance damage to the nerve trunk. Elastic intraneural elements indeed allow resistance against these forces until a certain level is reached. Then the nerve disrupts, and the nerve stumps retract a few centimeters to proximal and distal. Due to axonal re-growth capacity, the proximal stump turns to develop a neuroma, whereas the distal stump gets left behind with intraneural fibrosis. However, fibrosis also occurs in the proximal nerve segment above the neuroma. Therefore, most trauma cases end in large nerve gaps after resection of the entire scar segments so that the term "focal" no longer fits. You will find supporting and detailed modern imaging findings in these cases by means of high resolution ultrasound and magnetic resonance in Sects. 6.1.6, 6.1.7, 6.1.8, 6.2.3 and 6.2.4.

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It is entirely owing to Sunderland and his classification of nerve lesions in five degrees that we have an accurate impression of the possible types of structural damage and subsequent recovery potentials. For further details and references, please refer to Sect. 3.1. In addition, it is owing to Millesi that today we have an exact knowledge of the different types of intraneural soft tissue reactions which, secondarily can hinder the re-growth capabilities of nerve fibers. For further details and references concerning this field, please refer to Sect. 3.2.

In contrast to frequent and extended nerve disruptions, short distance lesions can for instance result, from a thrust with a knife, injury with a piece of broken glass, or a bite from a dog. Such injuries leave behind a circumscriptive trauma, and no physician can immediately differentiate whether the nerve remains in continuity or whether it is disrupted. Theoretically, nerve damage from grades I–V of Sunderland's classification could have happened following one of the above-mentioned injury causes.

In previous decades, two alternative treatments were offered to patients:

1. Subsequent exploration of the nerve lesion under general anaesthesia, neurolysis or nerve repair depending on the intra-operative findings
2. Wait for certain period of time so that we can test the behaviour of the axon sprouts exclusively by eliciting the Tinel sign (see also Sect. 4.1)

Palpation and percussion of the affected nerve trunk at different points enables to trigger an electric current-like pain that the patient describes in the sensitive area which previously belonged to the distribution of the injured axons. The maximum trigger point will coincide with the location of the maximum number of out-growing axon sprout ends. If this point of triggering moves downward during two different investigations, axonal outgrowth is proven and can be estimated as a positive prognostic

factor. If such a trigger point remains at the same level, the physician has to assume that fibrosis is blocking axonal outgrowth, and that a neuroma has developed. Unfortunately, the time span between nerve injury and potential or expected regeneration signs in previously paralysed muscles often lasts for several months. Electrodiagnostics are consequently rather useless during this period, because re-innervation signals can only occur when axon sprouts have arrived at muscle end plates. For details please refer to Sect. 5.6.

Because of today's abilities in high resolution ultrasound imaging demonstrated in Sects. 6.1 and 6.2, we are increasingly optimistic of improving on the time span just described. We are hopeful that new experience in these easily available techniques will help us in future to indicate neurolysis or nerve repair early enough when it is necessary, and to avoid surgery when it proves unnecessary. The timing of decision making influences the quality of regeneration. Muscle fibers which do not receive nerve signals undergo atrophy which is said to start becoming irreversible after 6–12 months, although different opinions exist today concerning this question [1, 2]. Nevertheless, the general opinion is that the prognosis for nerve regeneration and recovery essentially improves the earlier nerve repair starts.

11.2 Surgical Considerations

Even during surgical exposure with aid of magnification by microscope, it still remains difficult to determine whether or not nerve fascicles are intraneurally in continuity. In any case, our surgical goal is that remyelination and re-growth of nerve fibers start, and that this process again reaches muscle end plates. The affected nerve segment is presented as thickened and hardened (Figs. 8.8, 8.9, 8.13 and 9.9). Under magnification we can

longitudinally incise the epineurium in intact and neighbouring parts of this nerve segment. After this step, the external sheath of the epineurium can be partially removed so that several groups of fascicles come into view. These fascicle groups can be slightly separated from each other, a surgical step that now facilitates an approach to the damaged nerve segment. Nerve trauma results in histo-pathologically determined reactions of the connective nerve tissue, a process called fibrosis. If epineurotomy, epineurectomy, and interfascicular neurolysis as stepwise procedures succeed in overcoming the injured nerve segment, prognosis of recovery improves significantly. Circular compressing forces due to fibrosis hinder re-growth of axons. Our efforts aim at reducing such a barrier. By means of intraoperative nerve action potentials (NAP), the surgeon can now examine possibly preserved nerve fiber function. A healthy nerve segment is stimulated proximal to the lesion, and action potentials are deduced from a nerve segment distal to the lesion. The method is entirely credited to Kline and Hudson [3]. In the meantime, quite a new additional approach exists involving applying high resolution ultrasound directly onto nerve fascicles during the operation. However, further experience of centers will be necessary to determine exactly whether fascicle continuity is preserved either in the nerve trunk, or at least in certain nerve sectors [4].

As already mentioned in Sect. 7.1, pure entrapment syndromes predominantly need slight decompression only. However, if a circumscriptive trauma has happened in the patient's history, and if the patient then presents a pre-operative severe loss of function with deteriorated or lacking electrical conductivity, the surgeon must consider a nerve exposure which really needs microsurgical neurolysis. However, we must then take into account the plexiform structure of nerve trunks. Separation of fascicle groups destroys cross connections between them, followed by small neuroma after a period of time. This procedure can result in a painful nerve lesion in the

patient's future. The surgeon bears the whole responsibility and must decide during the operation how far to enter into the nerve and between different fascicles. The stepwise neurolysis should be stopped immediately when fascicles seem to expand under the microscope. Responsibility is high insofar as re-operations carry a high risk of making things worse. Perhaps more experience in intra-operative ultrasound will help in future to assess the type of lesion before microsurgical steps are started. For further details of this feature, please refer to Sects. 6.1.7 and 6.2.7.

Rare focal injuries can induce such a fibrosis and, despite preserved nerve continuity, nerve fiber re-growth can end in a neuroma in continuity, in other words, in a grade IV lesion from Sunderland's classification (see also Figs. 6.7 and 6.18). Such a situation needs the removal of the altered nerve segment or parts of it and repair of nerve continuity by means of grafts. Information on the consequences and microsurgical techniques of modern nerve repair is mainly due to the work of Millesi et al. [5, 6]. However, attention should be paid to the fact that the first nerve repair by means of autologous skin nerve grafts had already been successfully carried out in 1916 by Foester [7, 8]. Nevertheless, during a period of about 40 years, the development of nerve reconstruction unfortunately completely stagnated, despite this pioneer report. As an explanation, we have to consider that only a macro-surgical view of nerve structures was current. Surgeons tried to overcome the usual difference in caliber between the injured nerve trunk and a small nerve graft by introducing so-called "cable grafts". In order to achieve the caliber of nerve trunks, skin grafts were glued together or wrapped in a segment of an artery or vein so that the cable turned out to have the same diameter as the nerve trunk. These cables resulted in a central necrosis because the central sector remained unnourished. Because of its unsatisfactory results, end-to-end suture of the disrupted nerve trunk stumps was more and more preferred over "cable grafts" (Fig. 11.1).

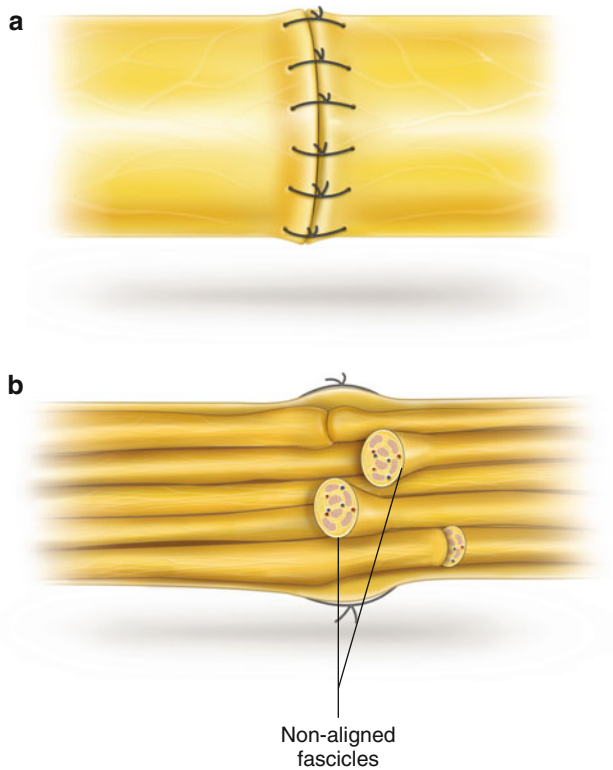

Fig. 11.1 (**a**) Epineural end-to-end suture. (**b**) Non-accurate fascicle approximation; a technique that can result in a suture neuroma

Extremities were then immobilised over weeks in a flexed position. As just mentioned, it is now owing to the work of Millesi et al. that the significant disadvantages of these historical techniques have been realized, and that our theoretical concepts have completely changed. The microstructure of nerve tissue has become well known.

However, even though microsurgical techniques had been introduced in peripheral nerve surgery by 1963 [9], it was more than another decade before Millesi's results could finally prove the accuracy of these concepts (Fig. 11.2) [6]. According to them, the quality of nerve fiber growth and final recovery depends on the following basic preconditions:

1. Accurate approximation of nerve fascicles. Size and distribution of the fascicular structures of the nerve stumps shall be taken into account. If a fascicle is of the same diameter as the nerve graft, co-aptation succeeds easily. If a fascicle group is of about double the diameter of the graft, two sural nerve grafts side by side fit on the fascicle stump. If a nerve trunk consists of only one large fascicle (mono-fascicular in cross-section), such as, for instance, the radial nerve at the upper arm, all grafts may be sutured side by side with the nerve end so that the whole surface of the stump is covered.
2. Fibrosis originates from the epineurium of the nerve stumps. It develops forces which circularly compress the nerve ends. Consequently, fibrotically altered epineurium at the nerve stumps has to be removed over a short distance.
3. Tension at nerve suture lines destroys accurate fascicle approximation, so that scar tissue occurs between insufficiently approximated nerve stumps. In order to avoid tension, grafts has become necessary, and their length should be a little longer than the nerve gap has presented after removal of the fibrotic nerve segments. To achieve an exact co-aptation between graft and fascicle stump, we then generally place one suture with 10-0-monofile suture material only. Too many sutures promote dislocation of fascicles against each other, and foreign body granulomas have to be minimized as much as possible. The patient's own fibrin glue additionally helps to maintain the stump approximation as accurate as possible.

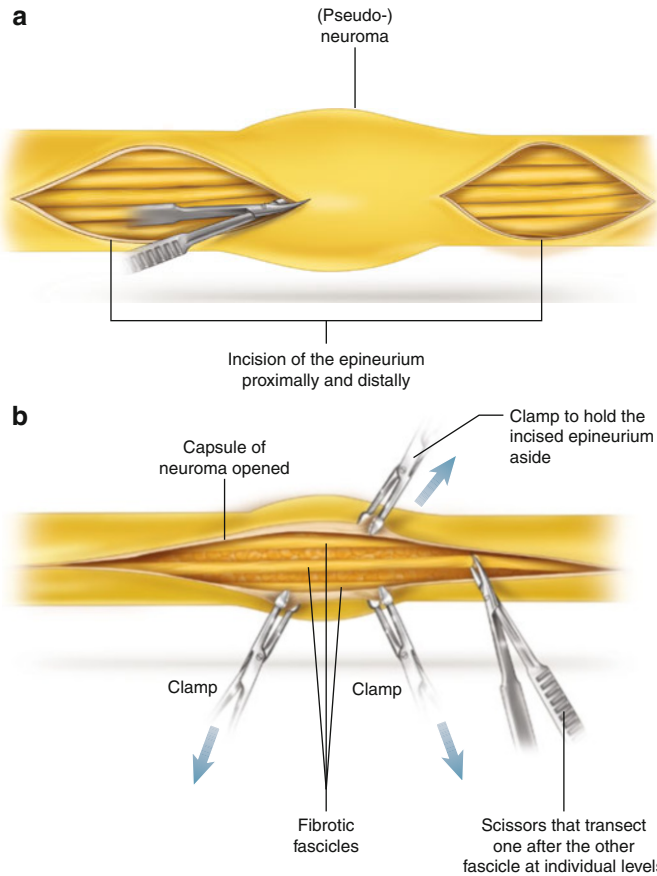

Fig. 11.2 Principles of nerve repair by microsurgical means. (**a**) First step: incision of the epineurium proximally and distally to the neuroma or pseudoneuroma. (**b**) Attempt to open the epineurium around the neuroma; assessment of the amount of fascicle fibrosis; transection of fascicles which need reconstruction. (**c**) Technique of grafting with exact fascicle approximation; the epineurium of proximal and distal nerve ends is removed

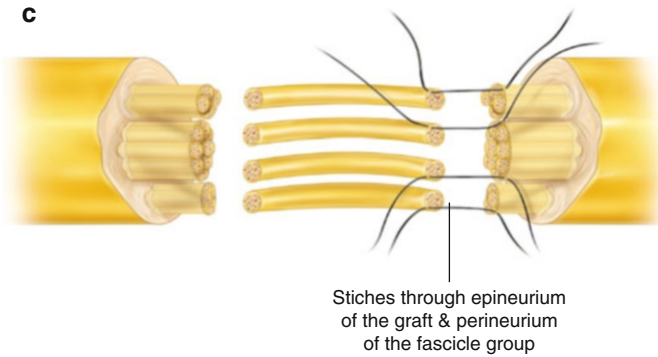

Fig. 11.2 (continued)

As an autologous graft, the sural nerve plays a role of first choice because of its length of about 30–40 cm in adults. We always use three or four small transverse skin incisions to remove it. Sometimes an anastomosis from the common peroneal nerve needs attention and one additional incision. The nerve has always to be extracted over its whole length in order to avoid a painful neuroma, which is located subcutaneously, when we only remove a short segment. Details with regard to each single traumatized nerve trunk of arm and leg, patient's positioning, skin incisions and approaches, and of course, graft removal are described in our previous book from 2004 [10].

11.3 Notes on Nerve Grafting

Arguments on nerve graft re-vascularization grew in importance for a certain period of time. In preceding decades, the so-called "cable grafts" of nerve trunk diameter remained unsuccessful because of insufficient re-vascularization from the recipient bed.

Small skin nerves as grafts are instead said to be re-vascularised in time. The term "in time" means that Schwann cells within grafts are enabled to remain alive by re-connection to a blood supply as early as they require. Functionally intact Schwann cells guarantee isomorphic regeneration and axon-sprouting within the tubuli by means of the growth factors they extrude. By 1946, experiments demonstrated how short the highly sensitive Schwann cells remain alive in culture with and without a blood supply [11]. We could recently prove that small vessels sprouting from the recipient bed generally achieve completed graft re-vascularization earlier than Schwann cells start to lose their viability [12].

We should summarize arguments to use autologous transplants, in other words, skin nerves that the patient sacrifices, with the following statements:

1. Results of graft repairs are better than of end-to-end suture of nerve stumps [6]
2. Re-vascularization of autologous nerve grafts takes place in time [11, 12]
3. Because of this statement: Schwann cells remain alive and keep their viability [13]
4. As re-vascularization starts from the whole recipient bed, the graft length does not play any role as far as adequate nourishment is needed [12]
5. Accurate and tension-free approximation of fascicles succeeds by means of grafts only [6].

Nevertheless, discussions and experiments go on to introduce bioengineered nerve grafts of different designs in order to replace autologous grafts with the following three arguments:

1. The autologous graft is a sacrifice
2. The availability of enough autologous nerve grafts can be limited in case of extended nerve trunk lesions [14]
3. Questions can arise whether bridging a short and focal nerve gap really needs to sacrifice another nerve

Meanwhile, everyone has realized that nerve fiber re-growth generally soon stops when Schwann cells are absent. For this reason, various experimental efforts of high quality are reported to culture and influence Schwann cells to incorporate them into artificial grafts. One group's therapeutic concept is based upon genetically modified adult human autologous Schwann cells [13]. One year earlier, the same group reported, via a rat model, how genetic modification promotes Schwann cells to over-express FGF-2 isoforms. They particularly succeeded in demonstrating which low and high molecular weight FGF-2 isoforms are able to promote and stimulate myelination and axon growth [14]. The second group's concept is based upon mesenchymal stem cells which are able to transform into cells of the mesodermal lineage such as into myelinating cells via transformation using a cocktail of cytokines. They succeeded in pointing out that these cells promote nerve regeneration in autologous muscle conduits [15]. Nevertheless, if artificial conduits are considered in future and in humans with long nerve gaps, an important – and in the literature until now unsolved – question will always rise as to how to vascularize these artificial implants because all kinds of Schwann cells need connection or re-connection to blood supply from the recipient tissue to remain viable. Therefore, in future, intensive efforts will have to focus on how to establish an early blood supply in such conduits.

11.4 Notes on Electric Stimulation

A short comment should be made concerning the potential of electric stimulation. In principle, we have to differentiate between electric stimulation of either nerve fibers which regenerate or of a muscle bulk which is in denervation. Despite clinical use of low frequency stimulation being current, convincing scientific proof is rather scarce [16]. Our personal experience is that large muscles or muscle groups which generate mass movements profit by

electric stimulation, whereas target regions of highly differentiated functions, for instance hand muscles, do not profit at all. Low frequency stimulation of nerve fibers just after repair seems to accelerate axonal out-growth in animal models [17] and even in humans according to experience gained from carpal tunnel release [18]. However, the question arises whether we really need this effect. Extraordinary disturbing effects are found if stimulation is applied to a repaired facial nerve and its muscles in rat experiments: it is reported that a collateral over-branching starts and results in severe synkinesias. According to the authors, accelerated miss-sprouting occurs within the level of target muscles and leads to additional mass reactions [19]. It is a reminder that patients frequently sustain such synkinesias when a Bell's palsy recovers, or when a partial facial nerve lesion has followed cerebello-pontine angle surgery. Experience has then shown that mechanical stimulation of facial muscles, application of ice, and exercises in front of the mirror are mostly favoured to promote recovery, whereas electric stimulation should absolutely be avoided. Supplemental experiments have, meanwhile, also succeeded in confirming the superiority of mechanical stimulation in rat vibrissal muscles during recovery [20, 21]. To summarize: the more diverse the dedicated functions that have to re-supplied are, the less electric stimulation seems to be indicated.

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Chapter 12
Nerve Torsion (Rotation)

Götz Penkert

For about 20 years, observations and reports have appeared about exceptional cases of suddenly rotated nerve segments [1]. The etiology of this phenomenon is unknown to date [2, 3]. Small nerves such as the anterior interosseous (see Sect. 8.13) and posterior interosseous (see Sect. 8.10) nerves, suprascapular and axillary (circumflex) nerves, and also the radial nerve trunk at level where it pierces the lateral intermuscular septum (see Sect. 8.9) can be affected [2–4]. An hourglass-shaped constriction occurs as a single event or even as multiple manifestations (see Sect. 10.3).

An acute pain is followed by a gradual onset of functional paralysis of the muscle group which the affected nerve supplied. As suspected pathophysiology, a local inflammation of unknown etiology (see also Sect. 6.2.8), perhaps together with repetitive or rotatory movements, are in discussion. An increased stiffness of the nerve fascicles is said to make them less adaptable to transversely running structures like vessels or fascias that can act constricting or fixing [2].

In high resolution ultrasound, the phenomenon appears as an hourglass-shaped structure of hypoechoic appearance with a

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proximal more than distal thickening without hypervascularization (Figs. 6.11, 6.24, and 6.25). Between both segments, a complete constriction of the nerve can be visualized [5].

The surgical approach to this scarce lesion consists of microsurgical exploration by means of epineurotomy as the first step. A careful derotation should follow, a procedure which was reported early as being helpful [6], but, in our own experience, subsequent segment resection and repair by grafts are needed [4], in agreement with the cases in literature [7].

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Chapter 13
Nerve Tumors

Götz Penkert

13.1 Types, Symptoms, and Diagnosis

Most types of nerve tumor are characterized by the same slowly proceeding symptoms that we consider as typical of all the other previously mentioned focal neuropathies. Therefore, it normally takes several months to find the correct diagnosis and to differentiate from alternative neuropathies. Either the patient notices a focal swelling in his extremity not on a particular day, or eventually increasing symptoms give reason for imaging examination which leads to finding a tumor.

In the following, we restrict ourselves to benign space-occupying lesions either located extra- or intraneurally for which a revised overview was recently published in literature [1]. In all of these cases, the slowly increasing space-occupying effect on nerve axons causes the typical electric-current like paresthesias, in other words, the typical Tinel sign, which characterizes all kinds of focal neuropathies with partial or complete loss of axon continuity (see Sect. 4.1). The point where these sensations are felt most strongly indicates the location of degenerating and

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regenerating neuronal sprouts. First, patients repeatedly notice the location of their sensation, and then they sometimes observe something like a tumor mass which is movable transverse to the nerve course. Later, it becomes more and more sensitive.

Neurological deficits occur rather late. Unfortunately, electrodiagnostic testing is of subordinate value to find the diagnosis because it cannot reveal the correct pathology (see Chap. 5). However, imaging either as high resolution ultrasound or as magnetic resonance imaging is of superior value, and it should therefore be arranged in any case (see Sects. 6.1.7 and 6.2.7).

13.2 Surgical Considerations and Prognosis

The introduction of microsurgery has extended our surgical horizons in tumor cases. Results have massively improved so that it seems increasingly difficult to justify cases where the involved nerve is completely sacrificed. As to be expected, secondary nerve repair after nerve sacrifice achieves a lower functional level than a microsurgical primary procedure with preservation of unaffected nerve fibers (Fig. 13.1).

Therefore, surgery seems mandatory in all kinds of benign histology and solitary manifestation. In most of all cases, imaging findings are sufficient to define the entity as a benign one what will facilitate to decide to operate on. The question about solitary or multiple tumor occurrences in a limb can be solved more easily by means of ultrasound because MRI is technically restricted for certain limb segments. Because most nerve tumors are revealed as solitary, they have to be assessed as easily and primarily accessible; thus we should not hesitate to apply our microsurgical experience and remove the pathology completely.

As a principle, the exposure of all kind of benign tumor first requires skin incisions large enough to identify the uninvolved nerve structure above and below the tumor. A few centimeters of the entering and exiting nerve segments should be visible,

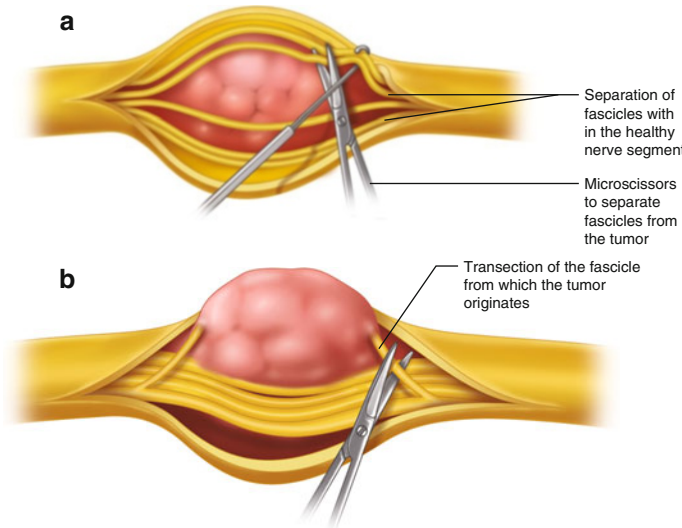

Fig. 13.1 Principles of microsurgery of a schwannoma. (**a**) Exposure of fascicles proximally and distally to the tumor as the first step. (**b**) Identification of the originating fascicles at the tumor poles, their transection, and then complete tumor removal as the final step

because the microsurgical procedure better starts from these healthy nerve segments. Its epineurium is longitudinally incised and perhaps partially removed so that a careful separation of fascicle groups is possible. The whole procedure is comparable to what we previously described in Sect. 7.1 as "interfascicular" neurolysis. Especially less experienced surgeons should make it a habit to start with these principles, because most of the fascicles and fascicle groups divide and disappear between the outer layers of the tumor sheath. These fibers are easier to identify and preserve when the surgeon arrives at the tumor mass with all fascicle groups in view from the beginning. They occur all over the whole tumor circumference. Under the microscope they can be carefully separated from the tumor and, sometimes, micro-scissors are needed for dissection. At the end, they

remain preserved anatomically and functionally. When you finally return to the entering and exiting nerve segments, at least in cases of neurinomas, you will identify one fascicle or a small fascicle group from which the tumor originates. This small structure must be transected above and below the tumor so that you can take out the tumor and close the wound [2].

As the anatomically visible fascicular nerve pattern at different levels does not simultaneously correspond to a functional grouping, the sacrifice of the involved fascicle or fascicle group does not result in a noticeable neurological deficit. Mostly, nerve fibers from which a tumor originates have already lost their function during the growth of the tumor. The functional loss is then commonly pre-operatively compensated. It is therefore reasonable that the prognosis of most benign tumor surgery is extremely good provided the tumor addressed early enough and no other surgeon has previously tried to remove it macroscopically.

Special considerations are needed if structures of the brachial plexus, especially at trunk level, are involved. The nerve tissue differs from more peripheral nerve segments insofar as the typical inter-fascicular pattern is absent in favour of a mono- or bi-fascicular structure. We find fewer collagen fibers within plexus trunks, and a perineural layer which envelops several fascicle groups is lacking. Therefore, supra-clavicular nerve trunks have less resistance to surgical manipulation. Nevertheless, by means of all our microsurgical efforts, a complete removal of a solitary brachial plexus tumor is less difficult than perhaps expected [3].

To summarize on prognosis, the surgical principles described above can always be applied to neurinomas (schwannomas) which represent the most common nerve tumor occurrence in humans. Consequently, they have excellent prognosis, independent of their location.

Neurofibromas, however, differ insofar as the nerve structure from which they originate includes more fascicles, sometimes

a whole fascicle group. Removal of such a tumor results in a slightly more functional sacrifice, but, nevertheless, its prognosis remains excellent [2].

In contrast to that, plexiform and multiple neurofibromas need quite another approach on prognosis than we would expect, taking into account the solitary tumors just mentioned [2]. The same holds true in rare cases of perineurioma, previously termed "local hypertrophic neuropathy" [4]. Both entities would theoretically need a complete nerve trunk resection and repair by grafts, but hardly any surgeon could bear responsibility for such a sacrifice.

Again, different comments on prognosis are needed in cases of intraneural multiple ganglia [5]. However, provided that microsurgical principles are kept in mind, the surgical prognosis is almost as good as for solitary neurinomas.

Special remarks on all benign nerve tumors will follow. It applies to all of these lesion types that a microsurgical step-by-step procedure remains the key to preserve the nerve function. Prognosis will then remain at a high level and independent of the availability of high-end diagnostic tools. Under magnification, the surgeon can, step by step, visualize all the details needed to decide and proceed correctly.

13.3 Special Comments on Different Tumor Types

13.3.1 Schwannomas (Neurinomas)

These tumors originate from Schwann cells of one single fascicle. Several manifestations within one nerve trunk may rarely occur, but then each time originate from another fascicle.

Imaging modalities are demonstrated in Figs. 6.10 and 6.20

Independent of the location, microsurgical removal always succeeds without any significant functional deterioration [2]. Even schwannomas of the brachial plexus in the supraclavicular area can be completely removed as described [3]. The surgeon then needs to remember that the fascicular pattern differs from nerve segments located in the periphery insofar as nerves at trunk level are mono- or bi-fascicular and with less collagen filaments inside. The challenge to microsurgical abilities is a little bit higher, but the prognosis to remove the tumor completely is excellent. A recurrence is only to be considered when fascicle involvement already starts at a very high level in the neuroforamen. These patients will need a follow up by MRI examination over the years and perhaps a second neurosurgical spine approach.

13.3.2 Neurofibromas

These tumors differ from schwannomas with regard to the histological pattern, and consequently, in some surgical aspects [2]. They are comparably encapsulated, but they originate from several fascicles or even some fascicle groups of the involved nerve trunk. Imaging modalities are shown in Fig. 6.21. At the end of the microsurgical neurolysis, not a small fascicle is found, usually several fascicles enter and exit the proximal and distal poles of the tumor. All these fibers have to be sectioned for tumor removal. Mostly, a numb skin area results, rarely a slight paresis which normally recovers within a few weeks.

Again, multiple manifestations within one or several nerve trunks are possible. They can sometimes be associated with a neurofibromatosis type I. Nevertheless, several and different surgical approaches are then necessary, because waiting deteriorates the prognosis; in association with a neurofibromatosis disease, there is a 15 % higher risk of malignant transformation so that a tumor removal would seem mandatory, particularly if its localization allows an easy approach [2].

13.3.3 Plexiform Neurofibromas

The term "plexiform neurofibromas" is used to describe a situation where multiple neurofibromas are localized within one circumscriptive nerve trunk segment. Each tumor has its own capsule, but all tumors can be surrounded by the nerve epineurium (see Figs. 6.11 and 9.7). Plexiform manifestations are more likely to be painful than solitary tumors. A complete removal would now need to sacrifice the whole nerve segment, and it would result in a paralysis [2]. Nerve repair as described in Chap. 11 should theoretically follow, but patients commonly do not accept such a procedure without a guarantee that the function will return to that of the preoperative level. Patients today favour a conservative view and require a follow-up by imaging. If doing so, explicit information about the 15 % risk of malignant transformation is necessary when association with a neurofibromatosis exists [2]. These patients need strict imaging follow-up (Fig. 13.2).

When a surgeon unexpectedly comes across such a situation, he should longitudinally open the outer epineurium and stop the procedure. Such a careful decompression may diminish the

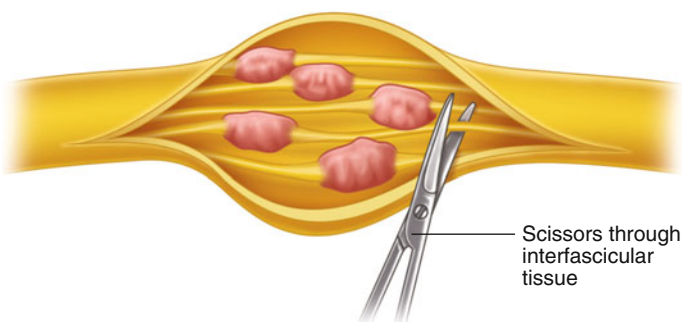

Fig. 13.2 Plexiform neurofibromas. Each fascicle is tumor-involved. Removal of all of them would result in a complete nerve sacrifice

patient's radiating pain. Further action must be thoroughly discussed with the patient.

13.3.4 Perineurioma

These very rare benign peripheral nerve sheath tumors have only been included in the WHO classification system since 2000 [6]. Previously, the term "localized hypertrophic neuropathies" was used [7]. Unaware of nature of these tumors, trials of neurolysis were first reported. A complete nerve segment sacrifice with graft repair was then temporarily recommended [4]. Because of the irreversible muscle atrophy following extended waiting behavior this kind of surgery was even advised as an early procedure. But as the borderlines between tumor and healthy nerve segments could not be defined for sure – not even under high magnification – tumor resection seems to get again abandoned and neurolysis together with simultaneously clarifying the histology is preferred today [6]. Late cases, even with loss of spontaneous muscle activity in myography, should better be treated by reconstructive surgery if the type of functional loss is suitable for tendon transfer. We do not have sufficient information as to whether the tumor itself can then remain in place, because reliable long-time follow-ups are unavailable. Unfortunately, differential diagnosis by means of imaging remains challenging at present (see Sects. 6.1.7 and 6.2.7; Figs. 6.11 and 6.22).

13.3.5 Nerve Sheath Myxoma (Neurothekeoma)

A new benign entity in neuropathology consists of tumors of predominantly cutaneous or subcutaneous localization. They usually occur in the head and neck region, but also sometimes intra-spinally with relation to nerve roots [8]. Occasionally they can arise from peripheral nerves in the upper extremity. Nerve sheath myxomas are immunoreactive for S-100 protein and

neuron-specific enolase. Neurothekeomas are not immunoreactive for S-100 protein, but their cells become stained after incubation with PGP9.5-antibodies. Ultra-structural observations and immunoreactions seem to confirm neuro-ectodermal origin.

We have no experience with behaviour and operability of these tumors. However, we were recently confronted with a 25-year-old patient who underwent removal of a nerve sheath myxoma in Guyon's canal. Because he complained of severe functional deficits afterwards, hand surgeons advised an interosseous anterior nerve transfer onto the motor branch of the ulnar nerve in the palm [9]. He asked for a second opinion. As some single voluntary potentials could be derived from the adductor pollicis muscle, we warned against doing so.

13.3.6 Haemangioma

An intraneural cavernous haemangioma occurs extremely rarely, but it can be completely removed if microsurgical principles are applied as described at the beginning of this chapter [10]. A combination of neurinoma and haemangioma elements has been described, e.g. in 1978 [11], probably one of the entities termed "hybrid tumors" in 2012 [1].

13.3.7 Fibrolipoma

Fibrolipomas in the peripheral nerve are hamartomas, comparable to fibrolipomas of the intraspinal terminal myelon. They are detected in children and young adults, and they mostly involve the median nerve and its branches in the palm [12]. In about 30 % of cases they are associated with macrodactyly [13]. Patients with median nerve involvement commonly present with a carpal tunnel syndrome so that a surgeon can be unexpectedly confronted with such a tumor mass. The surgeon then should remember these rare hamartomas and should stop after carpal tunnel ligament transection. Each microsurgical trial destroys

the nerve fibers such as in the myelon. The tumor has instead to remain in place for life.

Less frequently, fibrolipomatous involvement of other peripheral nerves of the upper extremity has been reported, such as in the radial nerve or the ulnar nerve (cited from [12]).

13.3.8 Ganglia

Focal neuropathies due to ganglia have the third position in terms of rate of occurrence of nerve-related tumors. They have a space-occupying effect, but they are no neoplasia. In contrary to this, they are cystic extraneural (Figs. 6.9 and 6.19) or even intraneural manifestations with hidden communication to a joint in the neighbourhood [5]. By the way, they have been well known for more than 110 years [14].

Extraneural manifestations have to be operated on by external "neurolysis" comparable to what we do for entrapment release. If possible, the communication to the neighboured joint should be ligated in order to avoid recurrence. Typical locations of occurrence are in the neighbourhood of the tibio-fibular joint with involvement of the common peroneal nerve, in the neighbourhood of the ankle with involvement of the tibial nerve in the tarsal tunnel, and in the neighbourhood of the elbow joint or wrist with involvement of the ulnar nerve. Rather rarely, the supraclavicular nerve is affected by single or multiple ganglia which communicate with the gleno-humeral joint. They can be punctured ultrasound guided (Fig. 13.3).

In contrast, single and multiple intraneural ganglia are a challenge for surgeons (see Figs. 6.12, 6.19, 9.19 and 9.20). Localizations of occurrence are the same as in the case of extraneural manifestations. The surgeon needs microsurgical equipment, and he has to apply all principles of "inter-fascicular neurolysis" over the whole involved nerve segment. Each cyst has to be evacuated only, and its membrane cannot be removed

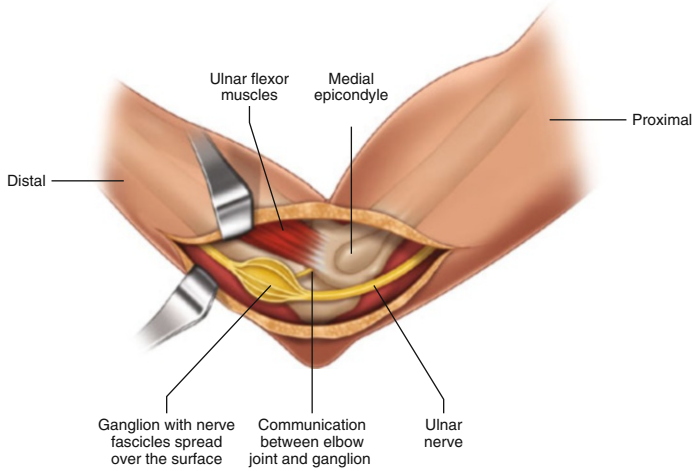

Fig. 13.3 Ganglion mimicking ulnar nerve neuropathy. Nerve fascicles spread over the capsule surface. A communication between the elbow joint and the ganglion is visible

because it contains invisible nerve fibers. There is no argument to sacrifice the nerve at all. On the other hand, no cyst can be neglected in order to avoid persistence of the problem. The exposure therefore needs to be long enough. Nevertheless, a recurrence is sometimes possible as described in the literature because detection of the communication to the joint remains the main problem [5]. We remember a patient whose deep peroneal nerve branch was involved over the whole course. A recurrence came after a few months despite ligation of a communication to the tibio-fibular joint. He was treated by a posterior tibial tendon transfer to restore the partial foot drop described as a reconstructive alternative [15]. The multiple cysts were left in place. To summarize: intraneural ganglia need real microsurgery, cyst evacuation, ligation of the joint communication, and imaging follow-up perhaps once or twice.

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Chapter 14
Focal Tumor-Like Lesions

Thomas Schelle

Similar to autoimmune neuropathies, plexiform growth variants of tumors of the peripheral nerve sheath (plexiform neurofibroma, perineurioma) may lead to a focal, mostly fusiform, nerve swelling with hypertrophic fascicles. High resolution ultrasound then evaluates a focal enlargement of the cross sectional area of the affected nerve and its fascicles as well as a hypoechoic appearance or even a complete masking of the normal echotexture. Moreover, tumors of the peripheral nerve sheath are characterized by a gadolinium uptake on MRI, whereas different reports exist regarding this feature in the case of autoimmune neuropathies. However, careful anamnesis survey, clinical examination, and additional electrodiagnostic testing will help to differentiate between these entities (see Chaps. 5, 6 and 10).

In addition, a few, mostly very rare (except leprosy) disorders may lead to a focal nerve involvement corresponding to enlarged nerve segments or nerve roots. Clinical impairment is dependent on the underlying etiology including the type of affection, either axonal or demyelinating, and the time course (e.g. relapsing-remitting, chronic-progressive, self-limiting, etc.).

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Due to the enhanced imaging capabilities, including high-field MR-neurography and high resolution ultrasound, nowadays we are able to depict nearly every single nerve or nerve root of the human body. Therefore, to an increasing extent we see subjects with a focal neuropathy/mononeuropathy in whom the usual differential diagnosis provides none of the common causes of focal nerve enlargement such as in case of nerve compression, nerve sheath tumor, traumatic nerve fibrosis, poly-neuropathy, etc. In these special cases, differential diagnosis remains challenging, but it remains important because of different treatment options. However, a fascicular biopsy will mostly still become necessary to detect the underlying condition.

According to our own experience and to the available literature, the following section represents the standard of knowledge about these rare entities at the time of completion of this book. Beside plexiform growth variants of tumors of the peripheral nerve sheath and autoimmune neuropathies focal, fusiform, tumor-like nerve enlargement has been described in the following conditions:

- Sarcoidosis affecting the peripheral nerve
- Leprosy affecting the peripheral nerve
- Amyloidosis affecting the peripheral nerve
- Extramedullary affection of the peripheral nerve due to a plasmacytoma
- Primary lymphoma affecting the peripheral nerve

Sarcoidosis is an inflammatory systemic disorder of unknown etiology, characterized histologically by the formation of non-caseating granuloma. In the majority of cases, lymph-nodes and lungs are involved, whereas affection of the nervous system counts for approximately 5–15 of the cases only [1, 2]. The overall incidence of this rare condition has been estimated to be around 20 per 100,000 [3]. Common manifestations of neuro-sarcoidosis within the central nervous system are non-caseating granuloma of the cranial nerves (mostly facial and optic nerves),

hypothalamus and pituitary gland [1]. In contrast, involvement of the peripheral nervous system has been considered to be rare, subacute with a wide spectrum of clinical manifestations such as large-fiber mono-neuropathies, poly-radiculopathy, distal-symmetric (predominantly motor) and small fiber polyneuropathy [4–8]. Granulomatous affection of the peri- and epineurium as well as an ischaemic vasculitis may lead to axonal loss or focal demyelination due to local pressure [6, 7]. To our knowledge, one recent report exists on focal affection of the ulnar nerve due to an epineural and perineural lesion, confirmed histopathology as non-caseating sarcoid granuloma. After surgery the condition improved [9]. We ourselves observed a case with involvement of the superficial radial nerve, clinically characterized by slight numbness with swelling and erythema on the left distal forearm as well as skin manifestations of sarcoidosis (erythema nodosum). High resolution ultrasound was able to demonstrate a significant hypoechoic segmental swelling of fascicles of the superficial radial nerve as well as an encasement by an (also hypoechoic) granuloma. Moreover, we were able to demonstrate the hyper-vascularization of the epineurium and the granuloma (Fig. 14.1).

Leprosy (HANSEN´s disease) is an infectious disorder leading among others to a granulomatous affection of the peripheral nerves. It is caused by *Mycobacterium leprae* and *Mycobacterium lepromatosis*. The disorder is common in a few countries such as, e.g. India, China and Egypt with about 250,000 new cases being detected every year [10]. Lepromatous leprosy usually affects the peripheral nerves diffusely, whereas the tuberculoid form is restricted to a smaller area of a nerve due to an active immunologic response [10]. The reactions mentioned above will lead to a clinically palpable nerve thickening. Therefore, it is no surprise that high resolution ultrasound is also able to demonstrate these alterations. Recently, it has been shown that in patients with leprosy the peripheral nerves and the surrounding epineurium were significantly thicker as compared to controls.

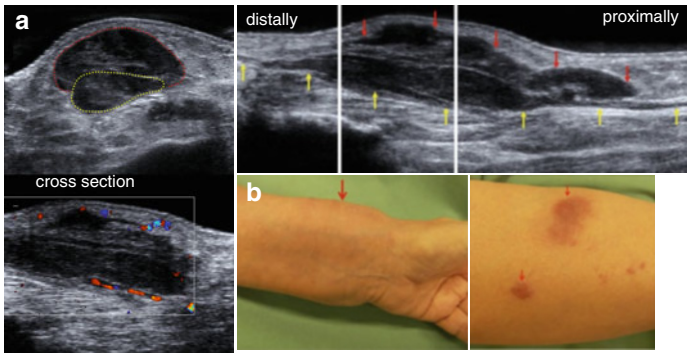

Fig. 14.1 Involvement of the superficial radial nerve in a patient with sarcoidosis. (**a**) HRUS, segmental hypoechoic swelling of the superficial radial nerve as well as effacement of its fascicles (*yellow arrows*, *yellow dotted line*). The encasement by the hypervascularized granuloma (*red arrows*, *red dotted line*) is clearly to be seen. (**b**) Clinical presentation with swelling and erythema on the left distal forearm as well as skin manifestations of sarcoidosis (erythema nodosum), *red arrows*

Furthermore, in 50 % of cases a partial or total loss of the normal nerve-echotexture, characterized by masking of fascicles or hypoechoic fascicles could be demonstrated. Moreover, an increased endo- or perineural flow suggestive of increased neural vascularity by colour-Doppler was observed in 26 % of involved patients, and it was always associated with leprosy treatment reactions in patients undergoing antibiotic therapy [11–13]. Therefore, in future ultrasonography may prove to be a useful technique for diagnosis as well as to assess the extent of the disease, the response to antibiotic therapy, and treatment reactions in patients undergoing antibiotic therapy. Surgery is restricted to releasing entrapment situations.

The term *amyloidosis* describes a rare condition wherein normally soluble proteins are deposited as insoluble amyloid in the extracellular space of body organs; they cause abnormal

protein build-up in tissues and may lead to organ dysfunction [14]. Therefore, acquired amyloidosis is not a disease of its own but associated with other diseases like renal failure, which needs long-term haemodialysis [15]. On the other hand, inherited forms have been described. The prevalence is said to be fewer than 200,000 individuals within the Unites States of America [14]. There are 27 amyloidogenic proteins associated with known human diseases. For a detailed overview of the clinical features, diagnosis and therapy of the condition, please refer to the specialist literature [14]. If a biopsy of the affected organ was carried out, deposited amyloid can be identified histologically by a pink appearance after Congo red staining using bright-field microscopy, whereas in polarized light it demonstrates an apple-green birefringence [16]. However, the different types of amyloid are indistinguishable using light microscopy and other procedures like immunogold electron microscopy or mass spectrometry are needed instead [17]. The main presentation of AL-amyloidosis is a generalized distal symmetrical sensory-motor neuropathy. In a patient with AL-amyloidosis neuropathy, electrophysiologically characterized by a pure axonal loss, high resolution ultrasound could demonstrate a generalized thickening of the peripheral nerves, caused by an increasing number of the identifiable fascicles, but with no specific pattern to these changes [18]. Focal mononeuropathy, especially of the upper limbs, is an uncommon presentation of primary amyloidosis and was confirmed by nerve biopsy (AL, light chain amyloidosis) in one case [19]. On the other hand, only a few focal tumor-like lesions in some peripheral nerves of the lower extremities or cranial nerves due to amyloidosis have been described in the literature so far, also referred to as amyloidoma. One paper reported amyloidoma of the sciatic nerve and the potential value of highfield-MR for detection [20]. Another paper stated that in two cases the amyloidoma focally affected the lumbosacral roots and plexus [21]. Both, one case report and a small study, identified four cases of an amyolidoma of the trigeminal nerve

affecting the Gasserian ganglion [22, 23]. Interestingly, almost all amyloidomas of the peripheral or cranial nerves were organ restricted neoplasms, unrelated to systemic amyloidosis and of the AL- lambda type. Their biological behavior was rather indolent, reminiscent of a lymphoma of mucosa-associated lymphoid tissue [23]. On MRI a gadolinium-uptake without significant surrounding tumor-oedema was seen. By means of high resolution ultrasound, we could identify a hypoechoic lesion of the common radial nerve involving its complete course from the axilla to the radial tunnel, with a marked segmental enlargement (CSA = 0.62 cm^2, diameter 7.6 mm) at the distal part of the spiral groove. In this segment, sonography indicated the presence of calcification which was later histologically confirmed to be a small metaplastic ossification. Furthermore, fascicular biopsy revealed amyloid deposition using Congo red stain and polarized light microscopy (Fig. 14.2). Curative surgical treatment by means of microsurgery was impossible.

Extramedullary manifestation of plasmacytoma is reported in 6–20 % of the patients at the time of diagnosis and during the course of the disease [24]. It commonly affects the nasopharynx, larynx, and upper respiratory tract [25]. Other manifestations including the peripheral nerve and the central nervous system are very uncommon [26, 27]. There is a recent case report of an extramedullary manifestation of plasmacytoma in the median nerve at the upper arm in a patient with IgA-plasmacytoma following stem cell transplantation with temporary remission of the disease. Together with a palpable mass at the medial part of the upper arm, a progressive loss of the sensory-motor function of the median nerve could be diagnosed. Electrodiagnostic testing of the median nerve revealed an axonal loss. Using MRI and high resolution ultrasound, a hypervascularized, rather long fusiform mass along the median nerve was seen with well-defined borders together with an inhomogeneous, rather hypoechoic echotexture with cysts inside (Fig. 14.3). A fascicular biopsy confirmed an extramedullary plasmacytome expressing the kappa light chain antigen [28].

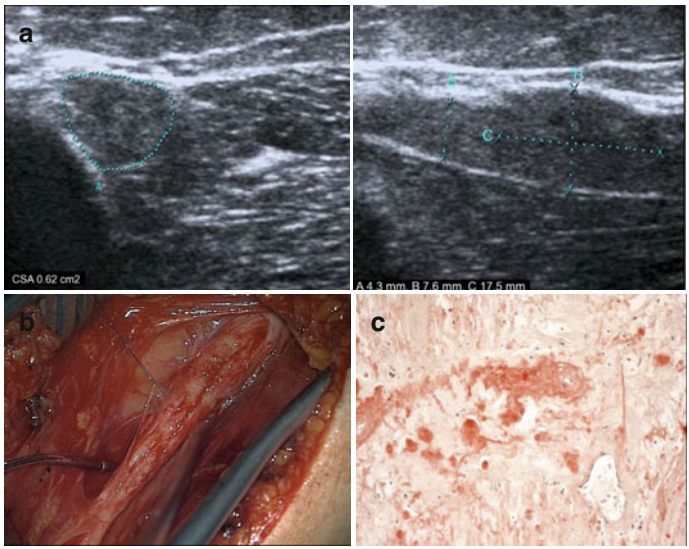

Fig. 14.2 Involvement of the common radial nerve by an amyloidoma. (**a**) HRUS images (*left* cross section, *right* longitudinal section) demonstrate a focal, hypo-echoic nerve swelling with effacement of the fascicles. (**b**) Intraoperative image shows a fusiform nerve swelling. A fascicular biopsy was carried out (Courtesy of TN Lehmann, MD (department of neurosurgery, Bad Saarow/Germany)). (**c**) Histological examination revealed amyloid deposition using Congo red stain and polarized light microscopy (Courtesy of S. Koch, MD, PhD (Department of Pathology, Bad Saarow/Germany))

Primary lymphoma of the peripheral nerve is a primary manifestation of a mostly b-cell non-hodgkin-lymphoma within the peripheral nerve. In the majority of the case reports or small case series the sciatic nerve was involved. Besides the clinical impairment electrodiagnostic testing shows signs of axonal loss. According to our own experience it is characterized by the same signs as mentioned above under extramedullary manifestation of plasmacytoma using HRUS. However, the vascularity may be less pronounced. The lesion shows contrast uptake on MRI [29–31].

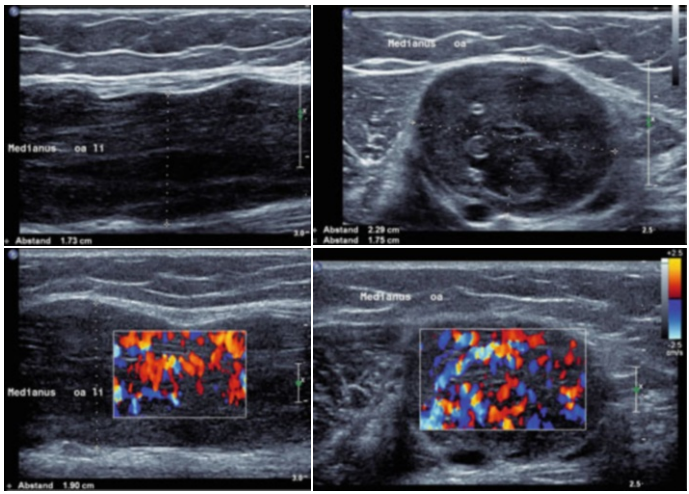

Fig. 14.3 Extramedullary manifestation of plasmacytoma in the median nerve at the upper arm in a patient with IgA-plasmacytoma following stem cell transplantation. HRUS, longitudinal sections (*left*) and cross sections (*right*) with the corresponding colour-Doppler images *below*. A hypervascularized, rather long fusiform mass along the median nerve is to be seen with well-defined borders together with an inhomogeneous, rather hypoechoic echotexture including cysts inside (Courtesy of Steve Dettmann, MD (Department of Neurology, Chemnitz/Germany))

Moreover we recently have observed a case of a metastasis in a female suffering from breast cancer within the median and ulnar nerves as well as the subpectoral and supraclavicular brachial plexus and also infiltrating the roots C7 and C8.

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