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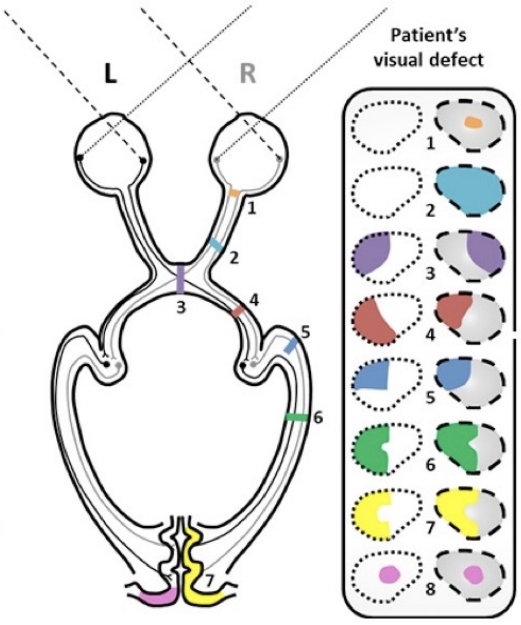
Manual of the Neurological Examination for Neurologists in Training



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Preface

The art of the clinical bedside examination has been a core feature of neurology for decades. Besides a clinical history proper examination against a background of clinical knowledge is a key aspect when making a diagnosis. Not very long ago, there were only few ancillary tests available, and yet remarkable diagnostic accuracy could be achieved.

With the recent advances in ancillary tests (CT scans, MRI scans, ultrasound, specific laboratory tests including searching for specific autoantibodies and gene mutations) young neurologists in training may be mistaken to believe that the clinical examination has lost its importance. We do not endorse that view. Indeed, the indication and appropriate selection for many of these ancillary tests depends on a clinical working hypothesis.

The clinical skills of the neurological examination will be communicated to neurologists in training with this clinical guideline. It does not replace reading more extensive books on the market.

This text contains only few standardized and (semi-) quantitative test scores or scales. These test scores and scales are essential in clinical treatment trials and are helpful when following up patients under treatment. The readers are referred to the available books describing quantifiable clinical scores and scales. Only few quantitative tests are part of a standard neurological examination.

How to approach the patient

There are two ways of approaching a patient: one is to **selectively** address certain aspects of the physical exam that puts the differential diagnoses to a critical test which the examiner takes into consideration after taking the history. This approach has been named ***hypothesis-driven*** examination (Kamel et al., 2011). Many experienced and busy neurologists use this approach. The other way is a ***screen-***

ing exam which is **supposed** to be unbiased, but, of course, is somewhat more time consuming.

We think that the more experienced an examiner becomes the more selective and *hypothesis-driven* the examination may be. Students and young residents and in any unfamiliar clinical field even the more advanced neurologist should prefer the unbiased *screening approach*. Why do we propose that? The diagnostic view, particularly of the inexperienced, is skewed towards disorders one is familiar with. This may become a trap if individual findings are neither consistent with the precocious hypothesis nor are they a plausible "best fit" of the findings. It is our experience that much time can be saved and costly or even invasive procedures may be avoided if the diagnostic hypothesis is generated in a logical way following an unbiased exam.

Terminology, abbreviations, and basic information on bedside testing devices

In the neurological exam terminology is key to communicate within the medical field. Many terms are based on ancient Greek words, others on Latin. In some countries they have been replaced by descriptive terms in the native language. For example, take some of the descriptors of weakness (cf. section 4).

Paresis

Paraparesis

Tetraparesis

Plegia (para-/tetraplegia)

General examination in the Emergency Room

No neurologic exam should go without a general medical exam. The emphasis is on signs that are obvious disease signs, such as short-

ness of breath, cyanosis, cachexia etc., and provide a general qualifier as to the obvious severity of the condition.

Neurological examination

Note: Do not go through an in-depth exam if you note abnormal vital signs or signs of overt psychosis. Here, most prominent symptoms and signs come first allowing for a quick idea about the underlying condition and emergency measures that need to be taken.

SECTION 1

EMERGENCY ROOM EXAMINATION

Wakefulness, stages of reduced consciousness, and orientation

Check first for 1) **alertness** and 2) **consciousness**, 3) **orientation** to person, time, place, and to the specific situation, and **circumstances** for coming to the emergency room. You should appreciate the mental status as a **priority**, for if a patient cannot comply with the exam it will be incomplete. ***Note: Any patient with problems in communicating adequately needs special care.***

Another category is presented by an awake or mildly sleepy patient with **neuropsychological deficits:** a patient who does not speak (mutism) or speaks with an abnormal language (**aphasia**). If the patient is able to communicate verbally, the quality of speech production and understanding will become obvious (fluent, non-fluent, misusing nouns and syntax [**motor aphasia**]) or not understanding any spoken commands and look irritated (**sensory aphasia**); with normal speech production the verbal expression may be disturbed by an articulation disorder (dysarthria) reaching from nasal speech to toneless whispering. If the patient can understand the commands but performs them inaccurately, **apraxia** may be present.

Loss of self awareness and disorientation, illusions, delusions, anxiety, irritation and other **psychiatric signs** are different categories and need to be appreciated very early in the exam by simple conversation on disease related topics and **casual rather than for-**

mal speech asking for events of the current day, the way to the hospital, self-appreciation of the current condition.

Note: Formal testing is not recommended at this stage if there are no clear signs of abnormality. Patients may be annoyed and discouraged if they are tested in more detail for neuropsychological and psychiatric abnormalities at this early stage of the examination.

Once a patient presents with overt signs of **altered consciousness** (such as closed eyelids, apparent drowsiness up to coma) a number of simple steps can be taken to describe the pathology. The degree of pathology is graded in a categorical way or by using widely used simple scales such as the Glasgow Coma Scale or FOUR Score (see section 11 and list of references) and a description of the test results.

First, the patient is addressed by increasingly **loud spoken simple language**, by **touching** hand and cheeks, and if no reaction ensues **milder, then stronger,** and eventually even painful stimuli, but injurious stimuli must not be applied. The reaction to painful stimuli is very helpful in comatose patients: **the deeper the coma, the less likely the patient will react to painful stimuli.** There are a variety of simple non-injurious ways to elicit a painful stimulus:

mild to firm supraorbital notch (glabella) pressure, e.g. with the blunt end of the **reflex hammer**;

pinching of the skin and muscle at the upper edge of the trapezius muscle;

nail bed pressure exerted by the shaft of a **reflex hammer**.

bilateral pressure on the mastoid may be used when lesions of the upper cervical cord or lower brain stem are a possibility.

The reaction may be **withdrawal** of the pinched **limb only** - away from the pinch stimulus; or of **both arms** in severe supratentorial lesions; or a **paradox and bilateral stretch** (extensor) reaction following painful pressure to **only one** limb in a severe brain-

stem lesion. With thoracic transverse **myelopathy** a strong pinch to the skin and trapezius muscle may cause ipsilateral bending of the elbows while the legs may stretch out or bend in the hip and knee joints. In general, nail bed pressure is less traumatic than pinching. If sensation is focally lost, e.g. in peripheral nerve trauma as part of a multifocal injury there will be focal non-responsiveness to nail bed pressure. With spinal cord or brainstem damage the response will be bilateral or even unilateral extension of the limb instead of withdrawal.

Hyperirritability of meninges (Meningismus)

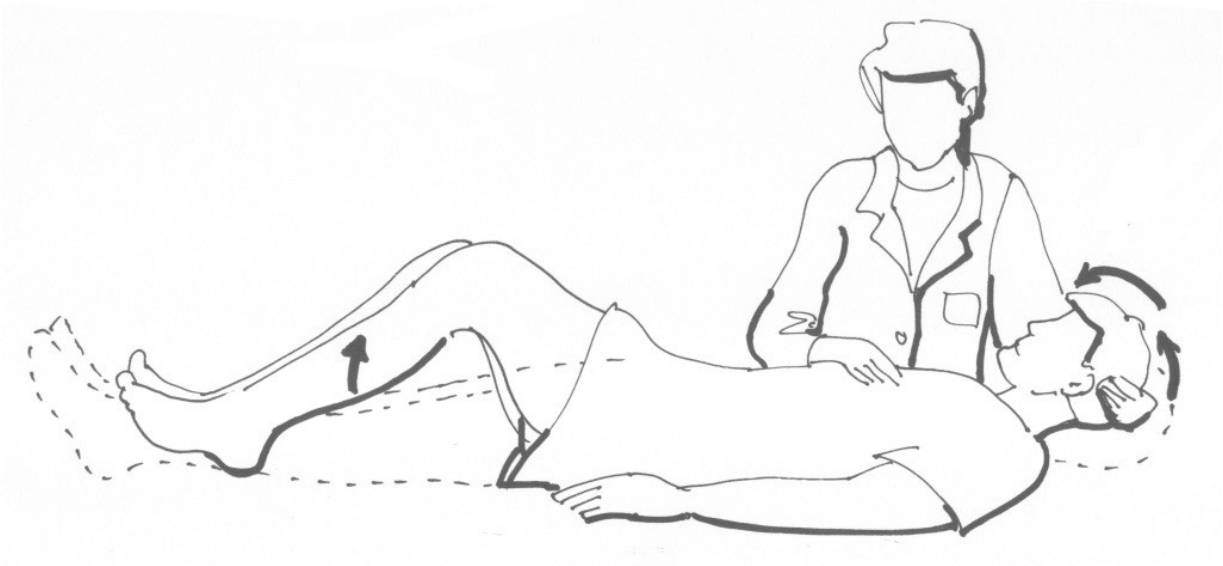
Note: this should be done in awake and in unconscious patients alike. Deeply comatose patients and elderly patients with meningeal pathology may not show meningism.

Meningeal irritation (meningismus) is best tested by observing spontaneous head and neck movements. In meningeal disease spontaneous movements are scarce. Afflicted patient **resists all passive movements** that will put traction on the irritated spinal meninges.

Formal testing is done by **gently** testing passive movements of the head in all directions while the patient is in supine position. When the head is bended to flexion, there may be resistance to further flexion (nuchal **rigidity**) and/or the patient's legs may flex at the hips and knees bilaterally, especially in acute meningismus (Brudszinski's sign). Kernig's sign is positive when the examiner induces back pain by trying to extend the lower leg while further extension of the leg is painful in a patient with the thigh thigh and the knee are both bent at an angle of 90°. Lasègue's sign tests painful resistance to passively flexing a stretched leg in the hip. The Kernig and Brudzinski signs have a combined **sensitivity of 5% and specificity of 95%** (Thomas et al, 2002) meaning that any resistance to passive neck and leg elevation are very specific but unfortunately

not sensitive in adult clinical practice. Therefore the differential diagnoses are important to keep in mind.

FIGURE 1.1.1 The Brudzinski's and Kernig's signs



in (a) the examiner lifts the head of the patient and feels a resistance in the neck muscles. The awake patient reports neck pain and as a secondary reaction both legs may go up (Brudzinski's neck sign);

(b) Kernig's sign is positive, when the examiner induces back pain by trying to extend the lower leg while the thigh and the knee are both bent at angle of 90°.

Note: In a stuporous (sleepy) or in a mildly comatose patient the mimics and other indicators of pain should be looked for.

Acute root compression by a cervical slipped disk or a recent whiplash injury may mimic meningism in the neck region but not in the lumbar region. Lasègue's manoeuvre is also positive with lumbar root compression, typically unilateral.

SECTION 2 – CRANIAL NERVES (CN)

General Terminology

Anosmia and Hyposmia

Miosis

Mydriasis

Anisocoria

OU/OS/OD

Papilledema

Anop(s)ia

Argyll-Robertson pupil

Adie pupil

Horner syndrome

Exo/eso/hyper/hypo-tropia

Exo/eso-phoria

Ophthalmoplegia

Gaze paralysis

Version

Duction and Torsion

Vergence

Nystagmus

Vertigo

Dysequilibirum

Myotonia

Anarthria and **Dysarthria**

See also Motor System (section 3), Reflexes (section 4), and Sensory System (section 5)

Olfactory nerve (CN I)

Smell is rarely tested in detail during a routine neurological examination. It can be tested, however, by offering aromatic compounds (**coffee, cinnamon, vanilla**) to one nostril by closing the other one gently. An anosmic patient is usually able to detect irritating compounds. **Hyposmia** is a common, early sign in degenerative CNS disorders, e.g. in Parkinson's disease.

Note: Test only in a fully awake and cooperative patient.

Optic nerve (CN II)

Visual acuity

(Test only in a fully awake and cooperative patient)

First, have the patient read your name tag in each eye individually. Then printed text can be tested with using reading glasses. For quantitative testing of visual acuity (VA) take a **standardized table** for near vision and distant vision, again allowing near and far glasses. Dimensions are in fractions or decimal figures (20/20 or 1.0 refer to normal values, respectively).

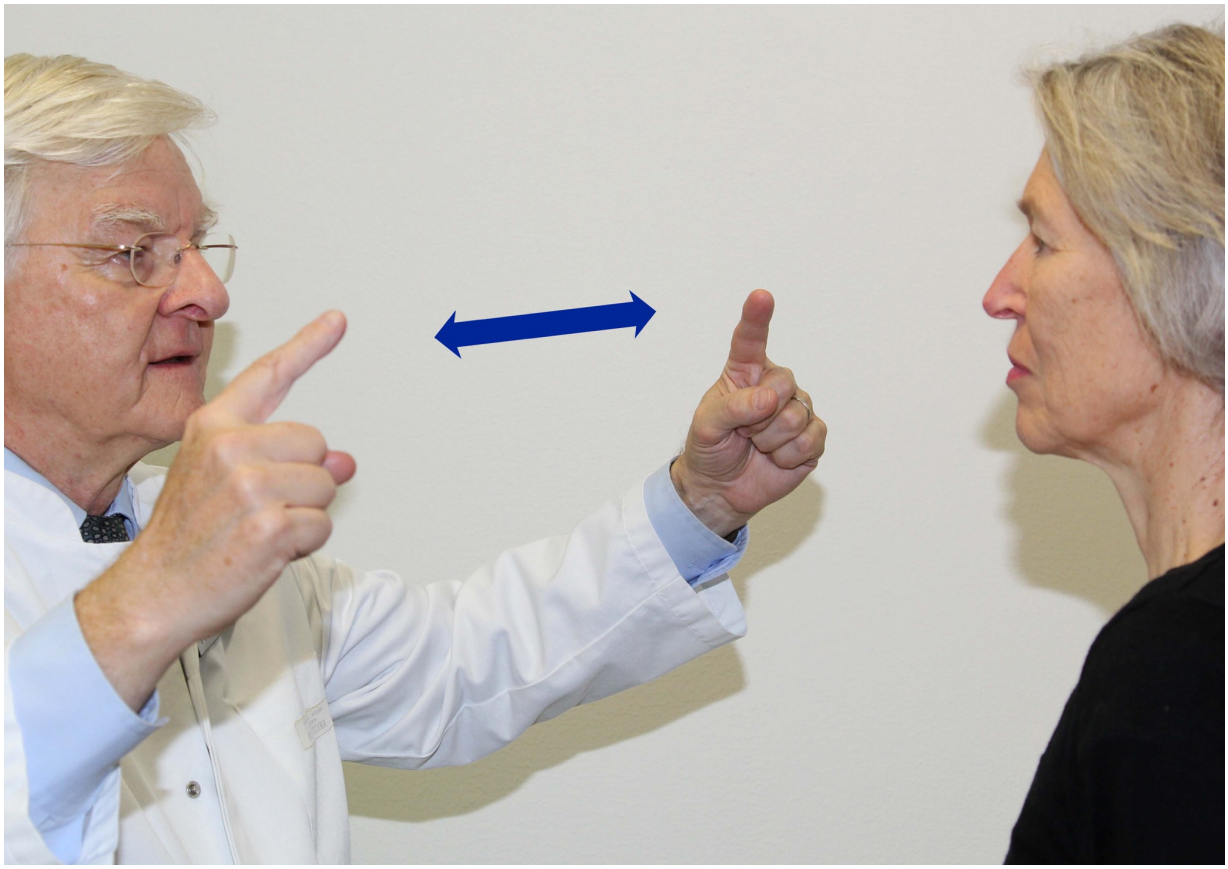
Visual Fields

Defects in visual fields include **homonymous hemianop(s)ia** (corresponding half fields afflicted in both eyes; e.g. patterns # 4, 6, and 7 in Figure 2.2.2), which indicates a retro- chiasmatic lesion of the visual pathway, or **upper/lower quadrantanop(s)ia** (one quadrant afflicted on both eyes; e.g. pattern 5 in Figure 2.2.2) found in lesions of lower/upper optic radiations, or **heteronymous hemianop(s)ia** (a different hemifield affected in both eyes (e.g. pattern 3 in Figure 2.2.2), which points to pathology in or near the optic chiasm.

For testing, sit or stand about **1 meter** away from and opposite to your patient and have him/her look at (fixate) your nose consistently. Stretch out your arms sideways in a horizontal plane **half way between you and the patient** to a position where you can still see your fingers well (Figure 2.2.1, Figure 2.2.3 and Movie 2.2.1). Then move your fingers slowly on one side and have the patient name the side of movement or point to it. Most patients will change fixation towards the hand that is moving, which tells you that **something has been perceived**. Ask the patient to again fixate your nose and try again. For a more demanding task, ask the patient

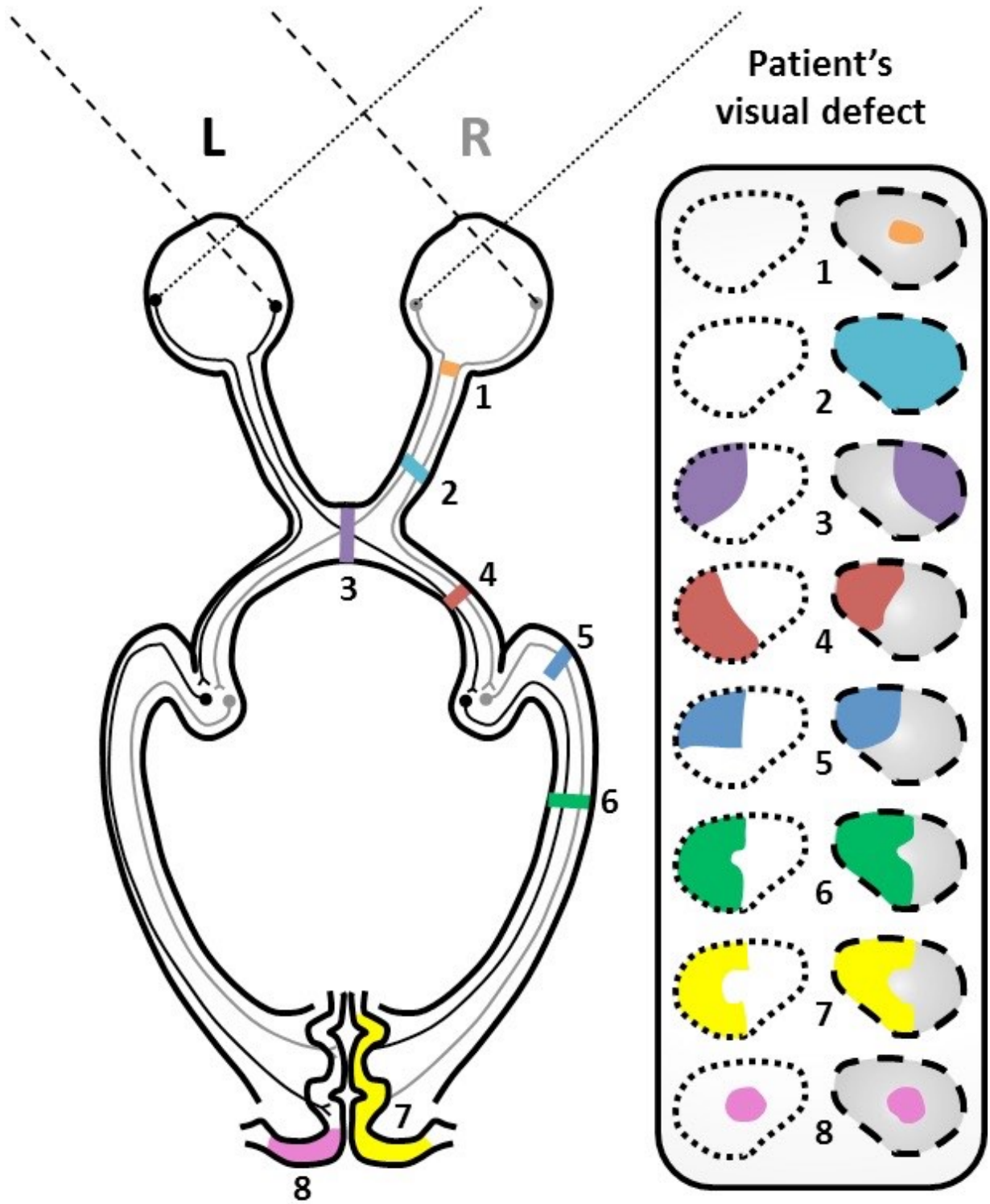
to count the numbers of fingers (only validated with 1, 2, or 5 fingers) presented by the examiner on either side. A healthy examiner should see all this (he/she is is the **control**). Then the arms are moved **down to test the lower** right and left quadrant fields and **up for the upper** quadrants.

FIGURE 2.2.1 Finger perimetric testing



The fingers are moved out until the patient cannot further see them on either side.

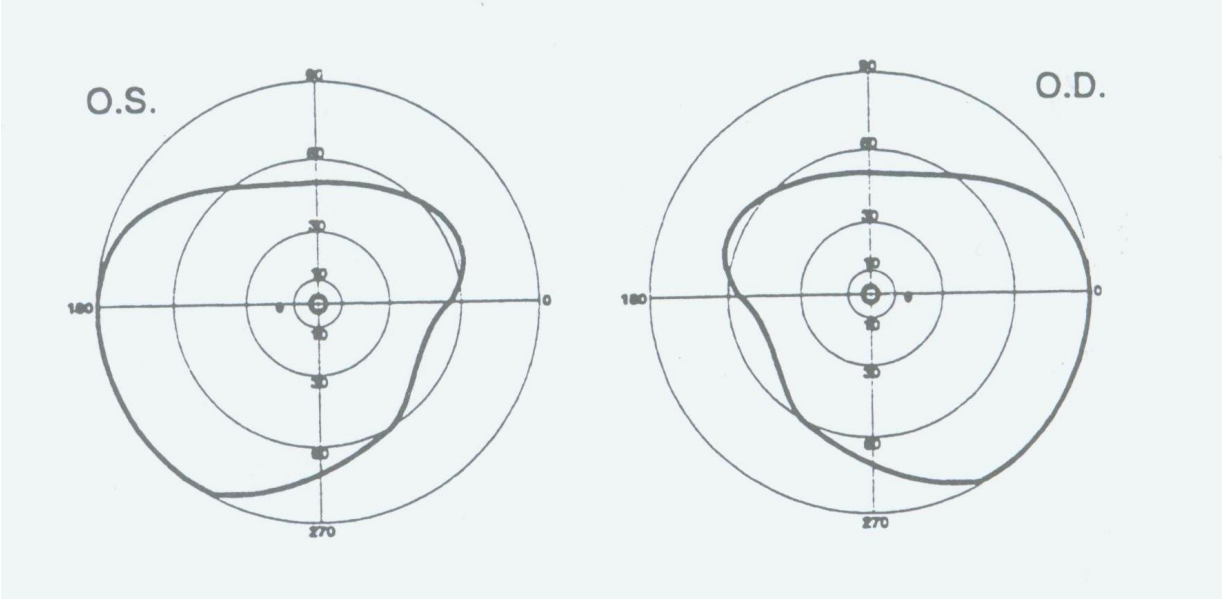
FIGURE 2.2.2 Typical patterns of visual field defects and associated lesion sites. Colours represent the defective fields and the associated sites of lesion.



MOVIE 2.2.1 Finger perimetric testing

Finally, the findings are documented in a standard scheme and archived in the chart (Figure 2.2.3).

FIGURE 2.2.3 Visual fields (normal)



Left (O.S.) and right (O.D.) eyes are shown with the normal visual fields as the patient sees it. This is outlined as a continuous line, as also shown in Fig. 2.2.2. The abnormal borders indicating a field defect are sketched with a marker.

Monocular testing: In any case of abnormality, or if the problem is thought to be more subtle, each eye should be tested individually in order to more precisely denote the degree of a field defect. The patient is asked to hold his palm in front of one eye at a time and the examiner closes his/her ipsilateral eyelid. A **red dot or a bright colour pen** is moved from the outside towards the central visual axis in all 4 orientations (horizontal, vertical, diagonal). The patient tells the examiner at what point the moving object is seen; again the examiner is the control.

Note: Proper perimetry by state-of-the-art automatic perimeters is essential if any type of defect has been identified at the bedside.

Visual neglect means reduced attention to a visual stimulus presented in a hemi-field – It is often confounded with homonymous hemianopia and may be associated for other types of neglect (**hemineglect**, see Section 8). It can be detected by simultaneous finger movements in both visual hemifields. It can only be tested if visual fields are largely within normal limits.

Extinction refers to a form of neglect when the patient is able to perceive unilateral stimuli on either side without problems, but cannot if stimuli are presented simultaneously.

Color vision can quickly be tested by asking for eye-to-eye comparisons of brightly coloured objects (such as coloured items in the room). Formal testing can be performed by using **Ishihara plates** in either the book form or as an app on the phone or tablet computer.

Fundoscopy is an essential part of the neurological examination. Neurologists prefer a **direct ophthalmoscope** (cf. Figure 2.2.5). Pharmacologic dilatation of the pupil is **avoided** because a paralyzed pupil **may mask** intracranial disease progression. In a **darkened room** the pupil will dilate and allow inspection with a low light emission with good contrast. For vessel inspection the built-in **green** filter may be used. Ideally, the **right eye** is looked at with the **examiner's right eye and vice versa.**

At first, the ophthalmoscope light is used from a distance of about 45 cm at a lateral angle of 15°. The pupil will contract and the fundus will light up in red. Then the examiner approaches the patient's eye and aims at getting the fundus in focus. The patient is asked to fixate on a **distant** point while the fundus is inspected with the lens dial adjusted at 0 diopters initially. If the refraction error in a myopic or hyperopic patient is known the dial is preadjusted to – or + diopters: a + 5 lens of the patient's glasses would be compensated by positioning the dial at -5.

FIGURE 2.2.4 Fundoscopic images of the retina and arachnoidea in intracranial hypertension

Image 0: Left eye with normal fundus; **Image 1:** Right eye with mild papilledema, with a more pronounced crescent of blurring of the papillary rims nasally, marked venous congestion and curly ves-

sels. **Image 2:** Left eye with moderate papilledema due to intracranial hypertension, with a more circumferential blurring of the papillary rim, with mild venous congestion. **Image 3:** Left eye with more pronounced circumferential papillary blurring and discontinuity of some, but not all, vessels as they pass over the papillary margin. **Image 4:** Left eye with marked papilledema with blurred rims, multivessel discontinuity, linear hemorrhagic areas, and exudates. **Image 5:** Right eye with a broad-based papilledema, massive papillary swelling with disc protrusion, obscuration of all major vessels on the optic disc, and peripapillary hemorrhages from obstructed vessels.

The examination includes the papilla (papilledema; pale optic disc), the main arteries (signs of hypertension; changes in calibre; cotton wool or blood deposits) and the major veins at the margin of the papilla (venous pulsations; congestion).

Note: The fundoscopic exam is considered a mandatory part of the complete neurological exam.

Formal examination of the pupil: The **shape and the size** (diameter in mm) of the pupil are recorded bilaterally at normal room light using a pupil size chart. Then a torchlight is used to shine only on one pupil and the contraction of both pupils is recorded; this is repeated by shining on the other pupil. This contralateral effect of constriction is called the **consensual light reaction**. Its absence is abnormal and may indicate a brainstem lesion at the pathway from the lateral geniculate to the tectum (parasympathetic nuclei). The ipsilateral reflex loss indicates a lesion of the optic nerve or tract.

With very small pupils (e.g. miosis in an unconscious patient) it is helpful to use the direct ophthalmoscope. The light beam is moved from the side to the pupil while looking through the **+ 15 or + 20** diopter lens to better recognize a minute constriction (Figure 2.2.5 and Movie 2.2.2).

FIGURE 2.2.5 Examination of very small pupils with an ophthalmoscope

MOVIE 2.2.2 Examination of very small pupils with an ophthalmoscope

For the evaluation of a lesion of the optic nerve or the retina, e.g. optic neuritis, the swinging flashlight test should always be done. In a darkened room the light is shining in one eye and the constriction of the pupils is recorded and if normal is expected to be the same on either eye. Then the light is directed to the contralateral eye and again the constriction is recorded. In a unilateral lesion, swinging

the flashlight to the affected eye will result in an apparent dilation of the contralateral pupil indicating an **afferent** pupillary defect ("Marcus Gunn pupil").

Note: these changes may be rather subtle, like a 1-2 mm side difference. A common trivial cause of abnormal pupillary diameter is through application of constricting (glaucoma) or dilating eye drops. Patients should be <u>asked</u> specifically if either situation applies.

Oculomotor nerves (CN III - oculomotor n.; CN IV - trochlear n.; CN VI - abducens n.)

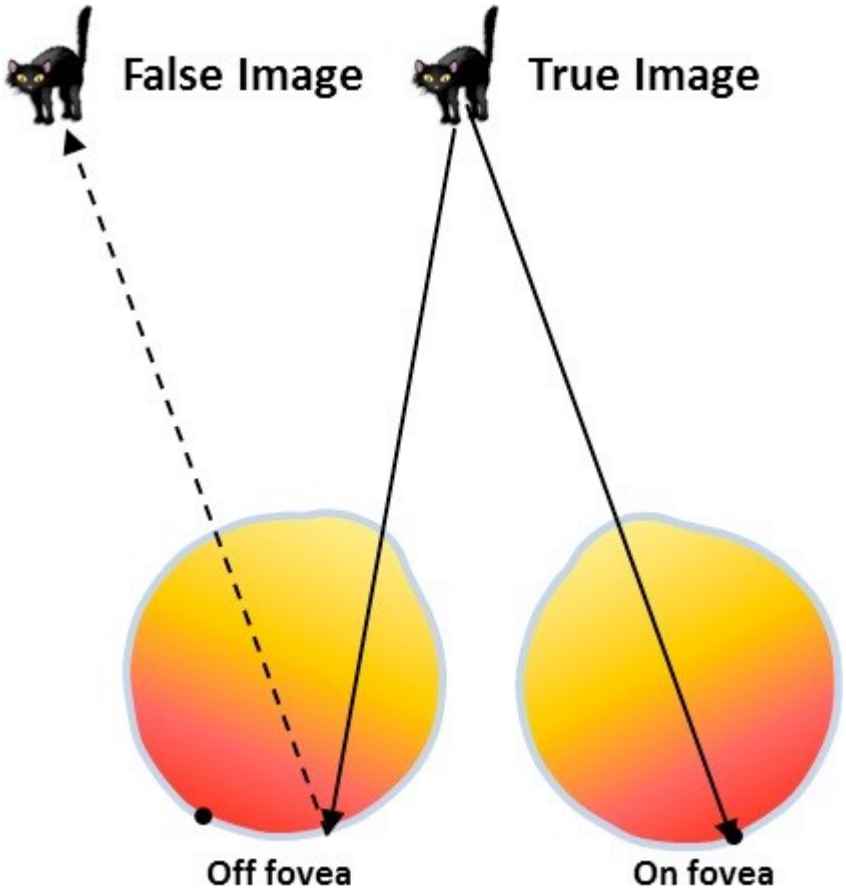
First, the eyes are inspected. Is there a squint (**heterotropia**)? Does the patient show a **head tilt**? Note that a head tilt can be compensatory, such as in trochlear nerve palsy. The **ocular tilt reaction** is the triad of i) head-tilt ii) skew deviation (lower eye on the side to which the head is tilted), iii) cyclotorsion of the eyes (toward the side of the lower eye, only seen with fundoscopy). It may occur with **central and peripheral** vestibular lesions.

Normally, the axes of both eyes are aligned in **primary position** and keep aligned on lateral and vertical gaze. Hence, any diversion from this binocular paradigm is named a specific type of **heterotropia** (eso- = in, exo- = out, hypo- = down, hyper- = up) or strabismus and is always **abnormal**. As a consequence, in any form of recent extraocular muscle palsy the patient will report double vision (**diplopia**). With longstanding palsies causing obvious squint, the "wrong" image may be suppressed (**excluded**) by the brain within days to a few weeks. Be sure to assess the **type of diplopia** (vertical, oblique, horizontal, bigger/smaller) and eye movements that worsen or improve the distance of the double images (i.e. does looking on one particular direction worsen or improve the double vision?)

Diplopia (Figure 2.3.1) can be subtle and may occur only in certain situations. In latent eye deviations (called **heterophoria**), the eye deviation is compensated while fixating an object and occurs only if binocular vision is interrupted. This is done by covering one eye and then rapidly uncovering it ("cover-uncover test"). The affected eye will deviate when the normal eye is covered. Heterophoria (eso, exo, hypo, hyper-- see above) does not necessarily indicate

nerve pathology It often decompensates when patients are tired or have perturbed sleep, or are under alcohol or drugs. Here it is also important to note up-down deviations of the eyes, as this **"skew"** is suggestive of **central** pathology.

FIGURE 2.3.1 An example of the false image in diplopia being represented as farthest from midline in lateralization of gaze



Notice that with left lateral gaze in left lateral rectus palsy, coverage of the impaired (left) eye would make the more lateral (false) image disappear. Note that the "on fovea "and "off fovea" images don't allow for cortical fusion, as the cortex tries to fuse images in the same relative position on the retina.

Another simple way to note misalignment of the eyes is through the **reflection** of the testing flashlight (or any other room light) on the corneal surface (Hirschberg's test). In the normal situation, the mirrored images should be at the same relative position on either eye independent of eye position. In Figure 2.3.2 a clear deviation is obvious and also illustrates two important facts: (1) the light reflection is off center in the non-fixating eye and (2) the de-

gree of diplopia (as seen by the patient) and heterotropia (as seen by the examiner) is more pronounced when the paretic eye is fixating.

FIGURE 2.3.2 Right superior oblique muscle underaction (paresis)

Upper image: the non-paretic left eye fixates and the right deviates upwards while the right light point is off center downwards. Lower image: the paretic right eye fixates and the left deviates down (inhibition of contralateral inferior rectus) while the left light point has moved up.

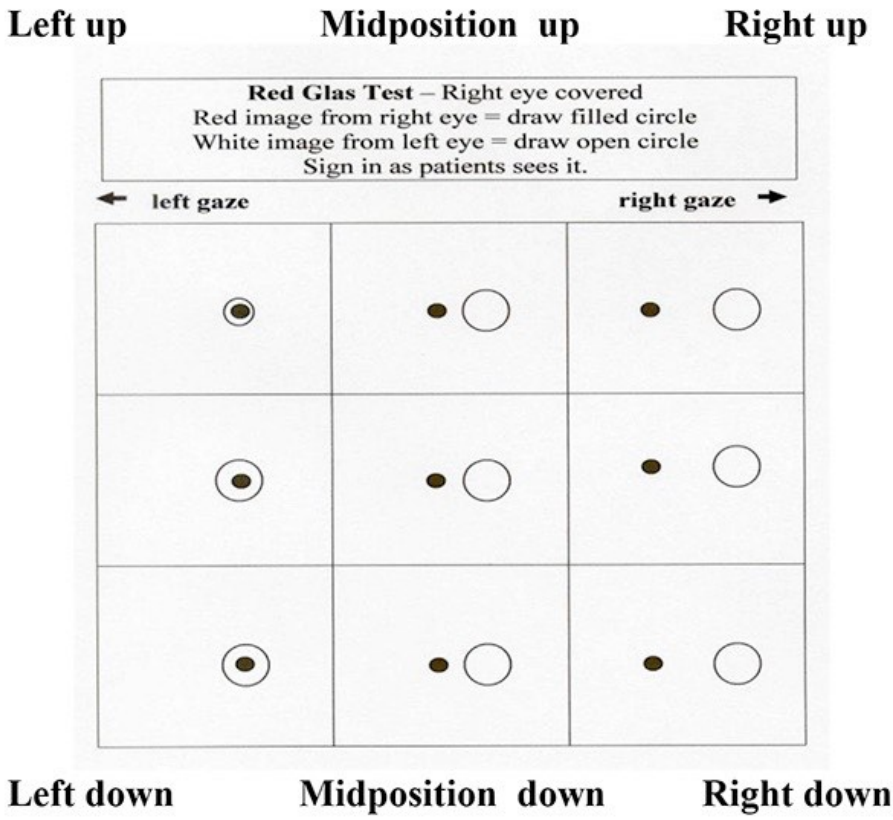
As soon as diplopia is reported and deviations become obvious during these tests a **fatigue test** should be done to look for ptosis and to test for variable or fatiguing extraocular muscle weakness indicating a neuromuscular **transmission disorder** (e.g. **myasthenia, see** Figure 2.3.4; Movie 2.3.2).

Once there is any type of abnormality, with or without reported double vision, further neuroophthalmologic examination is required. Consulting a more detailed text of eye movement pathology is recommended.

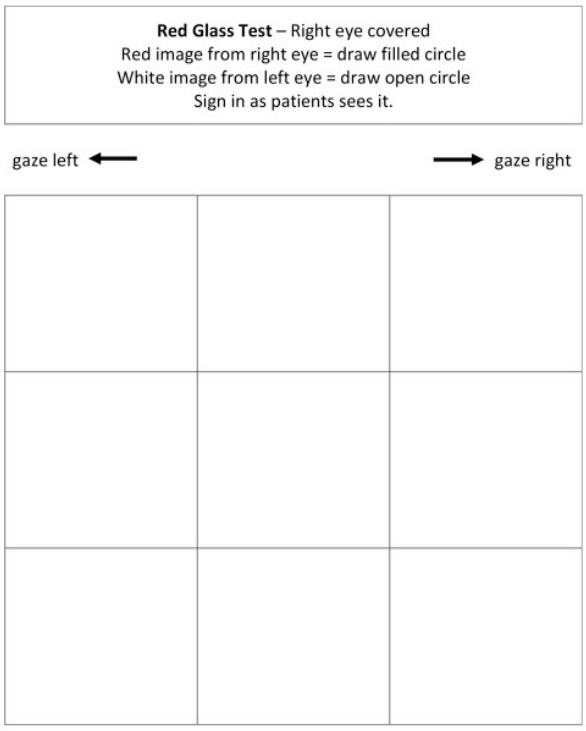
A **red-glass test** (Figure 2.3.3 and Movie 2.3.1) is a simple bedside test to document double vision. The examiner asks the patient to fixate a hand-held small flashlight with a red glass held before the **right** eye to start with. The red glass will allow the patient to easily tell the two images apart. The patient is asked to direct the gaze in 9 different fields (see diagram) and draw the perceived images into the diagram. Following Hering's rule that the distance between the two images is greatest in the direction to which the weak eye muscle(s) should be pulling the eye makes us understand the principles of double image deviations.

FIGURE 2.3.3 Red-glass test for double vision

In the standard test the red glass covers the right eye.



*Documentation of eye muscle weakness with double vision. Simple drawing of a **red-glass** test in a patient with a **left medial rectus underaction**. The red glass is in front of the **right** eye. The closed circle denotes the red image, the open circle the white image as seen by the patient. **Note:** Left and Right as the patient sees it.*



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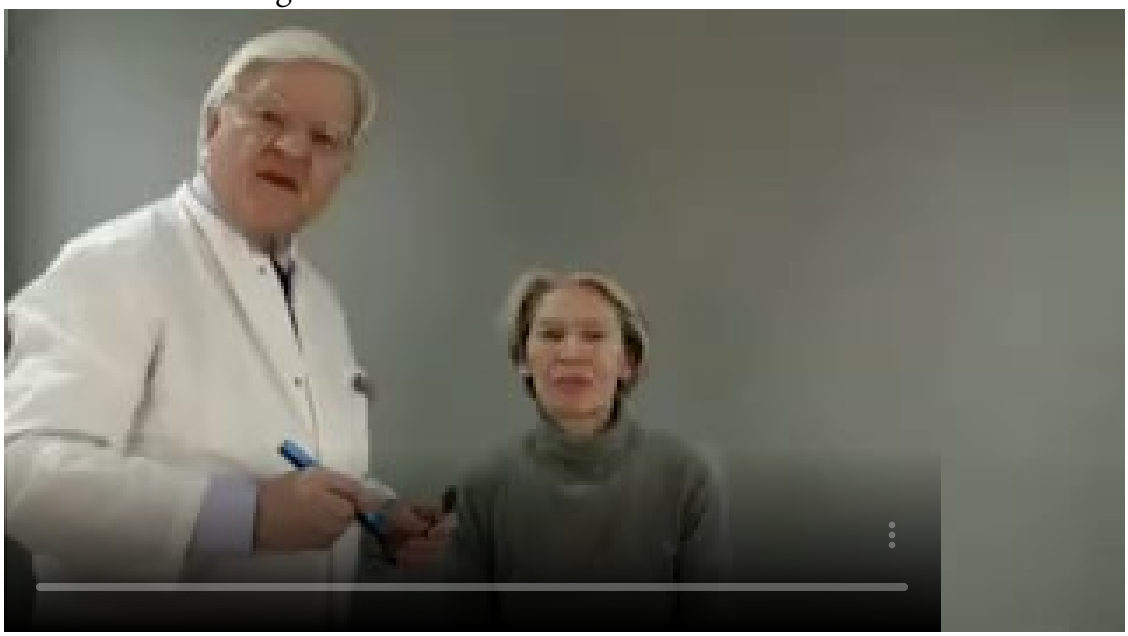
MOVIE 2.3.1 Red-glass test



When the red glass is moved to the **left** eye the distances between the two images may be the same, smaller, or larger. Following the rule that in monocular muscle palsy, double vision has a **larger an-**

gle of deviation when the **afflicted** eye is fixated, this right-left change may tell which eye has the muscle weakness.

Note, that usually, the eye with the red glass in front is the fixating one.

Moreover, rapidly changing distances of the double images or divergent results observed at serial tests help to diagnose a **neuromuscular transmission disorder** (see below). If you don't have a red glass or a pair of used green/red 3D glasses, then just covering up an eye to see which image goes away will help to diagnose which eye is impaired: the image that is **furthest** from the middle is the false image, if this one **disappears** when the eye is covered, you have just covered the impaired eye!

In **chronic** cases of ocular muscle palsies this **may not** be reported despite an obvious squint due to central **suppression** (exclusion) of vision in one eye.

Second, the eyelids are inspected. A spontaneous **unilateral ptosis** (hanging eyelid) is easy to detect and is rarely the result of a central/brainstem CN III lesion (as the nucleus for eyelid opening has bilateral innervation). This is typically associated with **wrinkling of the forehead** because the patient tries to overcome lid weakness and this may appear as a contralateral widening of the palpebral fissure. In addition, with severe ptosis, the patient may **hyperextend** the neck (bend head backwards, "astrologist's posture"). If ptosis is mild, it can be part of **Horner's syndrome**, due to malfunction of the sympathetic innervation of Müller's muscle (see section 7). This is usually unilateral and associated with a slightly smaller pupil(**miosis**). Aquired ptosis may have **many different** underlying pathologies.

Before any invasive work-up is initiated, a **disorder of neuromuscular transmission** should be formally ruled out (e.g. myasthenia gravis): the ptosis is fluctuating, often asymmetrical, and can be

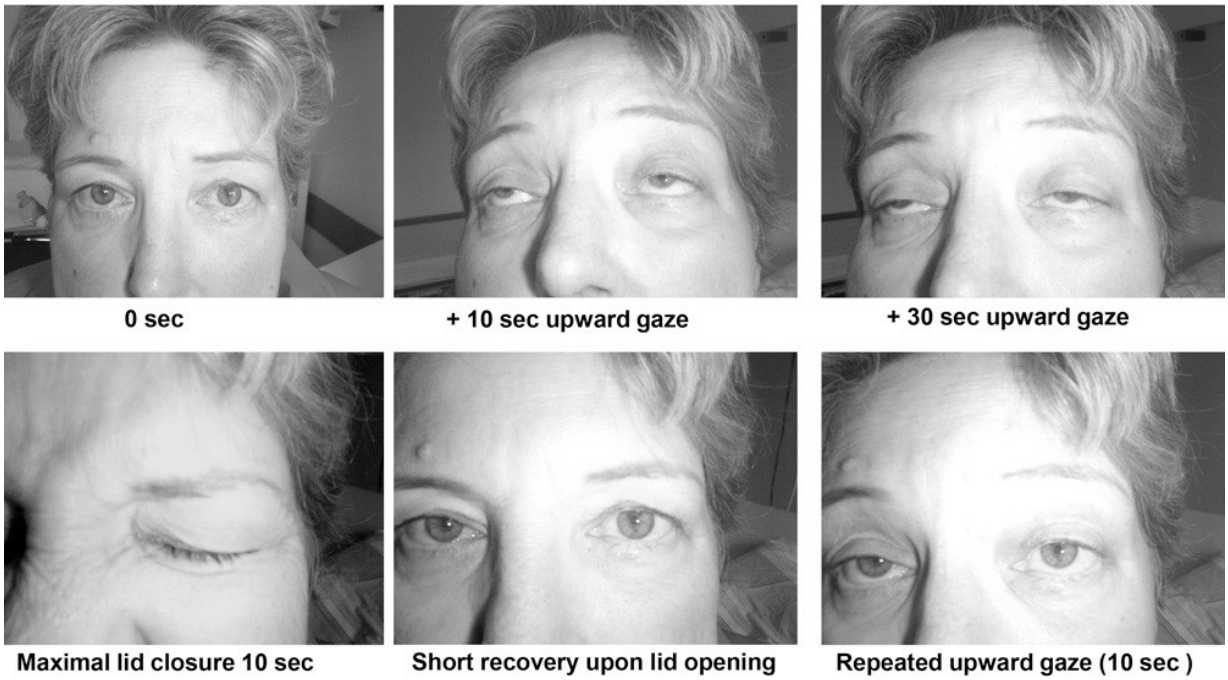
profound. A fatigue test is done by sustained upward gaze for up to 120 seconds. If ptosis stays more or less the same it is likely caused by a partial or complete CN III palsy. If it gets much worse and improves on subsequent squeezing of the eyes it is more likely to be due to a neuromuscular transmission defect (Toyka, 2006, Movie 2.3.2, Figure 2.3.4). After doing this, the eyelid may be **twitching** a few times on upward gaze probably indicating that fast twitch muscle fibers of the levator palpebrae muscle receive overshooting nerve impulses (**Cogan's lid twitch sign**). When doing the fatigue-recovery test, often other afflicted eye muscles are also fatiguing with the patient noting new or exaggerated double vision.

MOVIE 2.3.2 Fatigue & Recovery Test in real time

Patient with generalized myasthenia, 4 weeks after starting immunomodulatory treatment. No spontaneous ptosis. Slowly developing ptosis upon upward gaze and short recovery after maximal

eye closure associated with a two overshooting phasic lid elevations (equivalent to Cogan's lid twitch sign)

FIGURE 2.3.4 Fatigue and Recovery Test

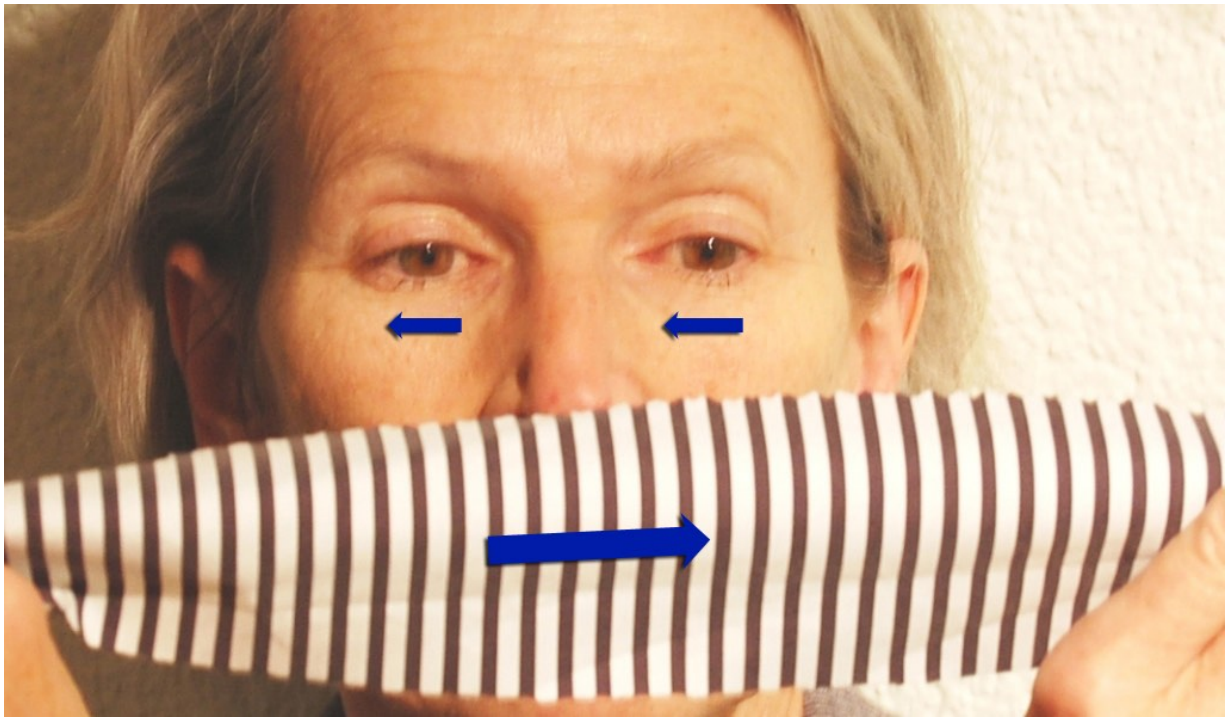


Upon upward gaze the ptosis progresses and recovers for a short time upon lid closure followed by progressive ptosis upon repeated upward gaze. Times are indicated in seconds.

Optokinetic nystagmus (OKN) is a normal phenomenon elicited when the patient intends to maintain moving objects in focus, i.e. on the fovea of the retina; it is normally observed if one looks at the "outside world" (e.g. out of a car) while moving along. Although it is properly tested only with whole-field stimulation, it can be tested at the bedside with a turning drum or just with a long stripe of fabric that has vertical black and white stripes on it (see Figure 2.3.5). In a lit room, the drum or the fabric is held in front of the patient, approximately 40 cm away, and is turned moderately slowly (1 turn per second) to either side or up and down. The fast phases (saccades of the **nystagmus**) are against the direction of the rotation. Any type of side-to-side differences, absence, or distortion of the OKN is abnormal. The downbeat OKN is often somewhat less pronounced than the other directions. OKN is lost or markedly altered

in brain stem disorders and is mildly asymmetric in hemispheric disease, e.g. stroke.

FIGURE 2.3.5 Testing for optokinetic nystagmus



A fabric or tape with dark stripes can be used. The fabric is placed in front of the patient and the patient is asked to fixate the stripes. Then the fabric is slowly moved from right to left (1-2 seconds for 50 cm of fabric, arrow) and a horizontal nystagmus to the right side (arrows) is observed. The direction of moving the fabric is reversed and finally changed to up and down for vertical nystagmus. Any side difference is a pathologic finding.

Third, **smooth pursuit** eye movements are explored with a flashlight or the examiner's fingers at a distance of about 50 cm. First, convergence is tested by having the patient fixate the approaching fingers of the examiner. There is an associated constriction of the pupils (miosis). The full extent of slow eye movements is tested in **horizontal, vertical, and diagonal** planes. The eye movements are observed and recorded. In case of any abnormality of the optical axis the patient will report double vision. There may also be a loss of smoothness with saccades interspersed (choppy, rapid eye movements), which usually is a sign of pathology except in the elderly.

Next is the search for a possible **nystagmus** or other abnormal movements of the eyes. The reader is referred to a detailed description in the textbook of Leigh and Zee (2006). When spontaneous **nystagmus** is present the patient may or may not report dizziness and **vertigo** depending on the underlying disorder. In acquired forms of **nystagmus** visual fixation will suppress its extent whereas preventing fixation with + 13-15 diopter magnifying lense while **covering one eye** or with Frenzel lenses (Figure 2.3.6 and Movie 2.3.3). A "poor man's" way of suppressing fixation is occlusive fundoscopy, in which the contralateral eye is covered while performing fundoscopy; you would expect to see the optic disc drifting toward the affected eye and then saccading away (the opposite of what the pupil will be doing). An even easier method is to gently hold the **examiner's fingers** on the closed eyelids and feel the **nystagmus** with the patient's eyes closed.

FIGURE 2.3.6 Testing for nystagmus and abnormal eye movements

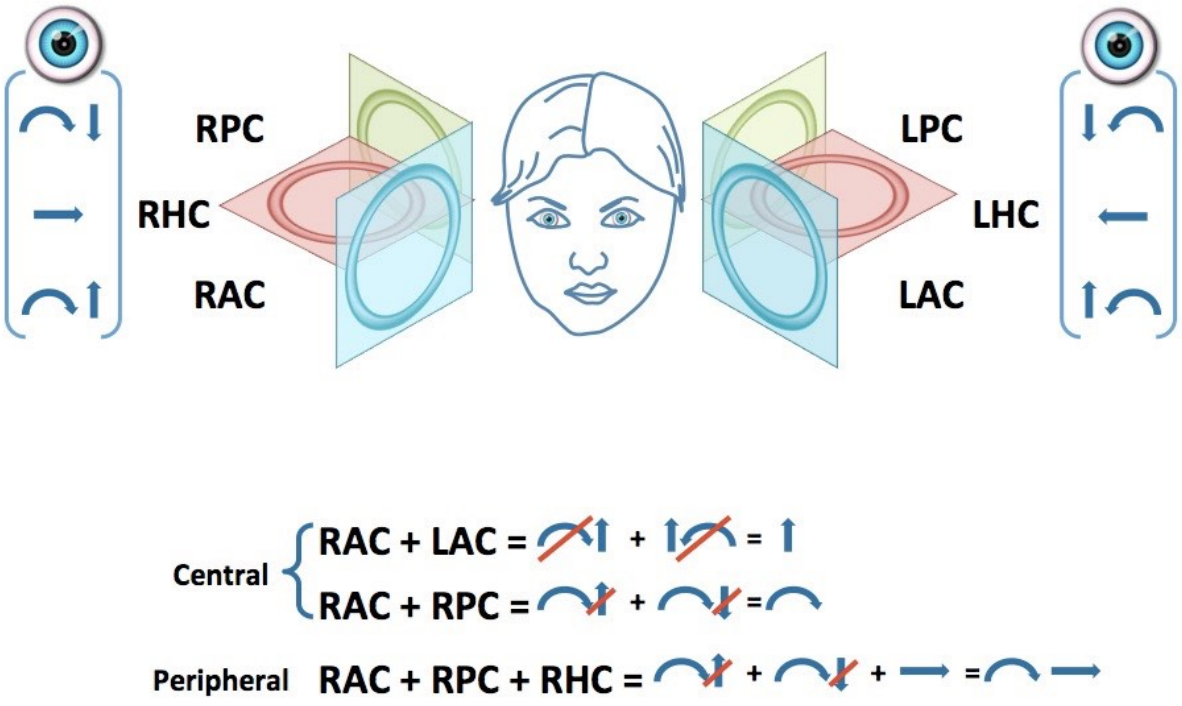
A modern version of the original lenses by Hermann Frenzel, an ENT specialist at the University of Göttingen (died 1967). Any abnormal eye movement can be identified by simple observation.

MOVIE 2.3.3 Rotational nystagmus (normal)

During examination of smooth pursuit eye movements the amplitude of the nystagmus is enhanced if gaze is directed towards the **fast phase** of the nystagmus, a feature more suggestive of peripheral lesions (known as Alexander´s Law). Some patients show no nystagmus in primary position but increasingly so on lateral or - less commonly - on vertical gaze. This is called **gaze-evoked** nystagmus, which may also have a rotational - clockwise or counterclockwise – component. Rotational nystagmus is most easily assessed by observing the movement of the scleral vessels. The important features of nystagmus to note are the **direction**, relative velocity of the fast and slow component, and changes with direction of gaze.

A standard test for vestibular disease is the **irrigation of either ear** with warm and cool water - provided that damage to the ear drum has been **excluded** by inspection of the ear with an otoscope. Findings are shown in Figure 2.3.7.

FIGURE 2.3.7 Oculomotor effects of stimulating SCC with the arrows indicating slow phases from the observer's perspective



Abbreviations: SCC - semicircular canal, PC – posterior canal, HC – horizontal canal, AC – anterior canal, R – right, L – left.

To use this figure: 1. Find the eye movement observed in the patient to figure out which canal is stimulated or 2. Predict what eye movements would happen based on stimulation/inhibition of a given set of canals.

This figure is a view on the face of the patient with the eye movements flanking the canal that causes them. The consequence of stimulation (warm water caloric, otoliths, etc.) results in a slow phase rotation (from the observer's perspective) in the direction indicated by the arrows under the eyeball with a quick saccade in the opposite direction (ex. stimulate the LHC and you will see the slow drift of the eyes to the patient's right and a corrective saccade back to the left). Notice that pure vertical and pure torsional nystagmus always indicates a central pathology because stimulation of just two of the semicircular canals at a time is all but impossible suggesting that the lesion is not peripheral but rather central. Conversely, a peripheral pathology (which knocks out an entire triad of canals or

may affect just one canal – most often the posterior canal in BPPV) usually has combined torsional-lateral nystagmus.

If no nystagmus is present yet the patient complains of rotational vertigo, a **provocation test** is useful. First, the patient is asked to shake the head vigorously from right to left in the horizontal plane for 30 seconds with eyes closed. Then with the 13 Diopter lens in place, provocation nystagmus can be seen. If this does not produce nystgamus the formal **„head shaking test"** is done where the examiner rotates the patient's head (eyes closed) laterally back and forth for 20-30 seconds or 10 right-left rotations. A nystagmus observed after this manoeuvre with or without the 13 Diopter lens in place is always pathological and usually due to an inner ear problem. If needed, a **caloric test** can be performed, also at bedside in comatose patients, after verifying with the otoscope that the tympanic membrane is intact. Instillation of 37°C water in one ear (stimulating the horizontal semicircular canal) induces a horizontal nystagmus physiologically beating to the same side, while after cold water the nystagmus beats to the opposite side. A simple mnemonic to remember the direction of the rapid phase and hence of the nystagmus is COWS – **C**old **O**pposite, **W**arm **S**ame.

Note: 3-4 beats of fading, laterally directed nystagmus may be normal, as occurs not infrequently in the elderly, while up/down beat nystagmus is never normal

The **vestibuloocular reflex (VOR)** is a reflex receiving its input from the vestibular organs and nuclei. In daily life it is suppressed by fixation. Suppression of the VOR is impaired in some disorders of the CNS, in particular of the cerebellum. The patient is asked to stretch out his/her arms (thumbs up) and fixate on the thumbs while smoothly turning the trunk and maintaining the head position relative to the trunk – like a robot or mannequin (Figure 2.3.8 and Movie 2.3.4). If a deviation of the eyes **against** the direction of the turns is observed or if a nystagmus with the fast phase

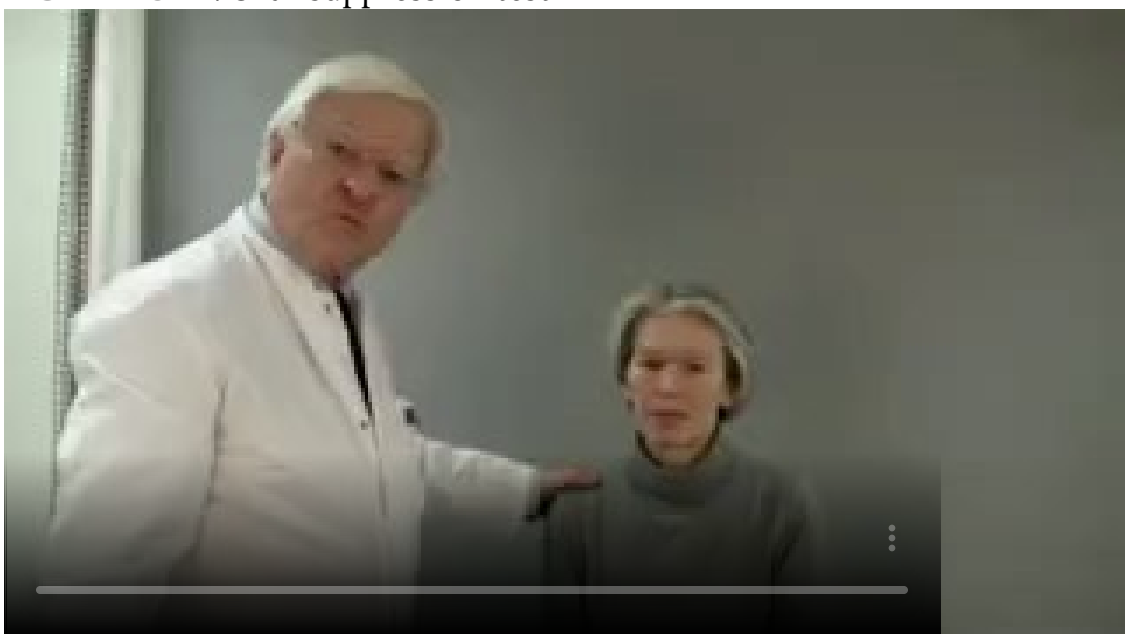
in the **direction of the turns** is seen the VOR suppression is lost. Its functional impact can also be verified by performing the same manoeuvre while having the patient hold and read from a book page or the Snellen eye chart instead of look at his/her thumbs. If more than a line of text or visual acuity is lost, then there may be a problem with the VOR.

FIGURE 2.3.8 VOR - suppression test

Note that the thumb is being fixated while the patient turns his/her trunk or the trunk is turned by the examiner at different speed. (a) primary position;

(b) turning right.

MOVIE 2.3.4 VOR - suppression test



Another test to examine the VOR is named the **head impulse test** (Halmagy-Curthoys): The patient is asked to fixate on the examiner's nose. Then the head is moved swiftly to one side by approximately 30 deg. If the eyes do not stay fixated on the target and corrective saccades toward the examiner's nose are noted, peripheral vestibular disease related to afferents from the horizontal semicircular canal is likely. The test is performed by turning alternatively the head to both sides. A video demonstrating proper execution of the head-impulse test can be seen at http://jnnp.bmj.com/content/78/10/1113/suppl/DC1.

Videos of an alternative version of how to perform the head-impulse test can be seen at http://emcrit.org/misc/posterior-stroke-video/.

Note: the VOR is also an important part of the examination of comatose patients (see section 11)

Trigeminal Nerve (CN V.1 - V.3)

Sensory fibers: (epicritical) sensation is tested by lightly touching the forehead, cheeks, and chin bilaterally. Pain is tested by using a wooden toothpick gently on either side. A quick screen test in each of the modality groups **protopathic** (pain, cold) and **epicritic** (light touch, vibration, proprioception, two-point) is sufficient.

Note: Only if the patient has reported sensory symptoms or the disease in question usually involves the sensory system a more thorough examination is done.

If one-sided **hyp(o)esthesia** is reported, the intensity of the stimuli are altered to evaluate the threshold for touch and pinprick on the afflicted side in comparison to the contralateral one in a similar dermatomal/nerve distribution (Figure 2.4.1 and Movie 2.4.1). It may help to ask the patient to compare side-to-side using a **percentage scale** with the normal side being 100% or a **monetary scale** with the normal side being 100 cents. The pinprick sensation is further tested by marching in steps of 1 cm from the hypesthetic side to the normal side, using the same gentle pressure throughout. The border between normal and abnormal sensation should cross the midline by about 1 cm into the afflicted side, due to some crossover and interdigitation of sensory fibers in the midline (Figure 2.4.2). If areas of **hypesthesia** are found, marching from abnormal to normal helps to more clearly demarcate the abnormality, as patients find it easier to say when something becomes normal, than when it stopped.

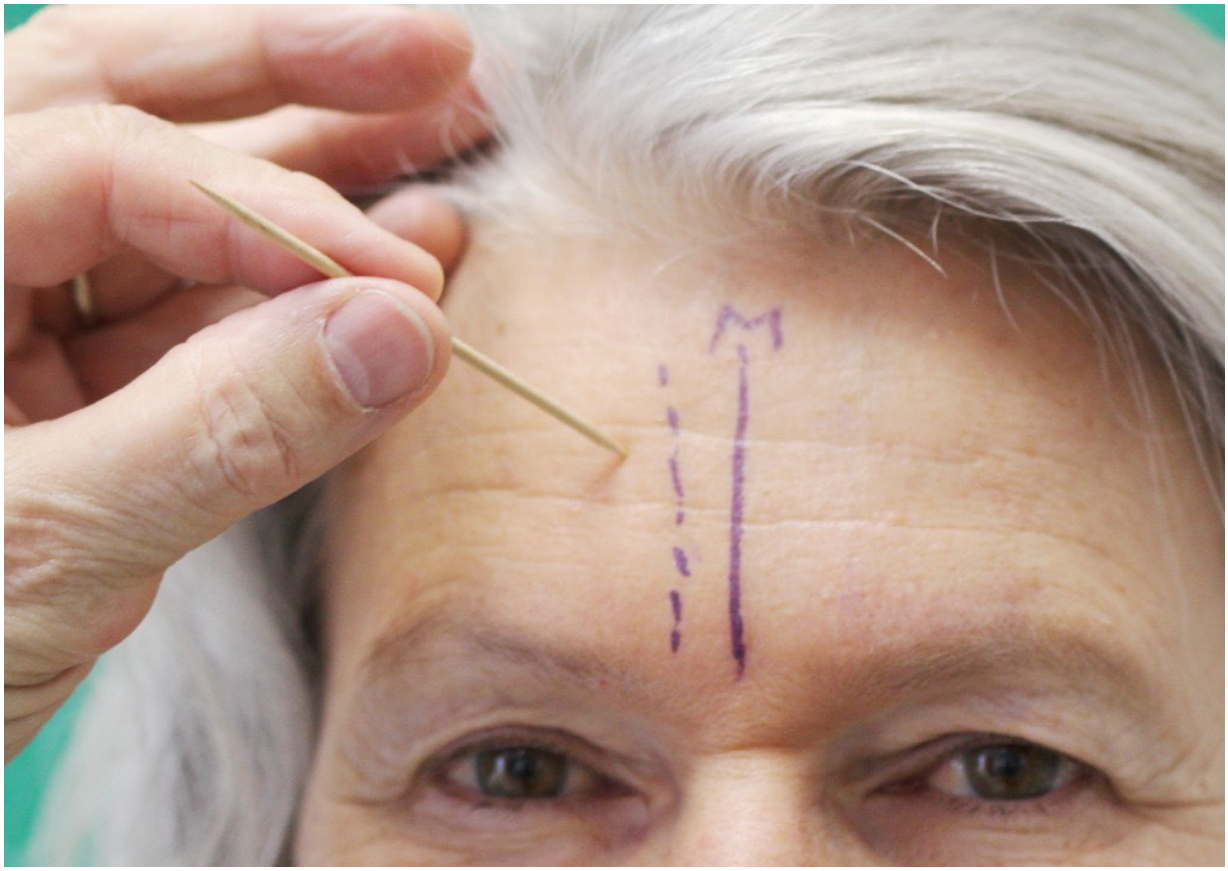
FIGURE 2.4.1 Pinprick

Pinprick is tested with a single-use tooth pick held in the pinch position with a gentle but constant pressure such that the pick may glide and slip through in case the pressure is getting higher (a vs. b). This procedure allows better comparison between left and right and between face and periphery.

MOVIE 2.4.1 Pinprick

Note: the innervation territory of the trigeminal nerve does not include the far angle of the jaw that is innervated by the C2 root. A sensory deficit in this area suggests that the lesion is not in the trigeminal system itself.

FIGURE 2.4.2 Trigeminal nerve sensory testing for pin-prick



*In a right-sided hemihypesthesia or with a right trigeminal lesion the border (dashed line) is about 1 cm before the midline (**M**) when coming from the afflicted side; when coming from the other side the border is past the midline and should be roughly identical.*

Note: all sensory tests are by definition subjective. Yet, if done repeatedly and reproducably it is likely real.

If abnormal findings do not match physiological nerve territories a **functional (conversion) disorder** may be present (see section 9). First, all tests should be repeated several times and in varying order and borders be indicated on the skin by a felt pen (to check for reproducibility). If a **conversion disorder** is under consideration, a **tuning fork** is placed on the middle of the forehead and on each side of the head independently. This should cause a global and midline maximum vibration due to the connection and structure of the skull bones, while some of the functional patients may falsely claim lateralization or decreased sensation over the "hypoesthetic" side.

Corneal reflex: This reflex has its **afferent** fibers in the ophthalmic (V1) portion of the trigeminal nerve; the **efferent** part of the reflex arc runs with the facial nerve, frontal branch (CN VII). The patients should be informed about this harmless but a bit frightening procedure. Take a **sterile cotton swab** or gauze and very gently touch the edge of the cornea approaching **slowly from the side** to avoid an unrelated blink. Normal saline eye drops (if falling directly on the cornea) can also be used for the many repeated exams needed in comatose patients, to avoid corneal irritation (Figure 2.4.3 and Movie 2.4.2).

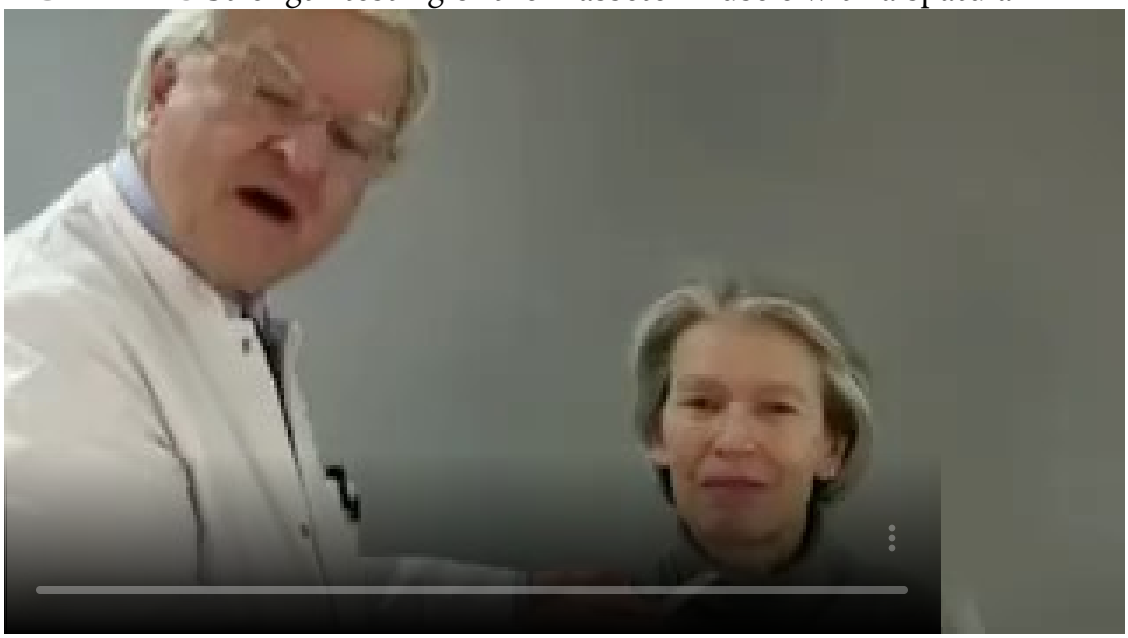
FIGURE 2.4.3 Eliciting the corneal reflex with a sterile swab

MOVIE 2.4.2 Corneal reflex

Jaw muscle strength testing: The masseter muscles are the strongest skeletal muscles. The temporalis and lateral pterygoid muscles are agonists to the masseters. To test for mild weakness a wooden spatula is inserted on either side of the mouth and the patient is asked to bite on it and not let go. If a **cracking sound** is heard from breaking the spatula the strength can be considered normal (Figure 2.4.4 and Movie 2.4.3). With mild or moderate weakness the patient may not be able to hold the spatula against the pull of the examiner. While doing the test the muscle bulks of both muscles can be seen and palpated. In the rare case of unilateral weakness of the pterygoid muscle the chin may deviate from midline **towards the weak side**.

FIGURE 2.4.4 Strength testing of the masseter muscle with a spatula

MOVIE 2.4.3 Strength testing of the masseter muscle with a spatula



Masseter reflex: This is the **only monosynaptic** reflex in the cranial nerves (Figure 2.4.5 and Movie 2.4.4). It is best elicited by holding the examiner's finger just above the chin and then hitting the finger with a **reflex hammer**, with the jaw half open and relaxed. The response is a short elevation towards jaw closure. As in limb reflexes it is graded from none = 0 to 4 = very brisk ("**spastic**"). The **reflex level** of a given patient is often based on the intensity of this reflex if one is certain that no cranial nerve pathology exists.

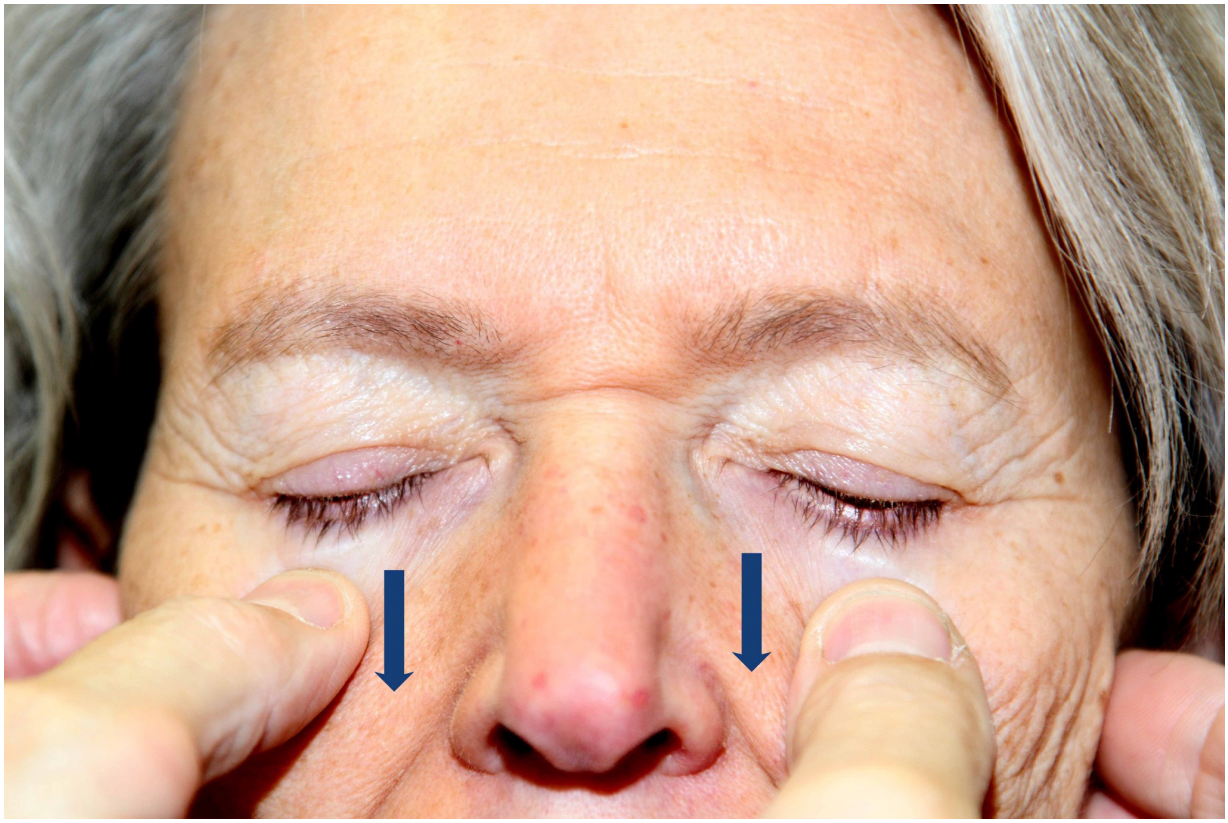
FIGURE 2.4.5 Masseter reflex

MOVIE 2.4.4 Masseter reflex

Facial Nerve (CN VII)

The facial nerve innervates the facial muscles and the tensor tympani muscle of the eardrum. The examiner first looks at the **mimics**. In **central** facial paralysis, facial movement may be almost or completely normal when the facial muscles are activated in an emotional context, such as laughter, suggesting the brainstem activation remains intact while cortical lesions may still exist. In Parkinson's disease reduced expression on one half of the face may be an early sign. Unilateral weakness due to **peripheral** facial nerve palsy is easy to detect but a bilateral weakness will **only come out** by formal testing: grimacing, wrinkling the forehead, squeezing the eyes shut, blowing up the cheeks, whistling, or showing the teeth. The ipsilateral skin folds (particularly the nasolabial fold) of the patient may be less prominent. Muscle strength can then be tested formally in the **frontal region** by firmly closing the eyes ("keep the soap out") while the examiner tries to open them by **gently, but firmly**, placing the thumb on the **lower lid** (Figure 2.5.1) and pulling downwards. **Do not** try a forced eye opening by pressing on the upper lid. With normal strength it will not be possible to open the eye.

FIGURE 2.5.1 Strength testing of the orbicularis oculi muscles



For the middle branches of the facial nerve the patient is asked to blow up the cheeks while the examiner pushes his/her index fingers gently onto the cheeks and checks how much force is necessary to overcome it until the air comes out Figure 2.5.2.

FIGURE 2.5.2 Strength testing of cheek muscles

In nerve inflammation (e.g. Bell's palsy) and in head trauma the **intermedius nerve** may be afflicted because its **fibers run with the facial** nerve. This may lead to loss of **taste** for sweet, sour, and salt. Testing is needed if there is doubt about the lesion site (peripheral vs. central). For testing, the tongue is stretched out and, then the examiner then holds it with a **large gauze** and then places a drop of sweet, sour and salt solutions on the tongue in sequence asking the patient to nod the head after being asked what quality he/she has tasted. Please note that pulling the tongue back into the mouth may move the probe to the unafflicted side allowing the patient to **falsely pass** the test.

Vestibulocochlear nerve (syn.: n. statoacusticus; CN VIII)

The eighth cranial nerve has two main components, the **cochlear (or acoustic)** nerve innervating the inner ear and the **vestibular nerve** innervating the vestibular organ.

The **vestibular nerve** is tested by various manoeuvres. In bedside testing, observation and elicitation of the abnormal eye movements (**nystagmus**) induced by gentle or more abrupt changes in head position is key (see section 2.3). Another simple test is having the patient "marching in place" (Fukuda or Unterberger test) or stepping 10 times forward and backward (Babinski-Weil test) with eyes closed. Any sideward deviation of more than 45° to either side is supposed to be abnormal. Normal limits may be different in the **elderly**. When the patient stretches arms pointing with the index fingers towards the examiner (Barany's test), a slow one-sided deviation may appear after some time, indicating unilateral vestibular dysfunction. More sophisticated testing includes a turning chair with Frenzel lenses (Figure 2.3.7) and cold-warm water irrigation of the outer ear (Figure 2.3.8). Additionally, the psychophysical reaction of the patient may include dizziness, **vertigo**, and nausea up to vomiting. In a dizzy and nauseated patient any maneuver should be done very gently if at all.

The **acoustic nerve** is tested at the bedside by simple hearing tests: normally spoken language and **whispering** presented to either ear with the non-afflicted ear as a control. Next is **rubbing** of the examiner's fingers directly adjacent to the outer ear. If this is not heard, a hearing deficit of **more than 15-20 decibel** can be assumed. Cooperative patients with normal hearing can detect a side difference of less than 10 decibel. If the patient cannot hear the finger rubbing or whispering, two simple tests can be performed before

sending the patient to formal audiometry and auditory evoked potential studies:

Weber's test: an activated tuning fork of 128 Hz or 256 Hz (latter preferred) is placed exactly in the midline of the scalp. If the vibration is heard better on the ipsilateral side of a unilateral hearing loss this indicates a problem in the sound conduction (**conductive loss**) of the outer and middle ear; if it is heard better in the reportedly unaffected ear, it points to a **sensorineural** (i.e. cochlear) **hearing loss**. The test is **not** useful in differentiating symmetrical hearing loss.

Rinne's test: an activated tuning fork is held firmly on the mastoid bone process. When the patient stops hearing the vibration, the fork is held before the outer ear and will still be heard except when the outer or middle ear is impaired by a conductive loss.

TABLE 2.6.1 Summarizes findings with bedside testing of hearing problems

Rinne	Weber w/o lateralization	Weber lateralizes left	Weber lateralizes right
Both ears AC>BC	Normal	Sensorineural loss in right	Sensorineural loss in left
Left ear BC>AC		Conductive loss in left	Combined loss: conductive and sensorineural in left
Right ear BC>AC		Combined loss: conductive and sensorineural loss in right	Conductive loss in right
Both ears BC>AC	Conductive loss in both ears	Combined loss in right and conductive loss on left	Combined loss in left and conductive loss on right

AC = air conduction; BC = bone conduction.

Glossopharyngeal and vagus nerves (CN IX and CN X)

The vagus nerve has a **motor component** to the pharyngeal and laryngeal muscles, a **sensory component** (nerve fibers running together with the glossopharyngeal plus an auricular branch subserving part of the outer ear), and a major autonomic component. Some of these can be tested at the bedside.

Hoarseness and toneless speech (**dysphonias**) are the signs of **uni**lateral or **bi**lateral **laryngeal** muscle weakness, respectively; drooling of saliva may be an indicator of pharyngeal muscle weakness, and a nasal speech (a type of **dysarthria**) indicates weakness of the soft palatal closure. Any of these signs may be caused by a lesion of the vagus nerve or its laryngeal branches. On inspection of the pharynx, poor lifting of the soft palate or an asymmetric elevation can be easily appreciated once the patient is instructed to sound a loud "aah" as in "large", and more clearly upon gently and very briefly touching the soft palate with a spatula, i.e. eliciting the **gag reflex**: The **afferent** path of the reflex arc runs via the glossopharyngeal nerve and the **efferent** path via the vagus nerve. If more than one cranial nerve is involved unilaterally, it is useful to examine the gag reflex by stimulating **either side** of the soft palate separately and check for asymmetry. With unilateral lesions, the uvula and the dorsal wall of the pharynx are **pulled to the normal** (i.e, not afflicted) **side** by the normally functioning levator veli palatini muscle. If the patient does not feel the touch of the spatula, a sensory deficit of that side is the most likely diagnosis. If it is felt perfectly it suggests a defect in the motor path (vagus nerve or innervated muscles) (https://www.youtube.com/watch?v= dPIkJ9XfgvU).

Note: abnormalities in mouth and throat motility are a common sign of many CNS disorders, namely of the brain stem, and of syste-

mic disorders of the motor system, but also of cranial nerve neuromuscular transmission disorders (e.g. myasthenia), and myopathies.

SUBSECTION 8

Accessory nerve (CN XI, in conjunction with anterior roots and motor nerves from roots C1, C2, and C3)

(see also section 3)

On inspection, **atrophy**, **fasciculations**, or **dystonic** movements can easily be seen in the sternocleidomastoid (SCM) and in the trapezius muscles. Muscle strength in the SCM muscles is tested by having the patient turn the head against the examiner's hand over the temporal fossa. The contraction of the contralateral muscle can be seen and felt by touching (Figure 2.8.1).

FIGURE 2.8.1 Strength testing for sternocleidomastoid muscles

Head turning against resistance tests for weakness of the sternocleidomastoid muscle and brings the muscle bulk out (arrows). Recall that shortening of the distance between the left sternum/clavicle and left mastoid results in rightward turning of the head.

Note: the contralateral SCM turns the head towards the examiner's hand. If weak, this may result from a lower motor neuron lesion ipsi-

lateral to the weak SCM. With upper motor neuron lesions weakness may be milder. There are variations as to contralateral or ipsilateral weakness in head turning because of variable pathways.

The **trapezius muscle** is tested by having the patient shrug the shoulders against resistance.

SUBSECTION 9

Hypoglossus nerve (CN XII)

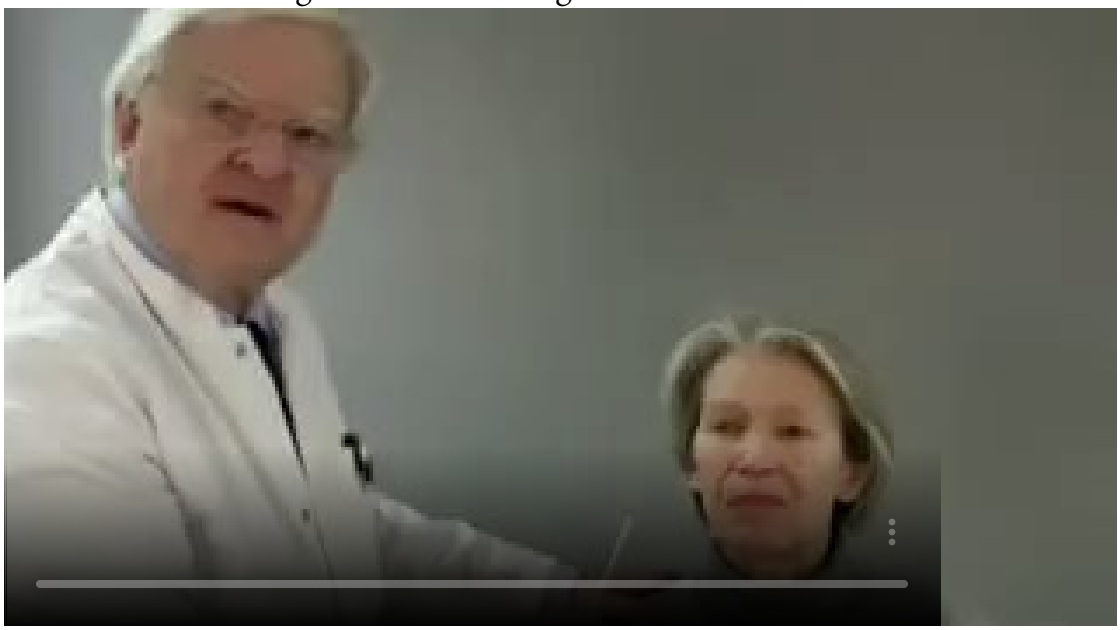
The tongue is best inspected with the patient in a resting position, sitting or supine, with the mouth opened and the tongue **lying relaxed** in the floor of the mouth. The tongue normally shows mild undulating movements. Muscle atrophy, **fasciculations** (sometimes referred to as "fibrillations"), and dystonic movements are best seen with daylight or a flashlight shining light from the side, along the surface of the tongue.

Muscle strength is tested by asking the patient to push the tongue into the cheeks from inside against the examiner's fingers positioned on the outside of the cheek. Next, the patient is asked to stretch out the tongue straight against resistance of a spatula. Unilateral lesions of CN XII lead to a deviation of the tongue toward the **afflicted** side because normally functioning intrinsic muscles push the tongue out of the mouth on the contralateral (not afflicted) side, i.e. having the patient push his/her tongue to the **right** tests for **left**-sided tongue muscles and vice versa. Mild weakness can be appreciated by asking the patient to push a spatula away against resistance (Figure 2.9.1 and Movie 2.9.1).

FIGURE 2.9.1 Testing for intrinsic tongue muscles

The spatula is pushed to the right by the left-sided intrinsic tongue muscles.

MOVIE 2.9.1 Testing for intrinsic tongue muscles



Tongue weakness, peripheral or central in origin, or incoordination may best be appreciated by listening to the precision of articulation (Dysarthria). Tongue motility is further tested by having the patient speak **lingual and glottal consonants** in fast reiteration (such as "te-te-te-te-te" and "ga-ga-ga-ga-ga", respectively) and moving the tongue **as fast as possible** from left to right and back. This will be slowed **and** speech becomes dysarthric despite near normal strength in **central** (supranuclear) lesions (pseudobulbar palsy) and becomes slow with combined severe weakness and atrophy in **peripheral** nerve lesions, neuromuscular transmission disorders, and lower motor neuron disease (bulbar palsy).

Concluding remark: there is a wide differential diagnosis and a thorough examination of all cranial nerves will help to narrow down the options for a diagnosis.

SECTION 3 – MOTOR SYSTEM (A)

General Terminology

(also refers to sections 5 and 9)

Astasia

Abasia

Ataxia

Dysmetria

Steppage gait

Spasticity

Festinating gait

Magnetic gait

MRC grades

UMN

LMN

Rigidity

Paratonia

Myotonia

Neuromyotonia

Fasciculations

Dystonia

Chorea

Ballismus

Myoclonus

Tremor

Postural tremor

Kinetic tremor

Intention tremor

Note: all complex tests of the motor system rely on pattern analyses used as screening tests for various types of abnormalities; for the inexperienced examiner, they usually do not easily differentiate between force (muscle strength), coordination, and underlying joint, tendon, and bone conditions (see section 5).

Some motor signs also apply to muscles innervated by cranial motor nerves (see section 2).

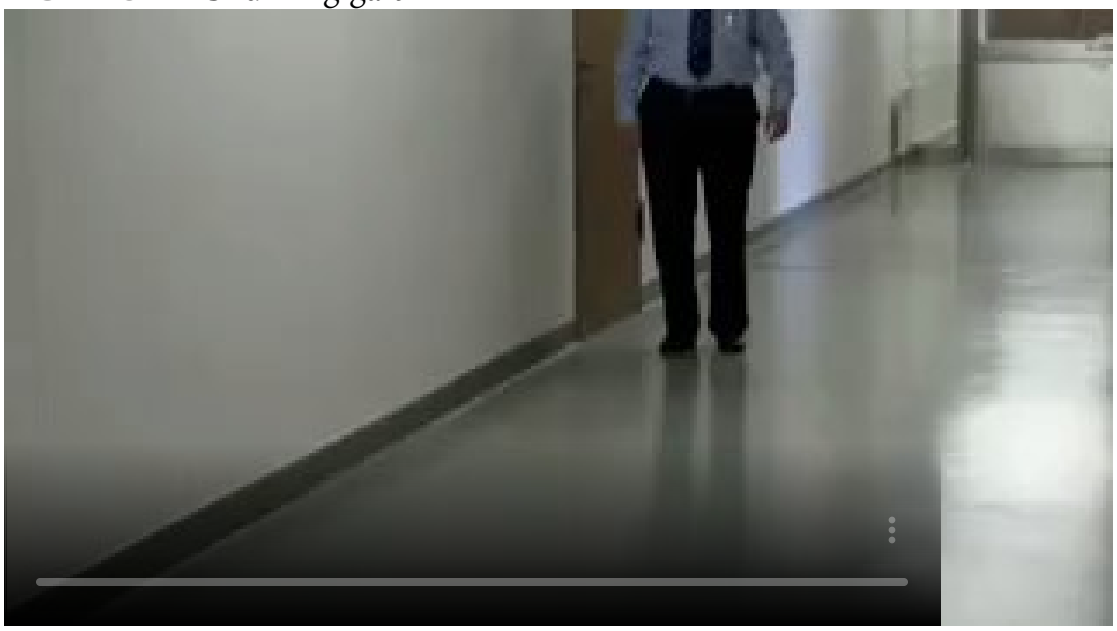
The examination aims at first detecting isolated or systemic motor abnormalities by **more global tests** (e.g. gait, stance) which then are dissected into a series of formal test procedures as outlined in the sections below. It is necessary to have the patient **undress** to see overt abnormalities such as muscle **atrophy**, circumscribed or generalized **hyperkinesias** including **chorea**, **fasciculations** and the like. It does **not** make sense to first examine all details, e.g. single muscle strength before getting an overall idea of the abnormalities from the patient or bystanders.

SUBSECTION 1

Gait and stance

The examiner will observe the patient when entering the room. If that is not possible, the **gait and stance** examination should be the **first thing** to do. Posture, stability of stance and gait, arm swing, and gait pattern, with the two phases of any step forward (stance and swing phase) are being observed and any abnormality is documented by narrative description. There is no need to diagnose at this point, but a simple description of what is observed will suffice. In all movement abnormalities, recording the patient's movement, after permission, on a video may be helpful. Commonly seen **abnormal patterns** include: **limping**, with reduced elevation of the leg when advancing for the next step (multiple causes), or with uni- or bilateral **foot drop** (weakness of dorsiflexion); abnormally slow and/or **dragging gait** with a reduced step distance and starting problems, combined with **abnormal posture** (e.g. in Parkinson's disease (see Movie 3.1.3 and Movie 3.1.4) and other movements disorders); broad-based shuffling gait (**dysbasia**, e.g. in subcortical encephalopathy, see Movie 3.1.1); unsteadiness with increased sway of the body (**ataxia**; see Movie 3.1.7 and Movie 3.1.8), stiff limbs while moving forward with uni- or bilateral dragging of the forefoot and circumduction (**spastic gait**; see Movie 3.1.2). Generalized hyperkinesia such as **chorea** in Huntington's disease may also produce a typical gait abnormality (see Movie 3.1.6) . These abnormalities can **co-occur** in variable **combinations**, and hence identifying the underlying gait patterns can be challenging. It is helpful to not only observe but also to listen to the gait pattern in a hallway (limping, staggering, or shuffling – all give typical sounds). The gait will also tell you about asymmetries and muscle tone abnormalities.

MOVIE 3.1.1 Shuffling gait



MOVIE 3.1.2 Spastic gait

MOVIE 3.1.3 Parkinsonian gait

MOVIE 3.1.4 Mild Parkinsonian gait

MOVIE 3.1.5 Dystonia

MOVIE 3.1.6 Chorea

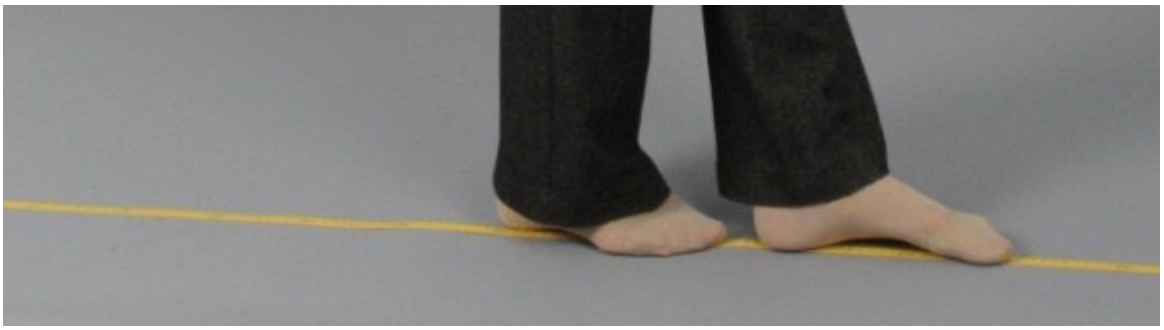
MOVIE 3.1.7 Gait ataxia (cerebellar degeneration)

MOVIE 3.1.8 Gait ataxia (MS)

Different gait abnormalities:

The patient is asked to **walk** by placing one foot in front of the other (Figure 3.1.1) **tandem gait**) on a **real or imaginary** line on the floor (as if on a tight rope or balance beam); any deviations are documented, because it is the description of what happened that is going to help with the diagnosis. If no obvious abnormality can be seen, the test is continued with closed eyes, but the examiner or an aid **must** be very close by and **ready to assist** if the patient tends to deviate and fall. This test is also part of coordination testing (section 5).

FIGURE 3.1.1 Tandem gait on a straight line



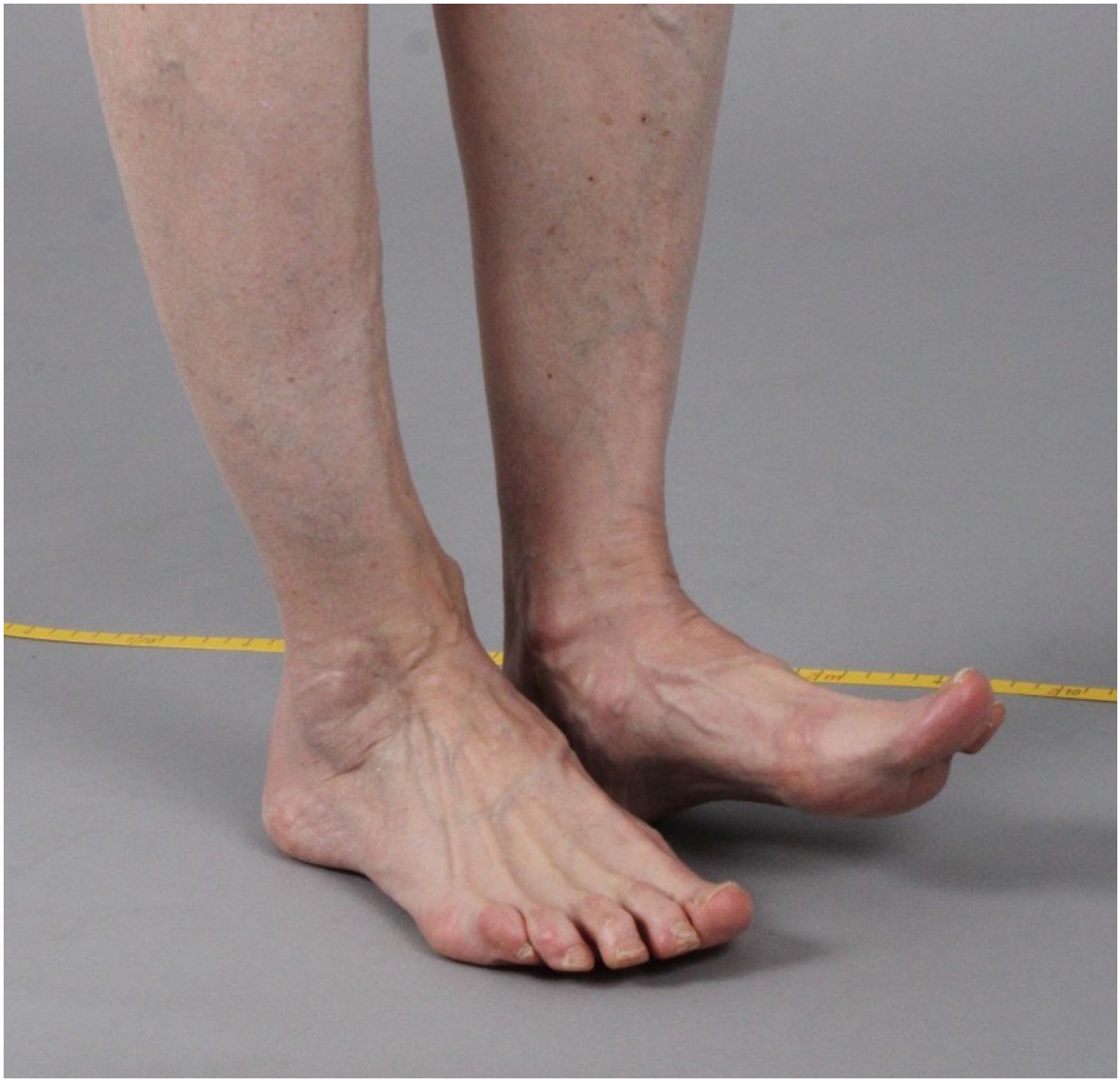
(a) Tandem gait on a straight line-eyes open. The examiner stands aside to provide immediate support if the patient is unsteady.

(b) Tandem gait on a straight line-eyes closed. The examiner stands aside to provide immediate support if the patient is unsteady.

Next, **tiptoe and heel-walking** are done which collectively test both the muscle strength in the lower limb and coordination. If the patient cannot walk on heels make sure you exclude a contracture of the Achilles tendon by testing passive range of motion. An example of a **unilateral** weakness of foot elevators (flexors) on attempted heel walking is shown Figure 3.1.2 and normal tiptoe walking is shown Figure 3.1.3.

FIGURE 3.1.2 Heel and tip-toe walking

(a) shows normal heel-walking;



(b) a weak foot elevation is seen on the right, e.g. in peroneal nerve palsy;

(c) normal tiptoe walking

Then, one should have the patient **hop on one leg alternatingly**, as well as have the patient squat with the thighs held at 45 degrees. If these tests are normal, any major muscle weakness is unlikely. Various limitations may be seen, and again this should be described and documented in the context of other tests.

FIGURE 3.1.3 Hopping on either leg as a globel motor test

This test examines muscle strength and coordination in trunk and lower limbs.

Note: any type of pain induced by and superimposed on these manoeuvres will influence the pattern or frankly prevent the patient from performing the test.

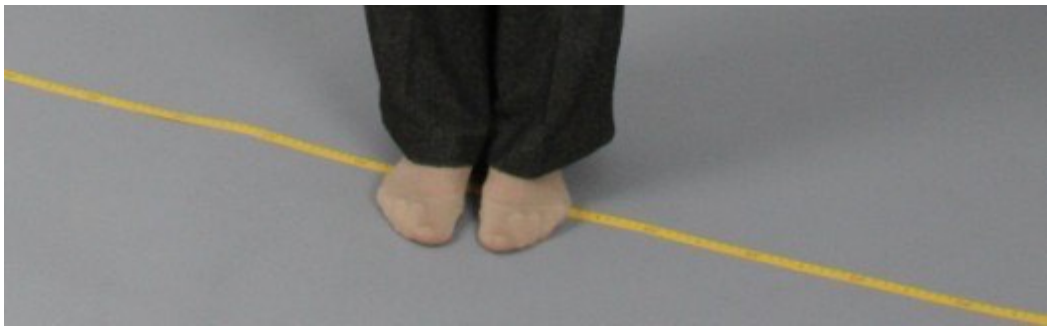
Occasionally, gait patterns become apparent that do not fit patterns in defined organic diseases. Here, a non-organic ("functional" or conversion disorder, see section 9) may be the basis. These **hints include**: highly variable patterns with stumbling or limping, deviating to alternate sides, stiffness of limb with alternating sides, gait with double canes or crutches despite good muscle power, or the classic wildly flailing or stumbling gait that collapses only when an examiner is nearby ("pseudo-astasia-abasia", see section 9). ***Note that the examiner must be ready to support the patient at any time when doing motor testing.***

Once a **conversion disorder** (see section 9) is in the differential, the patient is asked to walk slowly **backward**. If this can be achieved as good or bad as forward or even better, a functional pathology becomes likely. Better even, the patient is held and **supported** under his/her arm(s) or held on the hands by one or two examiners. A functional patient will not "take" the offered support in a constant and reproducible way but rather exert too much or too little power onto the supporting hands, and in addition, may breathe heavily as if over-exercised, or just give way.

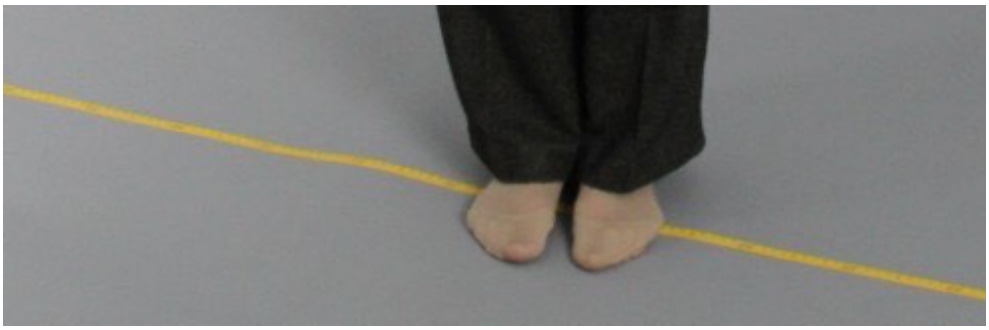
One test for abnormalities of stance **and** proximal central weakness (**paresis**) of the upper limbs is the modified **Romberg manoeuvre**; **do not try** this if the patient has an obvious major incoordination as evidenced by the previous tests. The patient is asked to stand still with both feet close to each other, and both arms outstretched and supinated (palms up, Figure 3.1.4). If the patient is **safe** with open eyes he/she is asked to close both eyes with **the examiner ready to support**. Everybody will be a bit less stable when doing this (i.e. exaggeration of the sway) but any **major** dissociation

between open and closed eyes points to a **disorder of sensory input** – either from the posterior columns of the spinal cord or from severe de-afferentation in the peripheral nervous system (historically named "tabetic" in late syphilis, **pseudo-tabetic** in other conditions). The basis for this observation is the fact that the visual system makes up for the de-afferentation of the PNS/spinal cord system. In severe cases or in cerebellar disease it cannot be compensated by visual control; thus stance and gait may look quite uncoordinated even with open eyes. If there is a high suspicion for instability, another modification of the Romberg manoeuvre can be used: one foot in front of the other with eyes closed to help bring out the deficit.

FIGURE 3.1.4 Modified Romberg manoeuvre (normal and abnormal)



(a) the patient stands safely balanced, with eyes open and closed;



(b) typical pattern in a mild cortical or subcortical hemispheric lesion: with eyes closed he right arm may slowly move down with partial pronation of the hand while the other side stays normal.

MOVIE 3.1.9 Leg lifting belly down (normal)

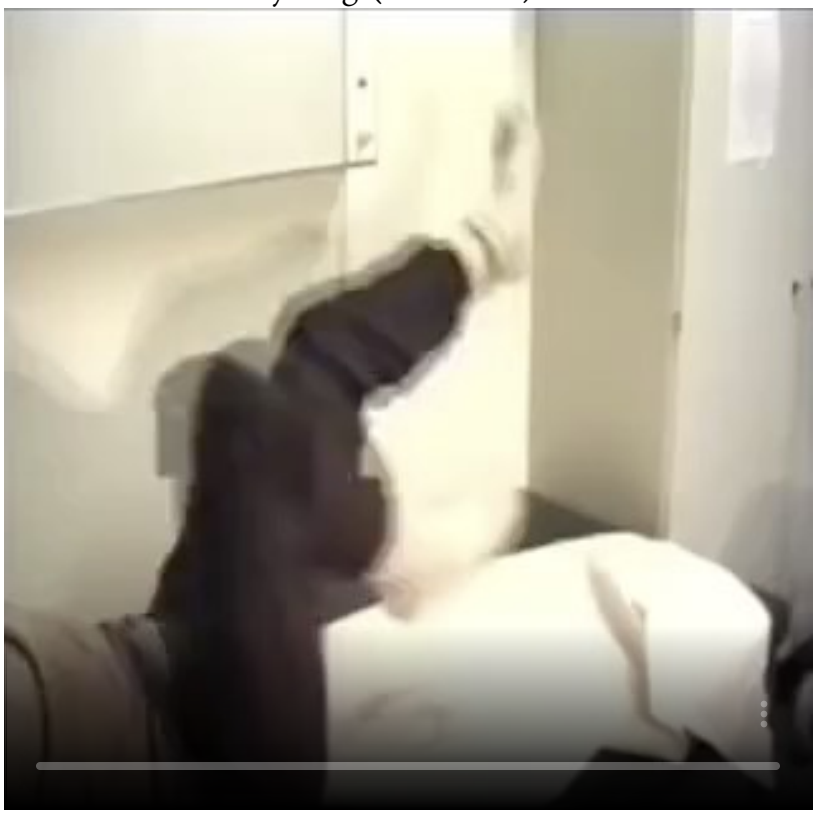
MOVIE 3.1.10 Leg lifting belly down (abnormal, left central weakness)

MOVIE 3.1.11 Leg lifting supine (abnormal, left central weakness)

MOVIE 3.1.12 Modified **Romberg manoeuvre**

MOVIE 3.1.13 Bicycling (normal)

MOVIE 3.1.14 Bicycling (abnormal, backwards >> forward, MS)



If the patient with a basal ganglia disorder or a cerebellar disorder tends to fall, the **pull-test** is a sensitive test for instability of stance. The patient is informed that the examiner will pull the patient backwards with **full protection**. Then the patient is pulled back abruptly and the reaction is observed. Normally, the patient will immediately swing one leg backwards to improve his/her balance (see section 5 and Movie 5.5.1).

Note: there is a 3-system (proprioception, vision, cerebellar) management of stance and a person needs 2 of 3 working systems (of which vision is the dominant, and most difficult to compensate for), if any of them are out and the eyes are shut only one is left, and the system fails, potentially resulting in a falling patient on the Romberg test.

In the case that all the above tests are normal it is **unlikely** that the patient suffers from any functionally relevant problems in

legs, cerebellar, and spinal cord motor function, or in position sense function.

Further tests of the motor system Muscle bulk, tone and strength

Bulk: in the sitting and supine position the muscle bulk is checked for focal or more generalized atrophy and documented as either proximal or distal muscle atrophy, symmetrical or asymmetrical, and graded from mild to severe. Or else, it may be just a very focal atrophy in the distribution of one peripheral nerve or a spinal segment. In any disease, atrophy will likely be a chronic or chronic progressive abnormality. In patients under treatment it makes sense to **measure the circumference** of the limbs in positions defined by bony landmarks (Table 3.2.1) as an objective measure. Some excellent locations in which to examine muscle bulk at first sight include the first dorsal interosseus muscle of the hand and the extensor digitorum brevis of the foot, the anterior tibial (Figure 3.2.1) and the calf muscles. Atrophy of the thigh muscles can be **masked** by subcutaneous fat tissue.

FIGURE 3.2.1 Shows severe muscle atrophy of the anterior tibial muscles in a patient with a symmetric vasculitic neuropathy leading to the "sharp shin" sign.

Note: in the elderly intrinsic hand and foot muscles may appear atrophic because of loss of "soft tissue" but may have normal strength and muscle bulk.

TABLE 3.2.1 Assessment of muscle atrophy by measuring the circumference of upper and lower extremities with a tape measure at indicated landmarks.

Circumference (in a relaxed state)	**right**	**left**
Upper arm (10 cm above the olecranon)		
Forearm (10 cm below the olecranon)		
Thigh (20 cm above the medial cleft of the knee joint)		
Lower leg (at the maximal bulk of the calf muscles)		

You can download this form for your own use here: https://www.albert-zwei.de/images/ebook/tab_3_2_1_en.pdf

Consistency and tenderness: the muscles can be palpated and their consistency rated (soft-flaccid, harder than normal) by side-to-side and proximal-to-distal comparisons. Other features which should be noted are subjective features and may not easily be verified by bedside testing: stiff and crampy; painful tenderness on minor pressure. If further clarification of these muscle findings is essential to make the diagnosis, ancillary tests including electromyography, ultrasound, and CT/MRI may help to differentiate these features.

Muscle tone: this is best appreciated in the sitting position for the **lower limbs** by letting the legs **swing back and forth** upon a minor push. In the standing position the **upper limb** muscle tone can be tested by gently **turning the trunk** with the arms hanging down. The tone in the hands is tested by passive **pronation-supination** and in the foot and in proximal parts of the limbs by passive **flexion-extension**. A subtle abnormality of asymmetric "spastic" muscle tone can be appreciated when each leg is **rolled passively** by

the examiner along its longitudinal axis while the patient is in the supine position (like rolling a log). In a patient with a mild hemispheric lesion the ipsilateral leg will roll out more than the contralateral leg. Normal foot swing in an axial plane (perpendicular to the longitudinal body axis) is usually quite symmetric with a mild lag at the ankle joint, whereas the amplitude of the foot swing is reduced and rolls with the leg as a whole on the abnormal side.

The following abnormalities can be classified by these tests:

- Spasticity: on passive extension of e.g. a flexed elbow or knee joint there may be **increased resistance** that builds up when increasing force is exerted (a massively exaggerated [ex]tension [or stretch] reflex) such that more resistance is encountered with **increasing velocity** and force of passive movements, while scaling the test down to slower movements may **revert** the muscle tone to the point of even feeling normal. At a certain point of increasing the force of passive movement suddenly the muscles may give way (clasp-knife phenomenon, best demonstrated here):

MOVIE 3.2.1 Testing for Spasticity

- Spasticity usually predominates in flexor muscles in the upper limbs, on extensors in the lower limbs. Additionally, spasticity may be so severe that the joint cannot be bent or extended passively by the examiner even with slower movements. In this case, a firm **sustained pressure** is applied to the distal body part to move the joint slowly while the patient is closely observed for signs of discomfort, if he/she is unable to communicate. The ability to move a flexed or extended joint rules out joint contracture, and may indicate that the patient can benefit from tone reducing measures. (Movie 3.2.1)
- Rigidity: the resistance to passive extension and flexion is uniformly increased throughout the entire range of the movement. Rigidity afflicts generally all muscles including those of the trunk. In mild rigidity this may only come out if the patient is asked to move the contralateral limb like opening and closing a fist, moving the contralateral foot or toes up and down (Froment's manoeuvre). Rigidity in Parkinson's disease may be mixed with slow frequency resting tremor (at about 3-6 per second, giving rise to the cogwheel phenomenon), in particular in the wrists (see Movie 3.2.16).
- Paratonia ("**gegenhalten**"): is a more or less constant and sometimes massive increase in tone throughout the **entire range** of passive movements (like through a highly viscous fluid – wax like). It is common in **advanced** degenerative brain disorders and may be mixed with rigidity or an inability of the patient to not assist the movement throughout the full range of motion ("**mitgehen**"). It is also often associated with a fixed limb phenomenon, i.e. the patient may maintain a given position after a passive movement, although this may occur more frequently as a sign of extreme rigidity. The concept of paratonia is pathophysiologically ill defined and is not felt to be useful to differentiate various disorders.
- **Hypotonia**: loss of muscle tone, in the awake patient as a sign of advanced muscle weakness from disorders of muscle, nerve,

or motor neurons, with (or less commonly without) obvious muscle atrophy. In the unconscious or intoxicated patient hypotonia may be a prominent sign but may be of no specific diagnostic value. Hypotonia is also associated with cerebellar disease.

- **Hypertonia**: Characteristically involves myotonia and neuromyotonia. In myotonia any abrupt movement or a tap by the reflex hammer typically induces local muscle bulking with cramping of the muscle. Active or passive decontraction is delayed. In some types of myotonia weakness and fatigue can be associated. If myotonia is in the differential, it can be tested by having the patient lay the tongue on a spatula, the examiner holding a second spatula upright with the thin edge sitting on the tongue and gently bouncing the reflex hammer on the upright spatula (Figure 3.2.2).
- **In** neuromyotonia the apparently increased resting tone may be the result of continuous muscle fiber activity, sometimes there is a wave-like movement that can be seen rippling over muscle areas (something also seen in rippling muscle disease, a rare genetic muscular disorder). **Stiffness and cramps** are also features of the stiff- person syndrome and are usually in the axial muscles. If hypertonia is unilateral or below the waist and flexor muscles are more involved than extensor muscles, the most likely reason is a spastic **hemiparesis** or paraparesis (e.g. hemispheric post-stroke condition or **spinal cord lesion**, respectively).

FIGURE 3.2.2 Test for tongue (percussion) myotonia

The patient is asked to place the tongue on a spatula. A second spatula is placed by the examiner above the tongue and this spatula is gently hit by the hammer. In myotonia a small muscle bulk will form and stay for a second or two.

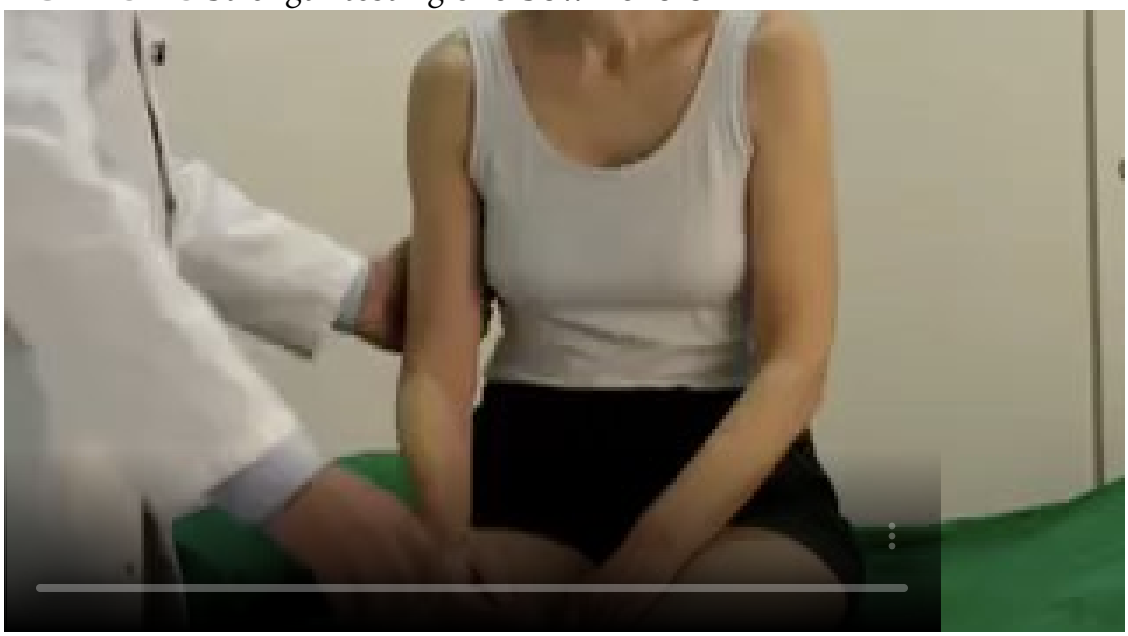
Formal muscle strength testing: the testing is usually done on **groups of muscles** that are representing the moving force in **one major joint** rather than testing individual muscles. The latter is done if a very **focal** weakness and/or identifiable focal muscle atrophy is seen. The patient is asked to perform a specific movement by bending or flexing a joint against the resisting force of the examiner's hand. The best position to start with is **half way** between flexion and extension of a given joint. An isometric strength test is performed and the more power the patient exerts the more is the resisting force, until finally the examiner will overcome the patient's force. To help localize weakness, the Nerve Whiz app is a free aid to help localize PNS lesions.

Note: neck extensors are amongst the strongest skeletal muscle groups. Strength is best tested bimanually.

FIGURE 3.2.3 Strength testing of the neck extensors with both forearms resting flat on the patient's chest. With normal strength extension can hardly be overcome or not at all.

MOVIE 3.2.2 Strength testing of the neck extensors

MOVIE 3.2.3 Strength testing of **elbow flexors**



MOVIE 3.2.4 Strength testing of the triceps brachii

MOVIE 3.2.5 Strength testing of **forearm and hand muscles**

The grading system is the one originally formulated by the Medical Research Council of the UK (**MRC**) (see General Terminology and "**Aids to the Examination of the Peripheral Nervous System**", see References). **This manual, completely re-edited by O'Brian and the Guarantors of Brain, contains drawings and photos of**

all muscles to be tested and therefore we do not reduplicate this here. Because most conditions are in the range between grades 3 and 5, intermediate **half grades** have been introduced (4-, 4+) but these grades are not precisely defined, though some feel that a 4+ is barely moveable by the examiner (while 5 is unmoveable), a 4 is able to be moved with full force, and a 4- requires a couple fingers of the examiner to be overcome. Since 3 means moving a limb in the **full range against gravity** a 3- has often been used if full movement is still possible but no more against gravity, e.g. full flexion in the hip joint when lying on the contralateral side or full abduction of the shoulder joint when in supine position.

Since grades 5 (and even 4+) **depend** on the maximum force exerted by the examiner it is advisable to use a more objectively quantitative measure for long-term follow up of progressive disorders or of those responding to treatment: **dynamometry** uses springs or strain gauges of various kinds. Two such validated dynamometers having been used in treatment trials are the Drachman hand-held dynamometer (Beck et al, 1999), and the Martin air pressure dynamometer for measuring grip strength (Merkies et al., 2000). Many others are on the market. A table allows you to fill in the various muscle strength number of the **MRC** scale or values obtained in kilograms (or pounds) when tested with a dynamometer (Table 3.2.2).

TABLE 3.2.2 Quantitative muscle testing in neuromuscular disorders and nerve injuries

right			Muscle	left		
Dynam.[1]	Atrophy[2]	MRC Grade[3]		MRC Grade	Atrophy	Dynam.
			Neck flexors			
			Neck extensors			
			Shoulder joint abductors			
			Shoulder joint exorotators			
			Shoulder joint endorotators			
			Scapula adduction			
			Elbow joint flexors (biceps and brachioradialis)			
			Elbow joint extensors			
			Elbow joint supinators			
			Elbow joint pronators			
			Hand flexors			
			Hand extensors			
			Finger flexors (base)			
			Finger flexors (middle)			
			Finger flexors (terminal)			
			Finger extensors			
			Finger spreading			
			Finger adductors			
			Thumb abductors			
			Thumb adductor			
			Thumb flexor			
			Thumb extensor			
			Thumb opposition			
			Abdominal muscles			
			Hip joint flexors			
			Hip joint extensors			
			Hip joint abductors			
			Hip joint adductors			
			Knee joint flexors			
			Knee joint extensors			
			Ankle (foot) flexors[4] (up)			
			Ankle extensors[4] (down)			
			Ankle pronators			
			Ankle supinators			
			Big toe flexor (down)			
			Big toe extensor (up)			
			Small toes flexors (down)			
			Small toes extensors (up)			
			Toe spreading			

[1] dynamometer, in kilograms or pounds [2] mild = +: moderate = ++; marked = +++ [3] standard MRC grade
[4] physiologically "flexors" but anatomically "extensors", and vice versa

Quantitative muscle testing in neuromuscular disorders and nerve injuries. Note that formal testing of functional muscle groups is useful in systemic neuromuscular conditions. Only in regionally restricted disorders single muscle testing is useful. Quantitative measures are helpful in evaluating progression or amelioration following treatments. You can download this form for your own use here: https://www.albert-zwei.de/images/ebook/tab_3_2_2_en.pdf

The innervation of skeletal muscles by an individual peripheral nerve or one or more root- or spinal cord segments (the myotomes) is also elaborated in the manual "**Aids to the Examination of the Peripheral nervous System**", see References).

Notes:

- ***With painful muscles, joints, and tendons the respective muscle strength cannot be tested reliably.***
- ***Occasionally rare mitochondrial cytopathies and neuromuscular transmission disorders may result in quite variable muscle strength on exertion that may mislead the examiner.***
- ***Highly inconsistent findings (changes over hours and days) may have a functional (non-organic) basis best explained by a conversion disorder that needs to be diagnosed properly.***

Further signs of motor impairment (see also Section 5)

Dexterity and fine movements of the hands and feet: after formal muscle testing for weakness has been done **fine motor** movements are tested by having the patient write and draw (see Movie 3.2.11, Movie 3.2.13, Movie 3.2.14, Movie 3.2.15), rapid finger tapping one hand at a time, or doing a "trill"-like alternate tapping of index and middle finger. **Loss of dexterity** may result from

muscle weakness (peripheral or central origin), or from

movement disorders (linked to the basal ganglia, cerebellum, or in the cortical and subcortical area) with little if any weakness on formal testing, and from

sensory de-afferentation with severe sensory neuropathy, in particular due to dorsal spinal ganglion disease (see above pseudotabes), or due to

spinal cord disease.

fine repeated/fractionated movements are also a **very sensitive** test for upper motor neuron (**UMN**) lesions (**spastic** syndromes).

For example, toe tapping is considered a more sensitive and specific test than the Babinski sign by one study (Miller and Johnston, 2005), but this test is dependent on a fully cooperative patient.

It is important to observe the hand movements carefully while the patient performs the tests, and report the observations in a descriptive way. Finger tapping can be measured in **taps per second** (normal around 4-6/sec), cadence/rhythmicity, force consistency (see Movie 3.2.6, Movie 3.2.7, Movie 3.2.9, Movie 3.2.10).

The accuracy of thumb-finger closures and openings is tested with another simple procedure, the thumb-finger test. A normal pattern is shown for the right hand but the left hand shows an abnormal pattern (see Movie 3.2.8). The read-out can be done in runs per second (normal value back and forth around 2-3 seconds).

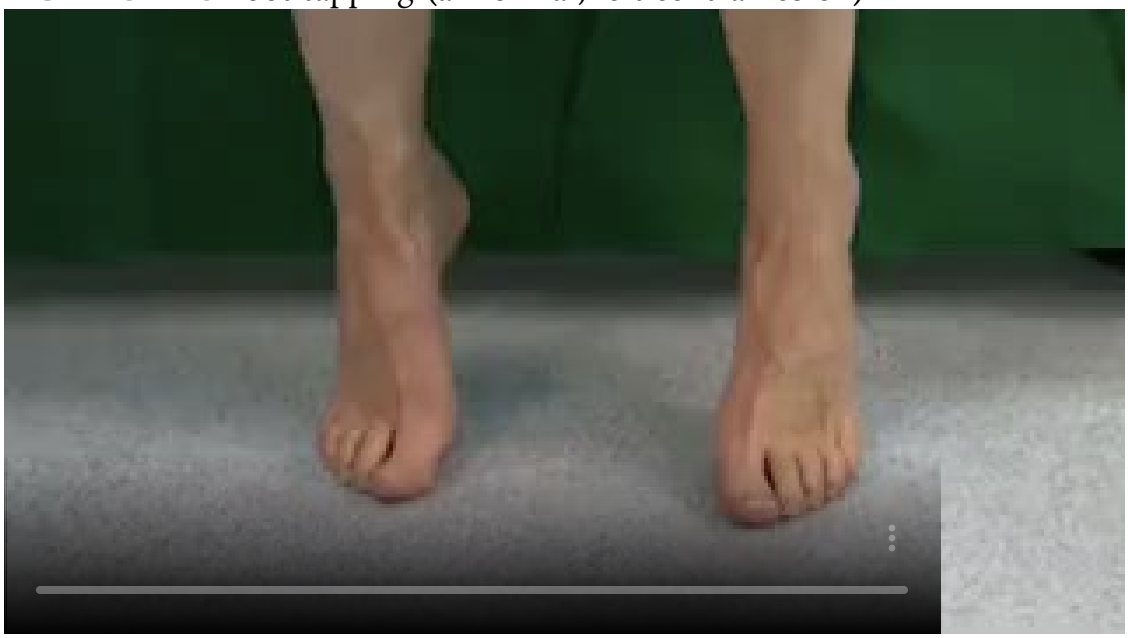
MOVIE 3.2.6 Finger tapping and trills bilateral (normal)

MOVIE 3.2.7 Finger tapping and trills unilateral (normal)

MOVIE 3.2.8 Thumb-finger test (normal with right hand, abnormal with left hand)

MOVIE 3.2.9 Foot tapping bilateral (normal)

MOVIE 3.2.10 Foot tapping (abnormal, left central lesion)



Task-specific dystonias (e.g., writer's cramp/musician's cramps) can also show a decreased performance in writing or finger tapping.

Checking the ability of a patient to **maintain limb position** with his/her eyes closed also helps to pick up on subtle UMN weak-

ness. If patients show a gradual decline (**drift**) of one arm **combined** with slow pronation of the forearm this indicates a **proximal, CNS pattern of weakness** most often associated with a hemispheric disorder involving the pyramidal tracts (see **Romberg test**, Figure 3.1.4Fig. 3.2.4, Movie 3.1.12) . The equivalent for thelower limbs is checked in the **supine** (lying on the back) position when the patient is asked to hold his/her legs up with knees and hips bent at 90°, eyes closed, and then the leg will show a gradual decline (**drift**) on one or both sides if there is a central weakness (Movie 3.1.11). This leg test is demanding and often cannot be done in the elderly, and so instead the patient can be placed in the **prone** (lying belly-down) position with the knees bent up, while watching for a gradual decline (**drift**) after a few seconds. In all these tests the patient is also asked which leg he/she **feels** to be heavier than the other. The paretic limb usually **feels heavier** and that corroborates the finding. All these tests are highly suggestive for a CNS disorder if the formal strength test has shown **normal or near normal** values (see below) and if no pain is present while doing the test. (see Movie 3.1.9 and Movie 3.1.10)

A simple and subtle sign sensitive to **UMN** lesions is brought about by asking the patient to rapidly lift up his/her arms or his/her legs upon a "ready-steady-go" command by the examiner (i.e. a **ballistic** movement): The affected side lags behind the intact side.

Orbiting around a weak limb when rotating fingers, arms, or legs forward and/or backward **as fast as possible** (twiddling the thumbs around each other or performing **cycling movements** like riding a **bicycle** while lying on the examination bed) suggests an **UMN** pattern of weakness if there are no structural or known peripheral nerve/muscle pathologies present. The **UMN** deficit will show a relative slowness and/or smaller radius of the movement on the affected side. Cycling **backwards** is even more sensitive (see Movie 3.1.13 and Movie 3.1.14)

Tremors, such as **action tremor** and goal-directed tremor/**intention tremor** (see Movie 5.1.1, Movie 5.1.2 and Movie 5.1.3)

may be disabling motor abnormalities and may represent a major handicap in daily living. They can be seen with several disorders. In **essential tremor** (Movie 3.2.17) the action tremor constitutes the core motor condition. The **tremor** can be observed when the patient performs translational movements, and may be more severe when performing mouth-directed movements, such as when drinking from a glass or cup. **Tremor** may also be more pronounced or visible while the patient performs precision movements such as writing, drawing, and eating. In contrast, in Parkinsonian resting tremor the tremor gets better when performing a goal directed movement or when drawing a spiral (Movie 3.2.16).

The abnormality during precision movements is documented by having the patient write (copying and writing name and address) and draw. Two normal tests are shown (Movie 3.2.11, Movie 3.2.12). Note the size of the writing, the correctness of the copied text, fluency of the writing, and any consistent axis of shakiness if demonstrated in the text. In Parkinson disease and in some other central motor system disorders the writing becomes slower and smaller than normal.

A simple test is **drawing lines** in between a spiral or between printed straight lines. A normal test is shown including total time (Figure 3.2.4). The patient starts drawing his/her line in the open space between the indicated spiral and ends at the innermost circle or in reverse order while instructed not to hit the printed lines and having the forearm and the elbow elevated without support from the table while drawing. Tests are taken first with the better hand and subsequently with the more afflicted hand on a new sheet. The time needed to perform the tests (in seconds) and the counts of hitting the printed lines can be recorded for both hands when quantification is intended for monitoring treatment effects, in addition to a video documentation (Movie 3.2.14, Movie 3.2.15, Movie 3.2.16).

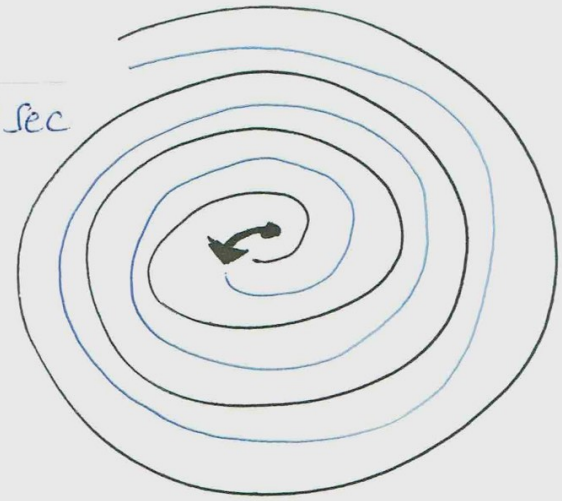
The same test can be used for **any other** motor disorder when dexterity is reduced.

FIGURE 3.2.4 Simple test to evaluate precision movements

Name: Smith, John Date 16 Nov 2015

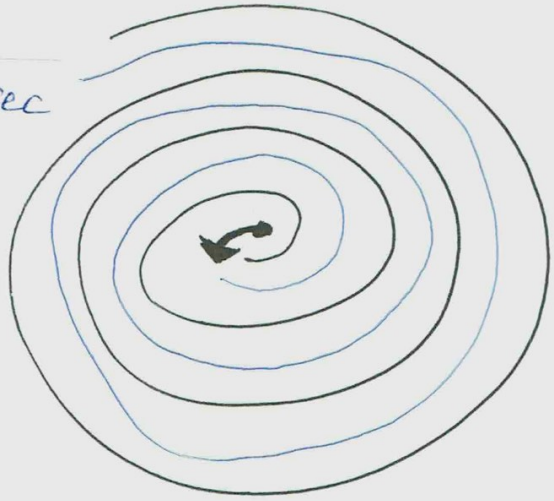
Medication: None

10.4 sec



Right Hand

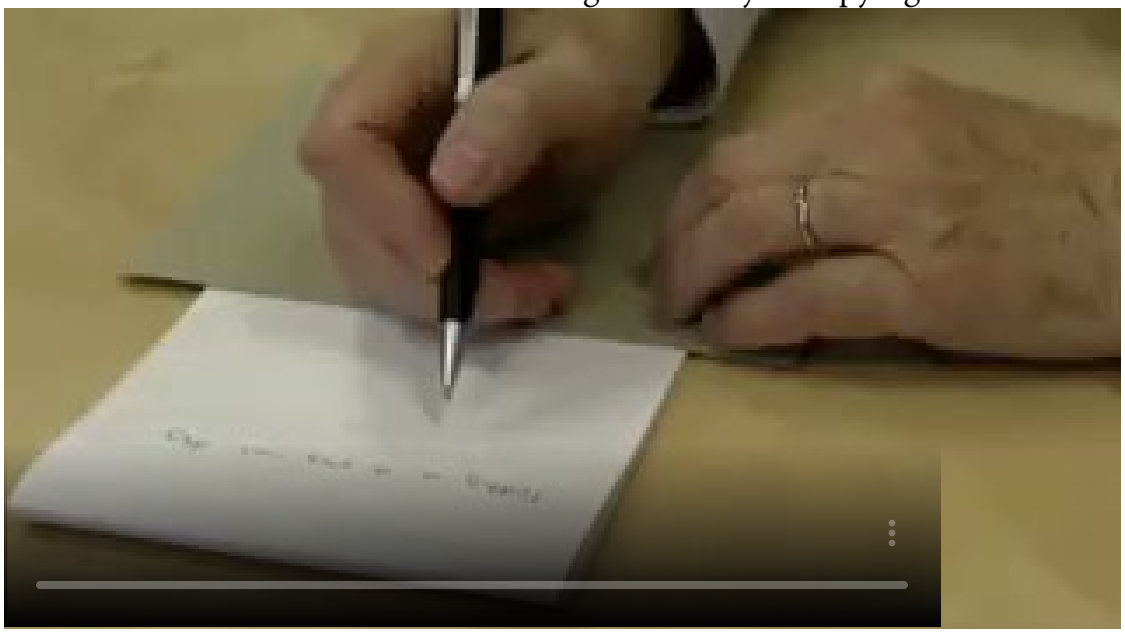
15.0 sec



Left Hand

The patient draws a line between the lines of the spiral from inside to outside without support of the hand and arm. The right and then the left hand are tested with two separate cartoons: the drawing is timed and any crossings of the given lines are counted with either hand.

MOVIE 3.2.11 Demonstration of testing the ability of copying a written sentence



MOVIE 3.2.12 Demonstration of tests to examine the patient writing (name and address)

MOVIE 3.2.13 Drawing spirals (normal)

MOVIE 3.2.14 Drawing spirals (abnormally slow)

MOVIE 3.2.15 Drawing spirals with action tremor

MOVIE 3.2.16 Parkinsonian resting tremor

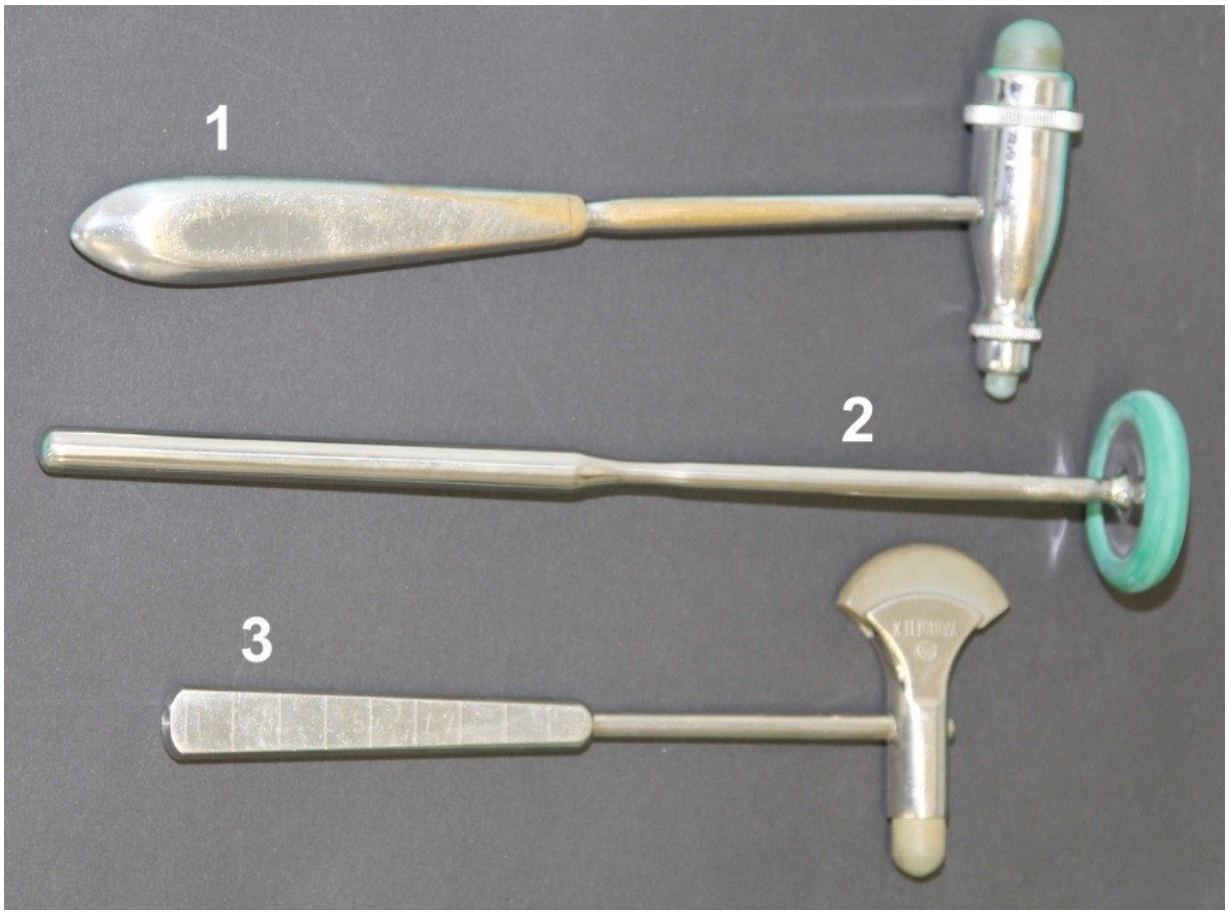
MOVIE 3.2.17 Action tremor (Essential tremor)

SECTION 4 – REFLEXES

General Terminology

Reflex hammer - There are various types of hammers around, some carry the names of respected institutions or of famous neurologists of the past (Lanska,1989) . A heavy weight hammer has the advantage of allowing a more reproducible tap.

FIGURE 4.4.1 Three types of reflex hammers



1 - Trömner hammer (Mayo hammer)
2 - Queen Square hammer
3 - Pediatric hammer
The Taylor hammer is not depicted.

Pyramidal signs - This denotes a **group of reflexes** that are positive when a suprasegmental or supraspinal (central) lesion exists. Unfortunately, the term is a **misnomer** as well-defined lesions of the pyramidal tract in animal experiments typically lead to flaccid paresis without "pyramidal" signs. The most important and sensitive is the Babinski sign **often combined** with the Chaddock reflex (Figure 4.1.3 d+ e; Figure 4.3.7; see Movie 4.3.3 and Movie 4.3.4). It is best elicited by a wooden spatula, or a thick wooden rod.

Monosynaptic (tendon) reflexes

Reflexes are elicited by using a **reflex hammer** tapping over the slightly tightened tendinous area close to the muscle insertion. Great care should be taken to tap repeatedly with the same intensity to end up with a clear result.

Note: reflexes are a critical aspect of the neurological exam as they serve to confirm suspected motor abnormalities (increased with upper motor neuron (UMN) lesion- associated weakness and decreased with lower motor neuron (LMN) or peripheral nerve lesion-associated weakness) and/or sensory findings. The appropriate patterns of pathology are a key to localization and differential diagnosis.

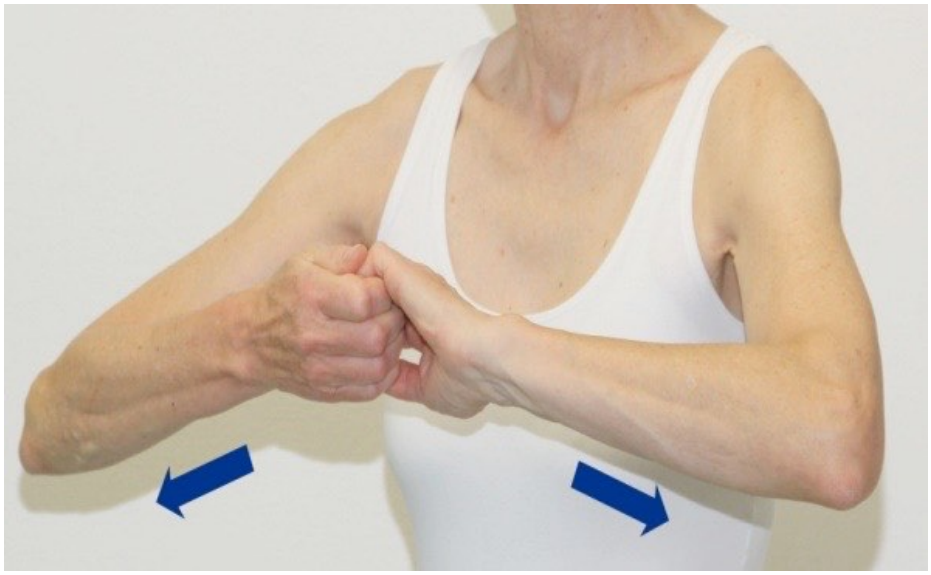
General points for testing reflexes

The reflex intensity (strength) in a normal person may be **variable** due to basic physiologic mechanism:

Effects of muscle coactivation: the "central motor drive" is enhanced, either by **slight voluntary innervation** of the limb tested or by an **overall increased muscle tone** (voluntary, anxiety-induced). Reliable results (see below) can only be obtained in a relaxed situation. Absent or marginally low tendon reflexes can be elicited if the patient voluntarily **coactivates** the reflex arcs by a strong jaw closure (as if biting on a spatula "clenching the teeth"; Movie 2.4.3), a strong contralateral fist closure, or similar other maneuvers such as the Jendrassik (Figure 4.1.1)

Position-dependant alterations: in **extensor** muscles reflex activity is **higher** in a **half-way extended** limb muscle while it is lower in **full** extension: i.e. the triceps brachii reflex or the quadriceps (patellar) reflexes are best elicited in a half-way **extended** upper arm position while sitting (cf. Figure 4.1.5) and in a half way extended thigh-knee position while the patient is in the **supine** position and the knee is bent at about **45° degrees**, best with **support** by a foam pillow. Conversely, in **flexor** muscles the tendon reflexes are best elicited in the **90° degrees** bended position.

FIGURE 4.1.1 Two manoeuvres to increase the central motor drive



Jendrassik manoeuvre (a)

and clench-your-teeth manoeuvre (b)

Further points

Each reflex is elicited **at least 3 times** to get an impression of its average strength, then the **same reflex** is tested on the **other side** and both are compared. Then, further reflexes are tested at the same limb until all reflexes have been tested and documented in a scheme (Figure 4.1.2 and Figure 4.1.3). The masseter reflex (see Section 2, Figure 2.4.5) may serve as a reference point for the limb reflexes; alone or in the context with other normal reflexes the masseter reflex allows defining an individual **reflex level** (low, intermediate, high). It is common experience that the reflex level **will go down** by about 1 grade after the patient has adjusted to the practice or hospital situation. It is impor-

tant to note both symmetry (side-to-side) and proportionality (upper-to-lower limb) of the reflexes, in order to **define normal** for any given patient.

Tendon-reflexes are called **abnormal** if they are

- very brisk with **clonic** responses, (**Grade 4+**)
- if there are clear-cut differences between **left and right**
- **more than** 1 grade lower activity in the lower limb than in the upper limb
- one single reflex is absent while the reflex level is at least grade 1 throughout.

The examiner may place his/her hand gently on the muscle that is expected to contract after tapping. This allows **feeling the reflex** response; especially the **dynamic** aspects of briskness may be much better appreciated than from only observing the response. It also tells the examiner whether the muscle is relaxed before tapping.

In hyperactive reflexes the muscle will contract briskly and the **area** from which the tap elicits a reflex is **widened**.

Clonus: a **rhythmic** jerking of the limb in which the jerk of the reflex always happens in the **same direction** as the reflex response (contrasted to myoclonus, which may be **non-rhythmic** and multidirectional). If reflexes are brisk there may be clonic contractions with or without gross limb movements. When tested in the foot the examiner holds and lifts up one leg at a time; then the foot is briskly bent up in the ankle joint and the number of rhythmic clonic movements (flexion-extension) are **counted**.

When eliciting the triceps surae reflex/Achilles tendon reflex (TSR/ATR), a clonus may follow the first jerk. In the knee joint, the patient is in the supine position while the examiner's arm is under both knees with both legs slightly elevated to an approximately 30° angle. Then the examiner lifts both legs up **abruptly**. This may cause an **exaggerated** extension (stretch) reflex or a **knee clonus** if a severe spastic paraparesis is present. The same may happen after eliciting the patellar reflex (PR) in this position.

Next, the complex **polysynaptic reflexes** are tested, all carrying names of the first describers, including the Babinski reflex (Figure 4.3.1, Figure 4.3.2, Figure 4.3.3, Figure 4.3.4, Figure 4.3.5, Figure 4.3.6, Figure 4.3.7 and Movie 4.3.1, Movie 4.3.2, Movie 4.3.3, Movie 4.3.4) and other pyramidal signs, and the cutaneous abdominal

reflexes (Figure 4.3.1) These are also documented in the scheme (polysynaptic reflexes).

Other pyramidal signs are considered less useful diagnostically than the Babinski sign, but may serve to complement the often "equivocal" toe response: Chaddock (running the stimulus along the edge of the lateral foot, can be **done together** (Figure 4.3.6) with the original Babinski reflex manoeuvre), Bing (pin prick to the dorsum of the big toe), Oppenheim (running knuckles or the hammer edge along the shin bone), Gordon (squeezing the calf). However, as noted in the Motor section (Table 3.2.1), **toe tapping may have a better sensitivity and specificity** to define upper motor neuron (**UMN**) lesions.

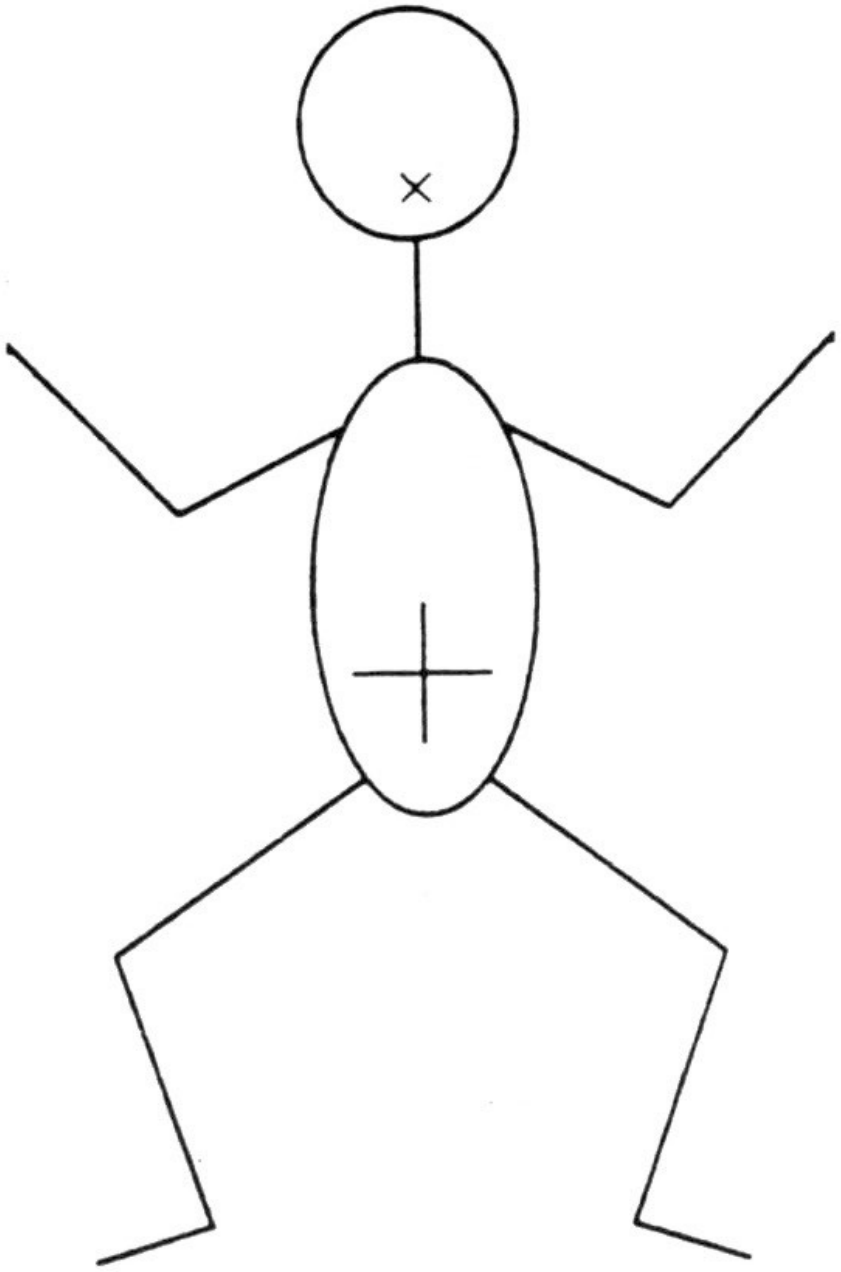
The **intensity of tendon reflexes** is **best expressed** and documented by using numbers of a widely used scoring system incorporated into a sketch (Figure 4.1.2); the following grades apply: as in the **MRC** grading section we must ***note here that there is poor INTER-rater reliability, but very good INTRA-rater reliability, and that is what matters in most clinical situations.***

0	not elicitable
0-1	absent on repeated tapping, present at low activity upon coactivation manoeuvres
1	weak response
2	intermediate response
3	mildly brisk reflex; some consider some spreading to adjacent muscles typical for grade 3
4	very brisk response; can be elicited even outside the usual tapping area; some consider spreading to the contralateral side of the body the point at which you

	can grade a 4. A few clonic after-contractions may occur in the foot
4+	very brisk response with longstanding clonus after a single tap over the Achilles tendon, some clonic after-contractions when eliciting the patellar and biceps reflexes, and tapping may even elicit neighbouring or contralateral reflexes (e.g. with the adductor (magnus) reflex, L 2.3)

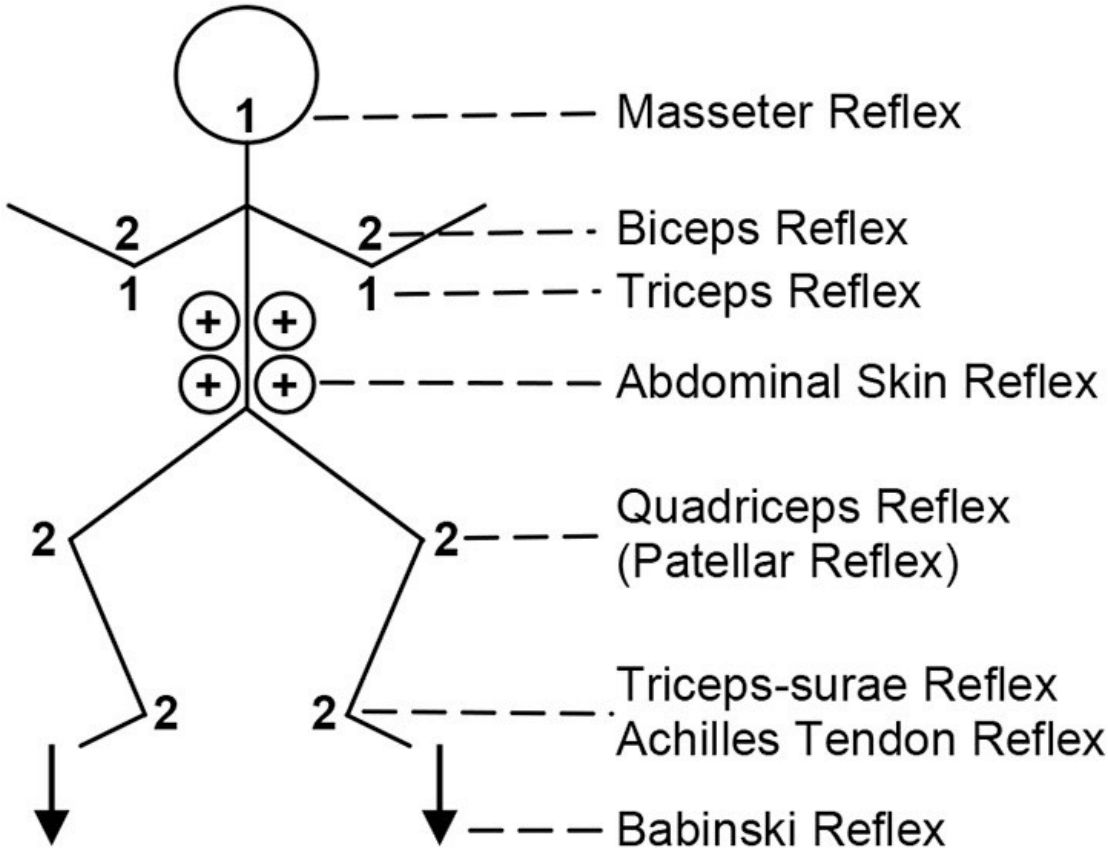
In the following drawings (Figure 4.1.3) the numbers of the reflex intensities are indicated. The Babinski (preferably plus Chaddock) reflex (Figure 4.3.6) is indicated with an arrow: an arrow pointing down (↓) is a normal down-going (a.k.a. flexor, plantar, "negative" Babinski) response while an arrow up (↑) indicates an up-going (a.k.a. extensor, "positive" Babinski) response (tonic or phasic/“stuttering” type also counts as a positive).

FIGURE 4.1.2 Reflex scheme

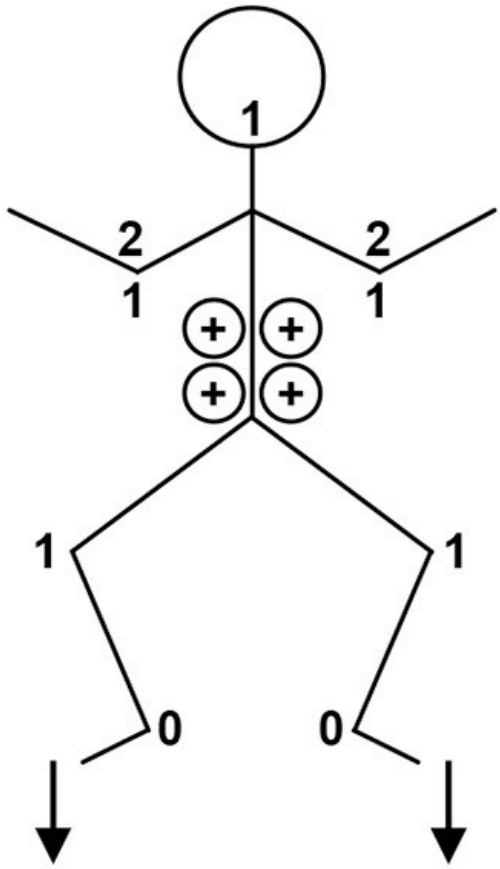


Reflex findings can be documented in this scheme/drawing and archived in the chart as shown in the examples below in Fig. 4.1.3. The cross indicates the 4 quadrants for the abdominal skin reflex.

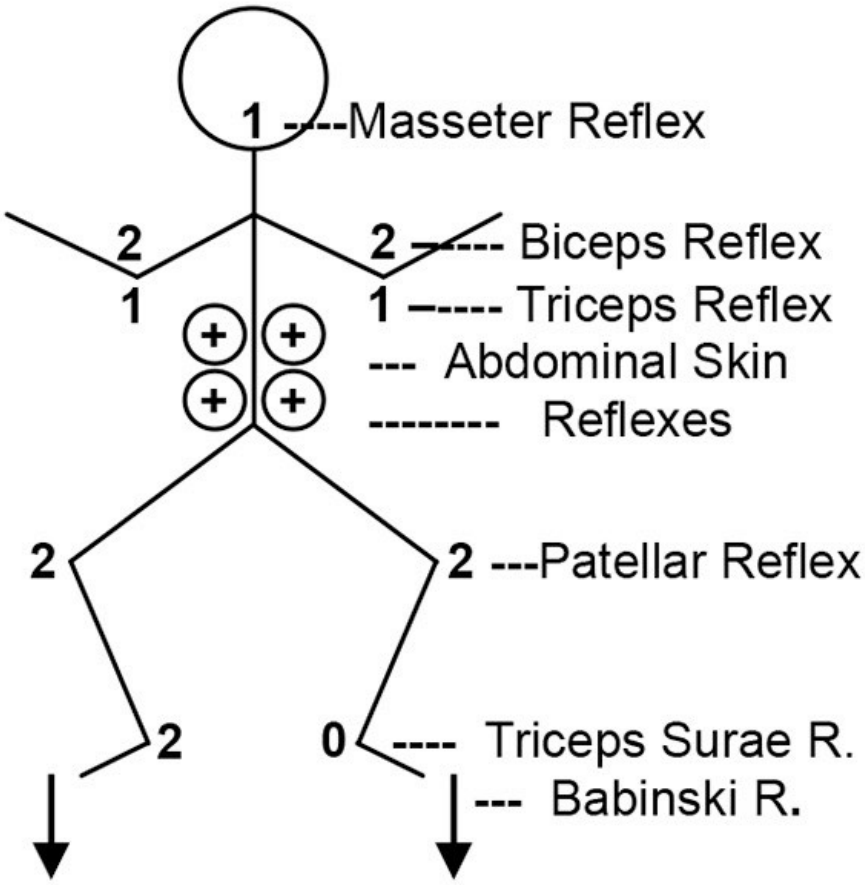
FIGURE 4.1.3 Types of reflex pathology in paradigmatic disorders



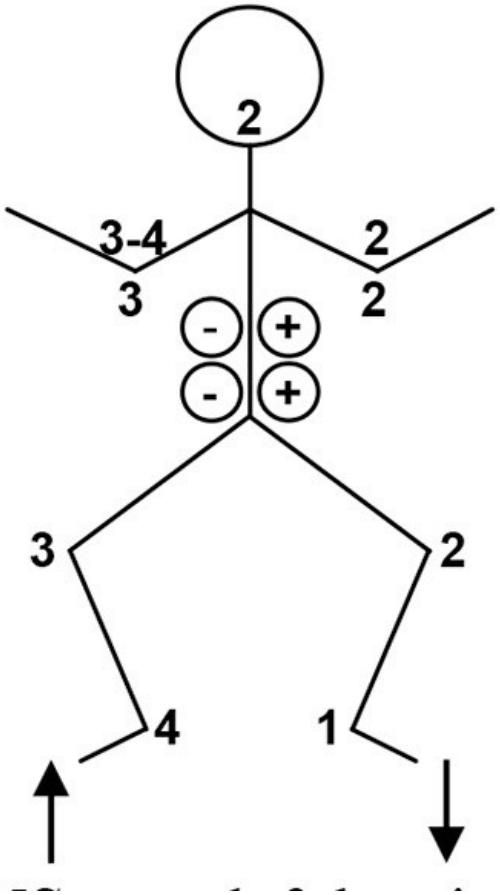
*A useful, simple drawing showing **normal** reflexes of intermediate strength. For grading see text.*



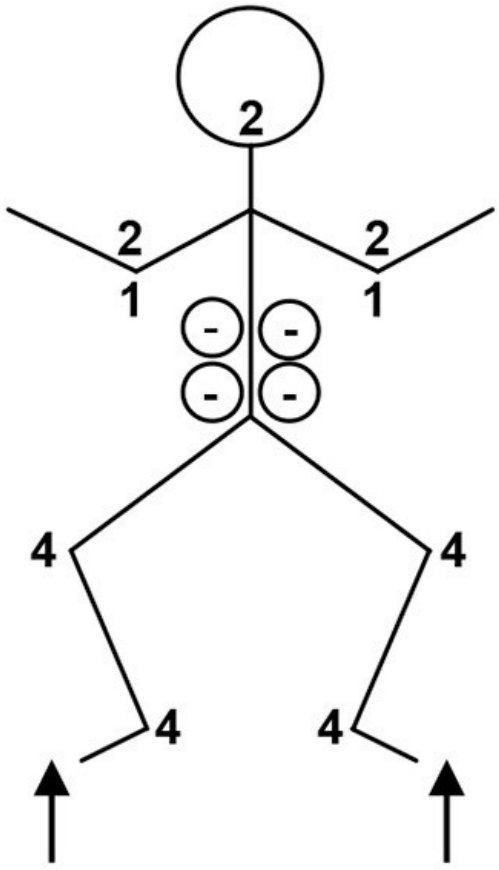
Polyneuropathy *typically results in distal reflex loss, due to the length-dependent process.*



Radicular-type - S1 root on left; *other reflexes of intermediate strength*



CNS-type *left hemispheric lesion., exaggerated reflexes on right*



Spinal-cord type *thoracic spinal cord, exaggerated reflexes below lesional segments*

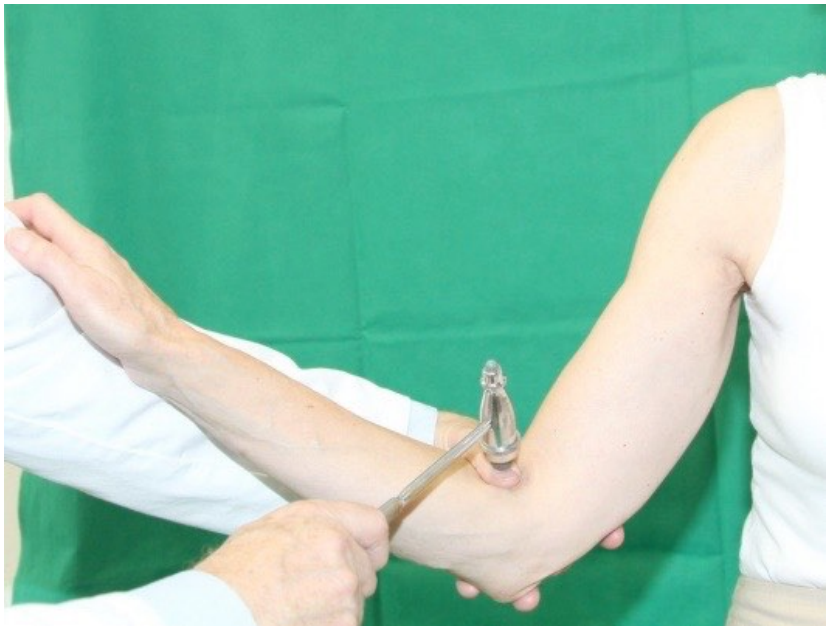
Monosynaptic reflexes in clinical practice

(associated segments of the spinal reflex arc are indicated; main segments **are in bold**)

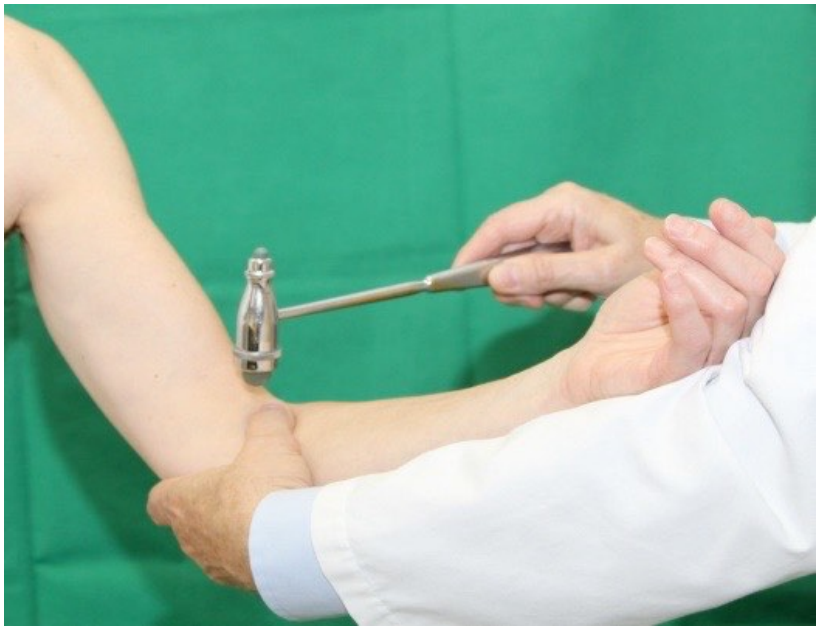
Note that main spinal cord segments may vary in different persons.

- **Biceps brachii muscle reflex** (BR, C5, **C6**): In the sitting or standing position the arm is bent at 70-90°; the examiner puts his/her finger on the biceps tendon and taps on the finger to elicit the reflex. **Note** that the positioning **is different for left and right.**

FIGURE 4.1.4 Biceps brachii reflex



on right (a)



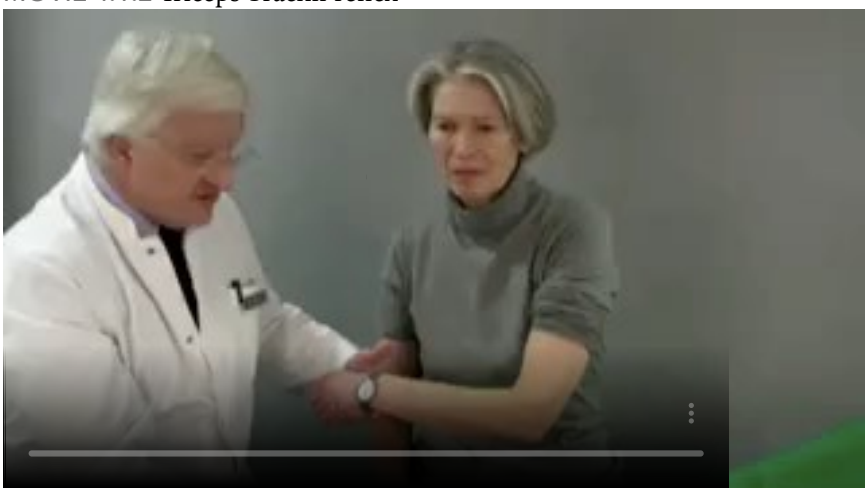
and on left (b) (Trömner hammer)

MOVIE 4.1.1 Biceps brachii reflex with teeth clenching

- **Brachioradialis muscle reflex** (BRR, C5, **C6**): In the same position as with the BR the distal end of the radial bone is tapped. If reflexes are brisk (grade 3-4) the BR will be coactivated when tapping the radial bone.
- **Triceps brachii muscle reflex** (TR, C6, **C7**, **C8**): The arm is positioned in half way extension with the palm over the belly. The triceps tendon is tapped just above the olecranon. Alternatively, one may hold the patient's arm up lying on the examiners arm in a pronated position and the tendon is tapped in this position.

FIGURE 4.1.5 Triceps brachii reflex – two different modes of eliciting

MOVIE 4.1.2 Triceps brachii reflex



- **Trömner reflex (C7, C8)**: This is partly a monosynaptic and partly an oligosynpatic reflex. The examiner supports the hand of the patient in pronation with his/her own left hand (right hand in left-handers) **just below the metacarpophalangeal joint/most proximal phalanx** (Figure 4.1.6 a,b). Then the examiner makes an abrupt flexion movement with his/her fingers in order to elevate (extend) the patient's fingers. The reflex response is a flexion movement of the fingers (the monosynaptic part) and of the thumb. If the reflex level is increased to grades 3-4, the long fingers bend more strongly and even the wrist may flex. A visible flexion movement of the thumb or fingers and thumb is called "Trömner positive" and an arrow (↓) sign may be included in the **reflex scheme** shown above (Figure 4.1.2). Right to left differences in intensity of the reflex movement are considered abnormal signs.
  A **variant** is performed by placing the patient's hand on a table or on the patient's own thigh above the knee, placing the examiners index finger sideways on the proximal phalanx of the gently flexed fingers of the patient's hand and tapping the examiners finger with a **reflex hammer**. The appropriate response is flexion equivalent to the standard Trömner reflex (Figure 4.1.6 c,d).

FIGURE 4.1.6 Trömner reflex – two modes of eliciting

(a) starting position. Arrows illustrate the reflex response.

(b) the reflex response. Arrows illustrate the reflex response.

Variant: (d) starting position.

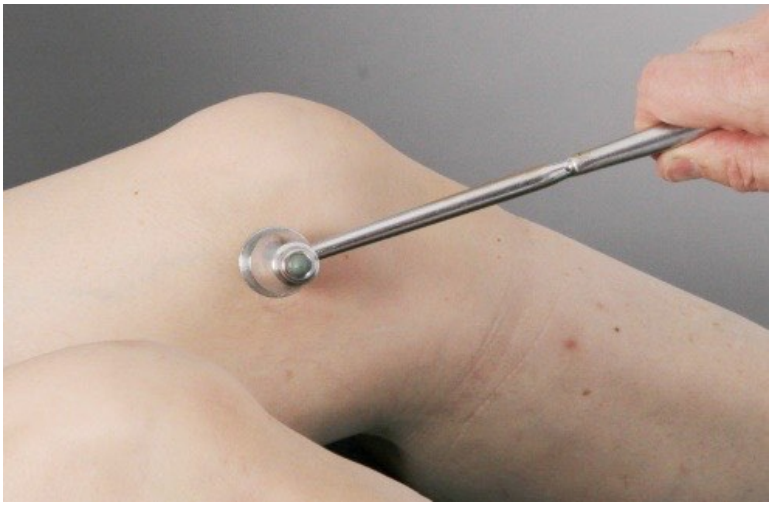
(b) the reflex response

MOVIE 4.1.3 Trömner reflex

MOVIE 4.1.4 Trömner reflex

- **Adductor (magnus) muscle reflex (L2, L3,** L4): The examiner puts his/her finger on the adductor magnus tendon just above the knee and taps gently on the medial epicondyle to elicit the reflex. With brisk reflexes (Grade 4 and 4+) the contralateral reflex is often coactivated.

FIGURE 4.1.7 Adductor (magnus) reflex – two modes of eliciting



The epicondyle is tapped;

In case of tenderness the hammer may tap on the interspersed finger of the examiner.

- **Patellar (Quadriceps muscle) reflex** (PR, L2**, L3, L4**): The reflex can be elicited in the sitting position by tapping the patellar tendon just below the patella. Alternatively and with a **heightened** strength, the reflex is elicited in the supine position with the knees flexed at about **135° while supported** by a cushion (Figure 4.1.8). The other hand rests on the thigh appreciating the briskness of the reflex. The reflex is elicited repeatedly to find define its strength (Grades 0-4). If the patellar reflex gets stronger with each subsequent tap, this is a typical in the Lambert-Eaton mysthenic syndrome (Movie 4.1.8).

FIGURE 4.1.8 Patellar reflex elicited with knee supported at a 135° angle

MOVIE 4.1.5 Patellar reflex

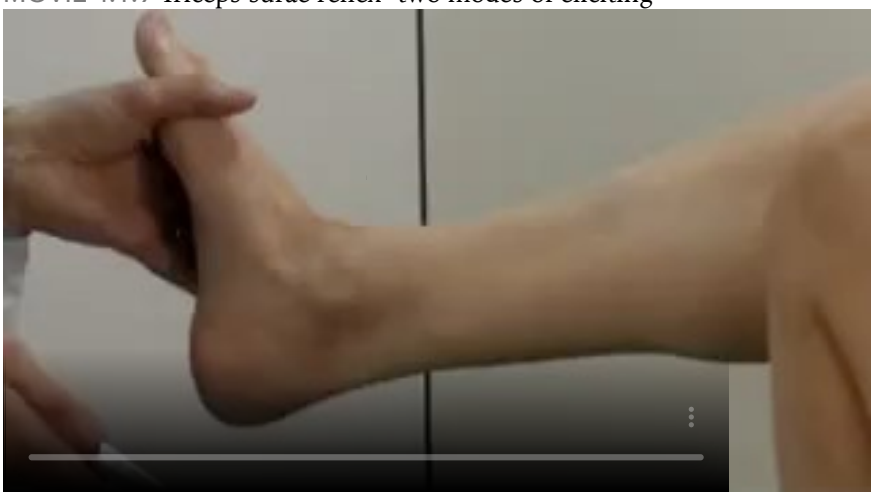
MOVIE 4.1.6 Patellar reflex (sitting) with teeth clenching

MOVIE 4.1.7 Patellar reflex (supine) with teeth clenching

MOVIE 4.1.8 Conditioning of PSR in Lambert-Eaton myasthenic syndrome

- **Triceps surae Reflex** (TSR; syn. Achilles tendon reflex; ATR, **S1, S2**): In the supine position one leg is bent in the knee joint and the foot placed on the outstretched contralateral leg. The examiner holds the foot at a **90° angle** (this implies a slight dorsiflexion in relation to the natural relaxed foot posture) and taps the Achilles tendon. If the reflex cannot be elicited in this way the patient is asked to **kneel** on the stretcher holding his outstretched arms against the wall. Now the Achilles tendon is tapped while the examiner holds both feet at a **90° angle** with his/her thighs/knees.

MOVIE 4.1.9 Triceps surae reflex- two modes of eliciting



SUBSECTION 2

Additional monosynaptic reflexes

(useful when testing for specific spinal segments of interest; main segments are in **bold**)

- **Deltoid muscle reflex (C5)**: This reflex can be elicited in the sitting and resting position by tapping the humerus bone on the lateral epicondyle in the resting position (Figure 4.2.1). At a low reflex level the reflex may not become apparent but may do so if the patient clenches his/her teeth (Figure 4.1.1 b).

FIGURE 4.2.1 Deltoid muscle reflex

- **Pectoralis Reflex** (C5, C6, **C7, C8**, T1): The examiner places his/her index finger on the tendon of the pectoralis major which can be felt below the axilla. The reflex can be elicited by tapping on the index finger or directly near the tendon. This reflex can sometimes be anatomically more informative if the responses to tapping the upper part (C5-7) of the tendon are

compared with those elicited by tapping the lower part (C8-T1) (Figure 4.2.2). The reflex strength is usually less than with the classic tendon reflexes.

FIGURE 4.2.2 Pectoralis reflex (normal)

upper segment (a)

middle segment (b)

MOVIE 4.2.1 Pectoralis reflex

- **Tibialis posterior reflex (L5)**: In the supine position the leg is bent in the knee joint and placed over the contralateral leg. Now the patient is asked to lift the foot slightly against the examiner's resistance into supination. The examiner taps the **posterior tibial tendon** just behind the medial malleolus or a bit more distally (Figure 4.2.3). In some healthy humans this reflex cannot be elicited (see Movie 4.2.2). It is judged as abnormal only if there is unilateral reflex loss (e.g. in lumbar disk protrusions with compression of the L5 root) or if the reflex is very brisk on both sides. With abnormal muscle weakness in the L5/peroneal nerve -innervated muscles an absent reflex helps to argue against a common peroneal nerve disorder but rather supports as L5 root problem.

FIGURE 4.2.3 Tibialis posterior reflex

The foot is actively but very gently supinated (here elevated) to increases the central motor drive.

MOVIE 4.2.2 Tibialis posterior reflex

- **Abdominal muscle reflexes** (T 6-12): In the supine position the examiner taps the lower rib cage (image); then the hand and index finger are placed on the middle of the belly and finally above the hip - here the examiner taps the index finger of his/her own hand (Figure 4.2.4). This reflex will only be present if the reflex level is high (grades 3-4) but can also be released using the clench-your-teeth manoeuvre (Figure 4.1.1 b). If there is a transverse spinal lesion these reflexes may help to establish the segmental level of the lesion.

FIGURE 4.2.4 Abdominal muscle reflex – monosynaptic-plurisegmental

upper section by tapping on the rib cage (a)

middle section tapping on abdominal muscles, lateral to the umbilicus (b)

- **Anal sphincter reflex (S3-S5)**: This exam is done if there is a suspected **sacral cord or a pelvic peripheral nerve lesion**,

e.g. after spinal cord trauma or a fracture of the pelvis. The test is done as an anal digital examination. After the **patient is informed** about this painless procedure and the need for it, the examiner wears two superimposed latex gloves and, with a good amount of a soft crème, inserts his/her index finger first gently, then moves the finger back and again inserts it quite abruptly into the anal foramen. As a result the anal sphincter muscle will contract and this can be felt through the examiner's finger. A **unilateral decrease** in muscle contractioncan also be appreciated.

This reflex testing should **not be done** if there are painful hemorrhoidal masses or local inflammatory changes. As with all ano-genital testing a witness should be around.

SUBSECTION 3

Polysynaptic reflexes

The reflex arc may include several spinal segments and intersegmental spinal pathways. Some of these reflexes have a **tendency to habituate** on repeated testing.

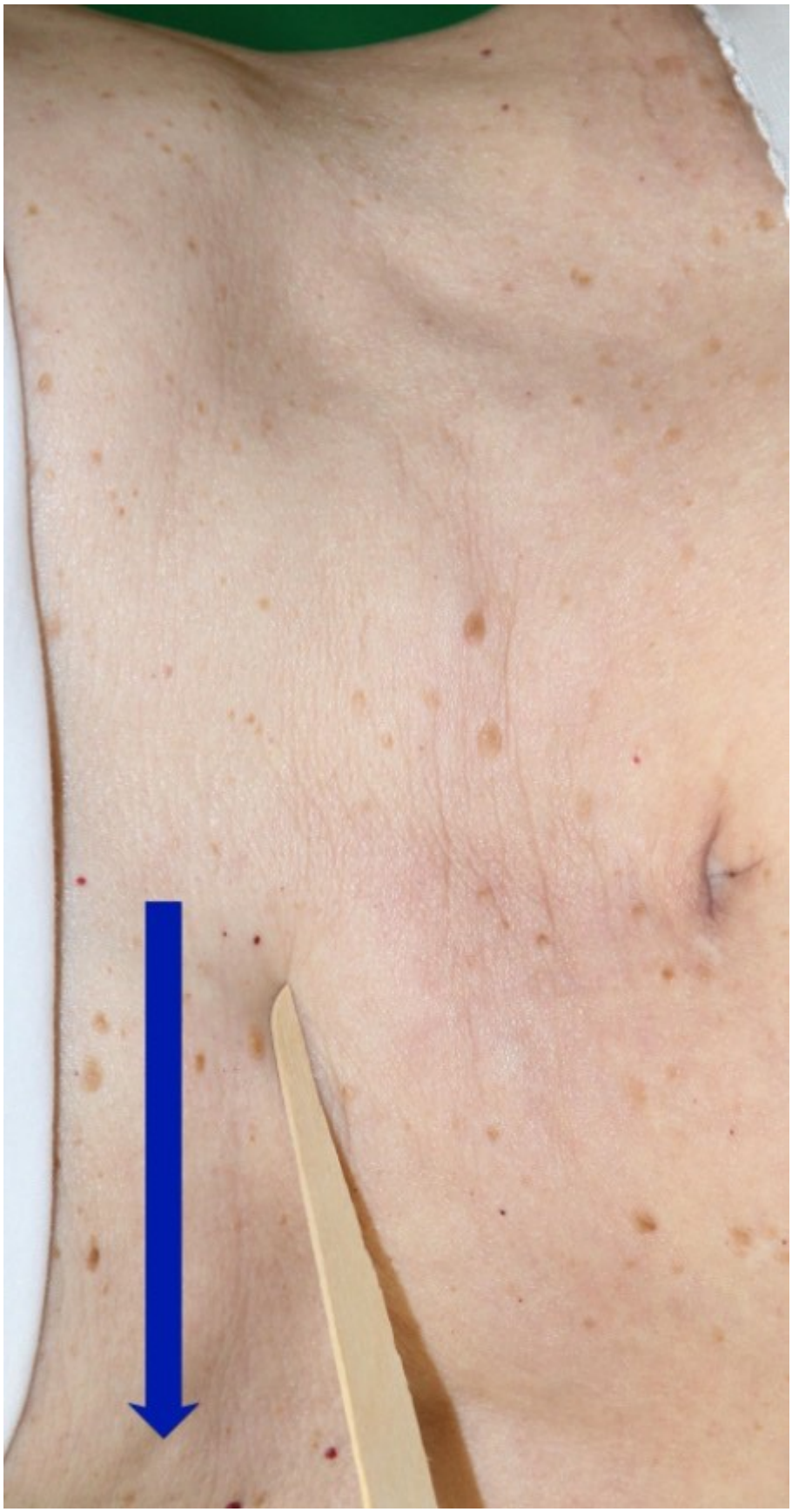
Diagnostically important polysynaptic reflexes include the following:

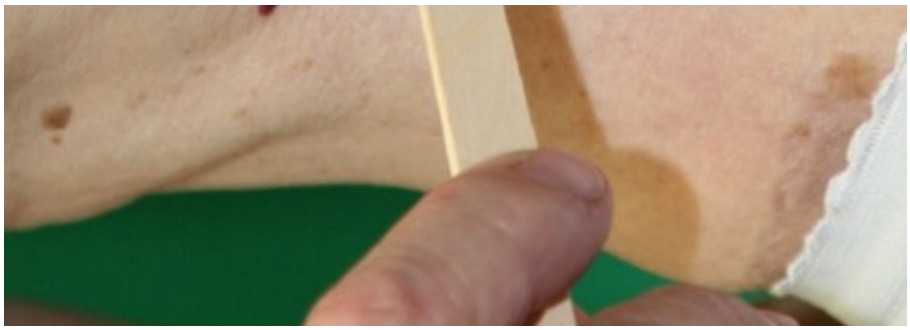
Corneal reflex (afferent CN V1, efferent CN VII): (see section 2, Figure 2.4.3)

Gag reflex (afferent CN IX, Efferent CN X): (see section 2)

- **Abdominal skin reflexes (T6-T12)** (Figure 4.3.1 and Movie 4.3.1): A broken spatula or a wooden rod is used to rub over the belly from midline to lateral or in reverse order rather gently but swiftly. This should be done at two or three levels: below the rib cage (approximately at the level of the dermatome T8), at the navel (T10) and if needed above the hip (T12). If the reflex is absent in one segment it is likely that the patient suffers from a spinal cord process above the segment of the reflex loss. This reflex may be very week or even absent in healthy people. It is prone to adapation, i.e. it may go away after a single reflex response.

FIGURE 4.3.1 Abdominal skin reflex

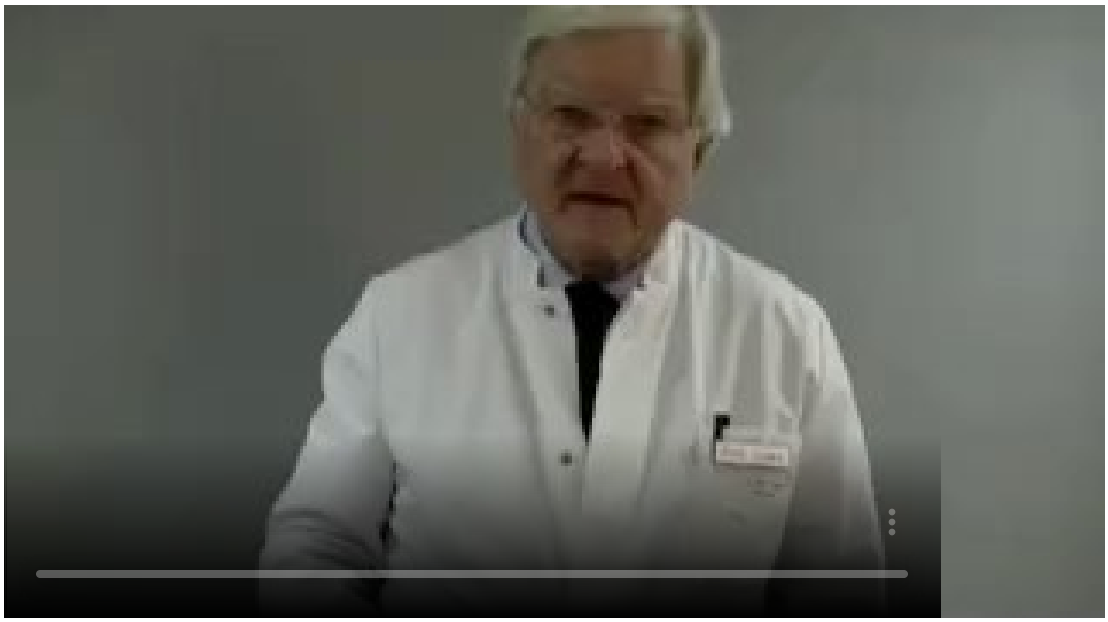




(a) upper segment

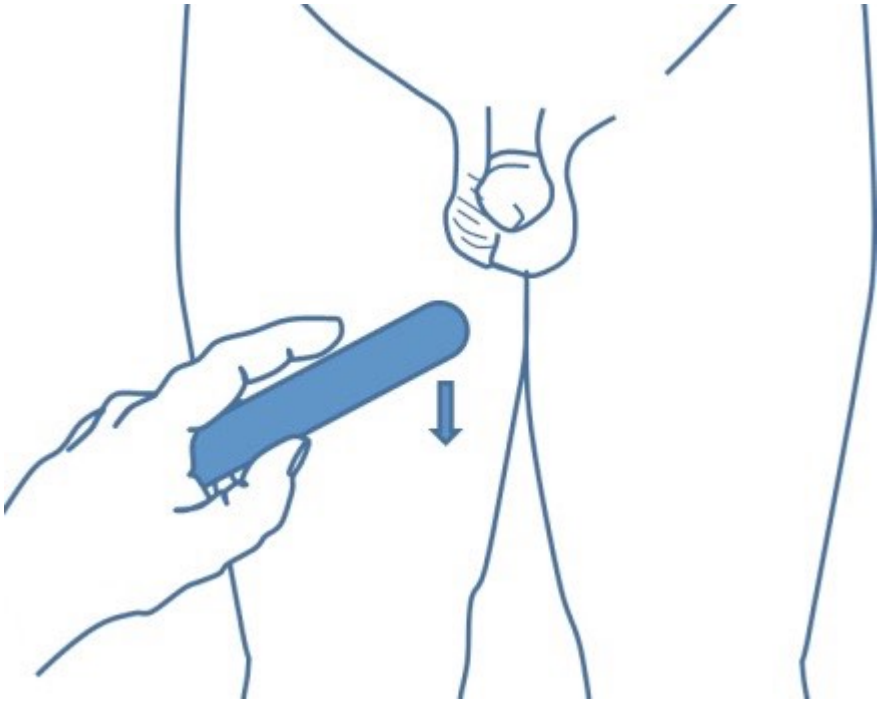
(b) middle segment. The broken spatula is best moved swiftly and gently from inside to outside or in the reverse direction.

MOVIE 4.3.1 Abdominal skin reflex



- **Cremasteric reflex (L1, L2,** L3): The reflex is elicited by moving a spatula swiftly down the medial aspect of the thigh distal to the inguinal crease. The reflex response is a **contraction of the cremaster muscle** with prompt **elevation of the testis** on the ipsilateral side. In addition, the ipsilateral **skin of the scrotum** may contract (Figure 4.3.2). This reflex is lost in sacral cord, medullar conus disease and with peripheral nerve injuries. As with all ano-genital reflexes a witness should be around.

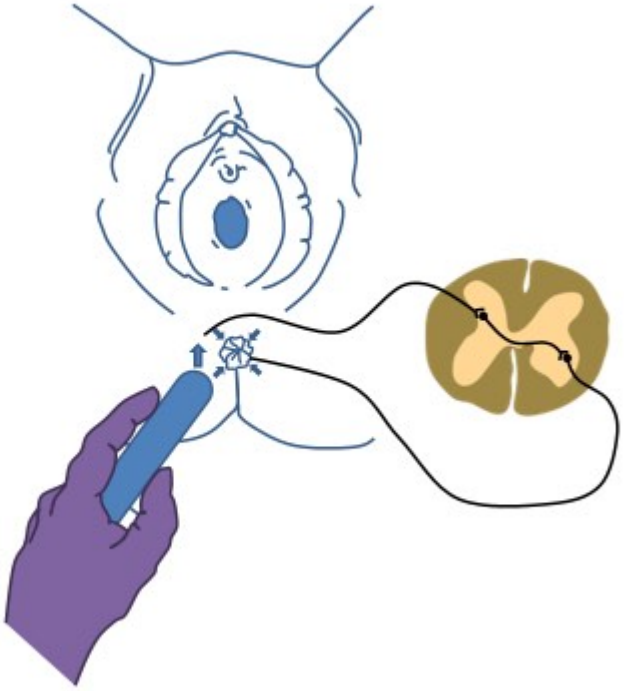
FIGURE 4.3.2 Cremasteric reflex



The ipsilateral scrotum and the levator muscle contract (hatched area).

- **Anal skin reflex** (S2, **S3-S5**): The patient is examined in the sideward position away from the examiner with the hip and knee joints bent (Figure 4.3.3). After informing the patient the reflex is elicited with a wooden rod by gently scratching the perianal skin. The reflex response is a contraction of the sphincter muscle. This reflex is lost in sacral cord or medullar conus disease but occasionally also in healthy individuals. A unilateral loss is always abnormal.

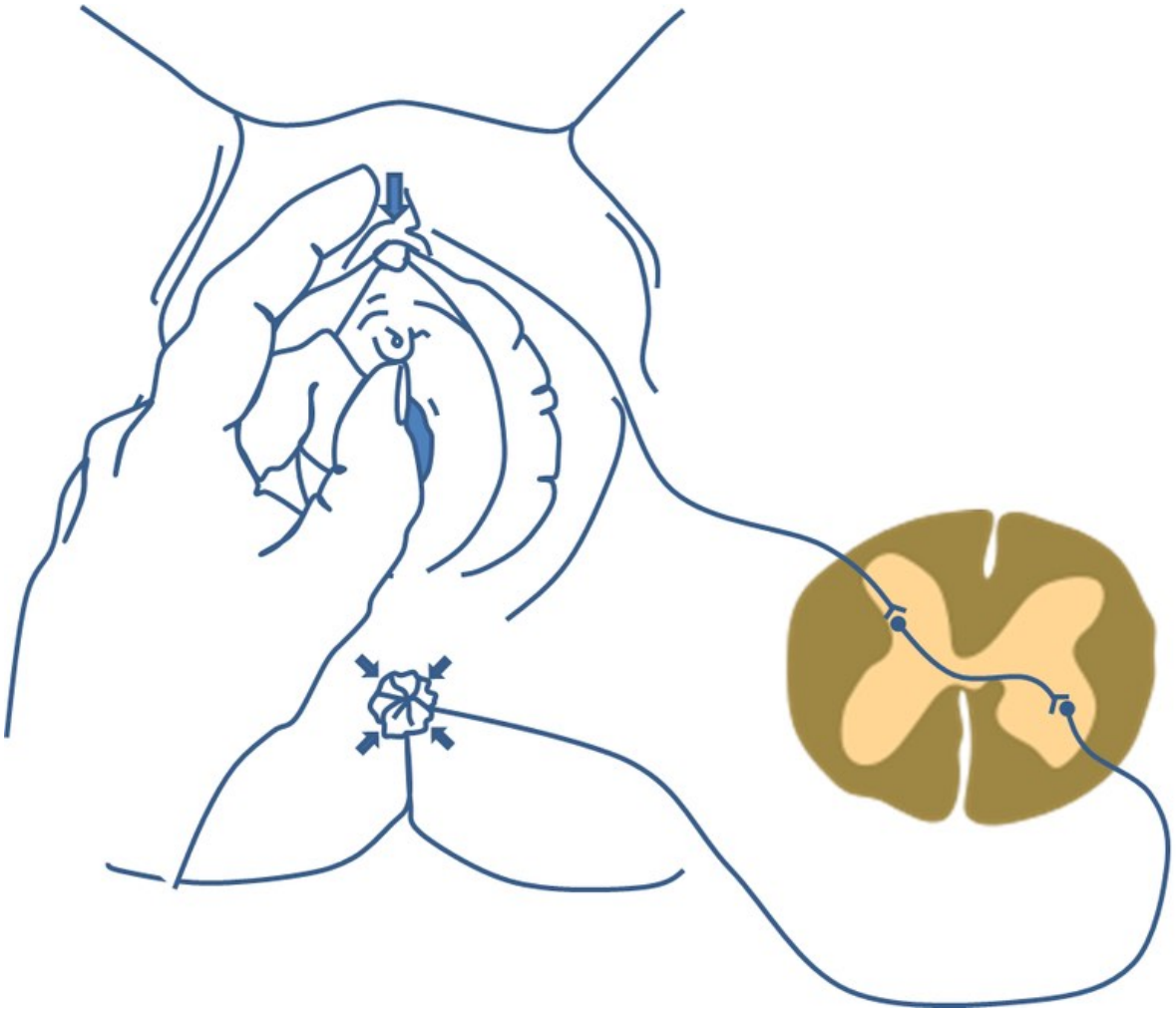
FIGURE 4.3.3 Anal skin reflex



The broken spatula is moved gently and swiftly as indicated by the arrow along the skin lateral to the anus.

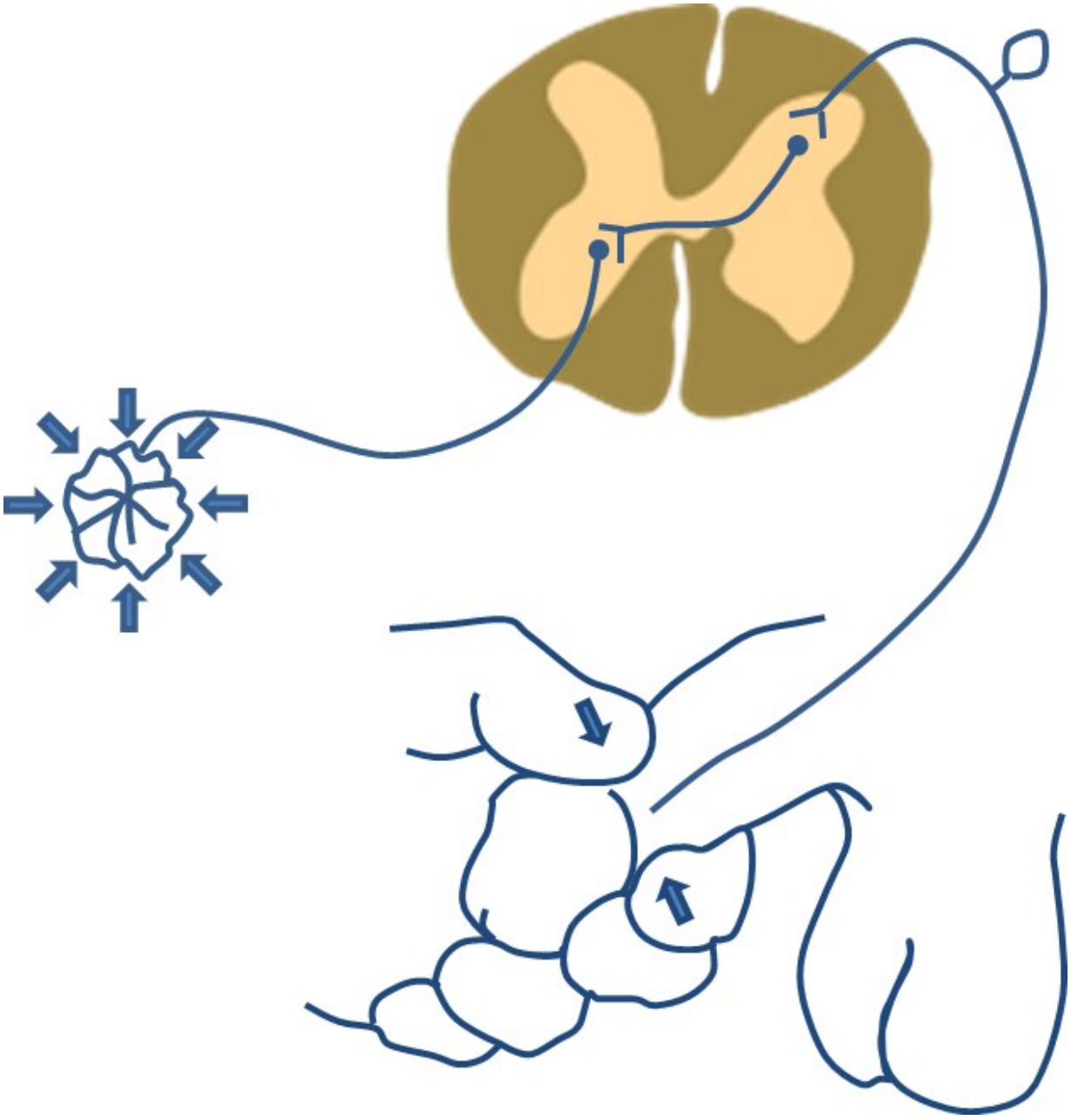
- **Bulbocavernosus reflex** (S2-S4): The patient is precisely informed about the procedure and its need, and is specifically asked for **consent to touch the genitalia** (Figure 4.3.4 and Figure 4.3.5). It is advised that a second professional is witnessing the examination.
  Absence could indicate a lesion or injury of the conus medullaris, sacral nerve roots, and perineal peripheral nerves. If traumatic spinal cord injury is being investigated in a patient with acute paralysis, the absence of the reflex indicates spinal shock, whereas the presence of the reflex suggests cord severance. A common reason for its temporary or even permanent absence is a pressure lesion in the perineal region, e.g. during prolonged bicycling.

FIGURE 4.3.4 Bulbocavernosus reflex in females

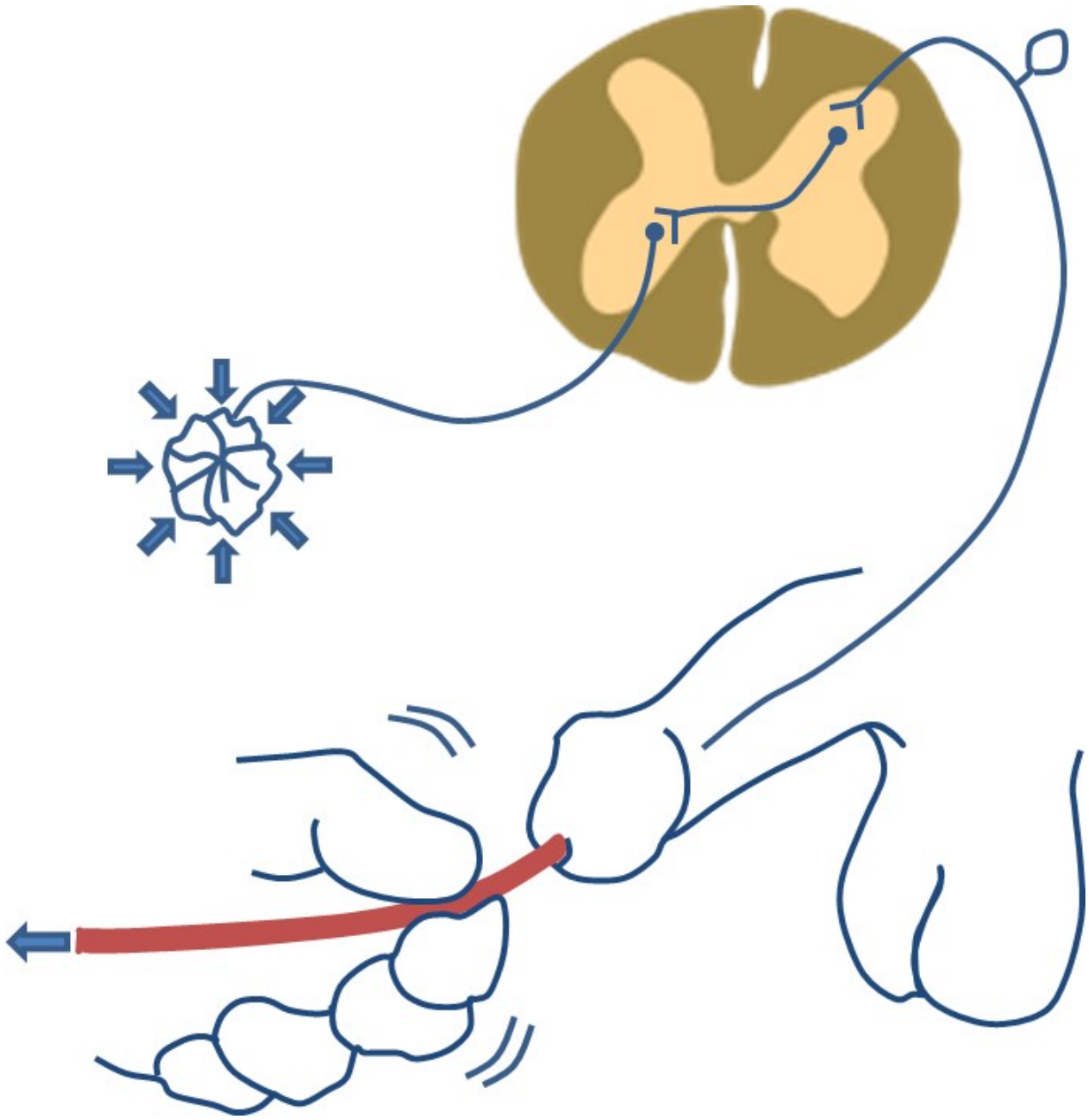


The examiner's hand gently compresses the clitoris. The reflex response is a contraction of the anus.

FIGURE 4.3.5 Bulbocavernosus reflex in males



Test #1: The examiner's hand gently compresses the glans penis. The reflex response is a contraction of the anus.



Test #2: The reflex is tested when a catheter is being exchanged. The examiner moves an inserted catheter outwards. The reflex response is a contraction of the anus.

The Babinski reflex group

Of special importance is the **plantar response** described by Babinski: According to JM Lance "the Babinski sign is an indication of withdrawal of supraspinal control of flexor reflexes in the lower limbs... It is associatet ... with inactivation, transient or permanent, of the upper motor neuron, which term implicates cortico-reticu-

lospinal fibres as well as the pyramidal tract anywhere in its path from cerebral cortex to termination in the cord." (Figure 4.3.6)

The Babinski reflex is best elicited in a **modification**. The Babinski mid-sole stimulus is applied together with another stimulus at the lateral part of the sole (also called Chaddock reflex if done alone) using two not-too-sharp wooden rods or the two broken pieces of a wooden spatula, which are **discarded** after examining the patient (Figure 4.3.7 and Movie 4.3.4).

One should take care that the patient can **tolerate** the manoeuvre and does **not have a withdrawal reaction** due to tickling or pain where all toes may go up and the patient may even pull back the entire limb.

Note: a patient with severe spasticity may typically pull back the entire limb in flexion as an exaggerated reflex response together with an upgoing big toe. This may be elicited with only mild pressure of the spatula.

FIGURE 4.3.6 Babinski reflex response

(a) the response is "downgoing toes", i.e. a negative Babinski (sign) , a normal response

(b) the response is an "upgoing toe" with a slight spreading of the other toes, a "positive" Babinski sign, an abnormal response.

The **Babinski response** is "positive" (abnormal) if the toes go up either tonically or in the form of a few dorsal extension movements ("stuttering" movements). The other toes may show spreading movements. The expression "upgoing toes" is preferred. The finding is documented by a vertical arrow pointing upwards in the scheme

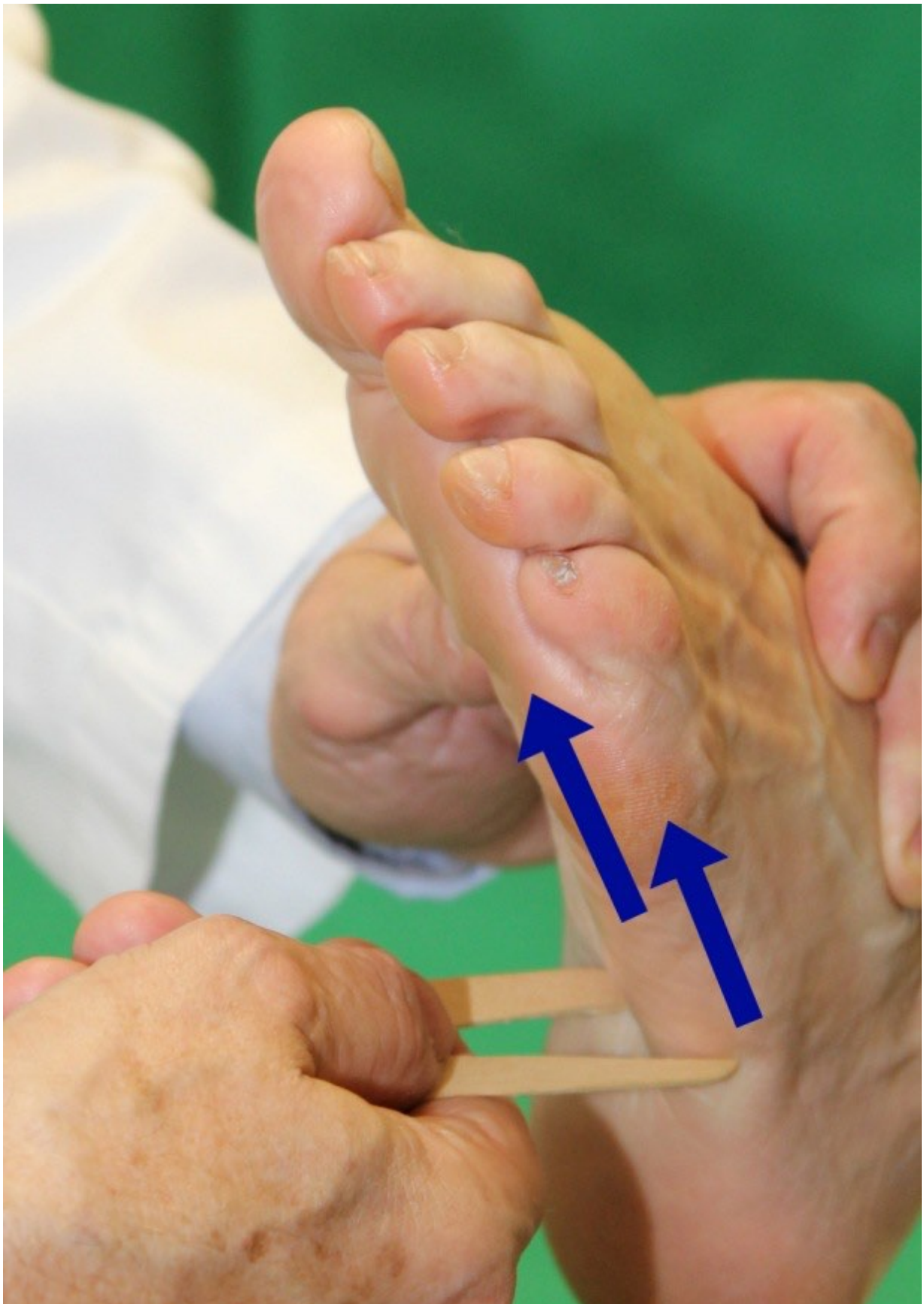
shown in Figure 4.1.3. A "normal" Babinski response is best termed "downgoing toes": a vertical arrow pointing downwards in the reflex scheme (Figure 4.1.3).

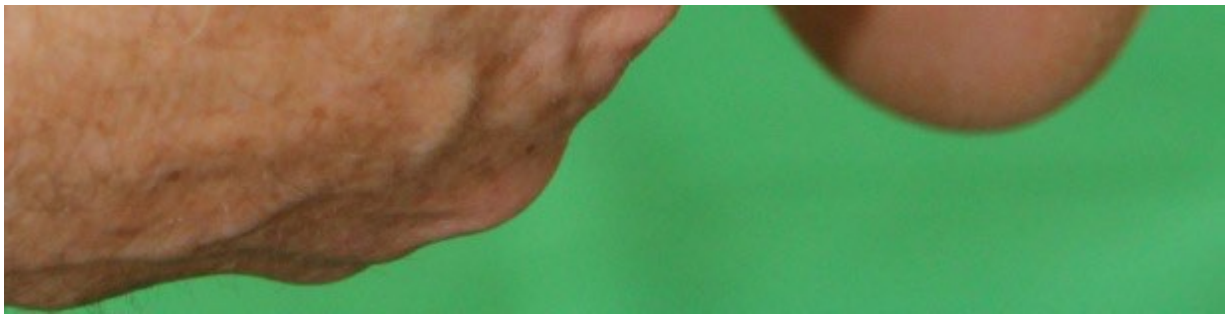
MOVIE 4.3.2 Babinski reflex

The **Chaddock reflex** is essentially a variant scratching the lateral side of the sole (Movie 4.3.3), but the **simultaneous** plantar and lateral skin stimulation seems the best approach (see above). Alternative but less specific manoeuvres include the Oppenheim reflex (rubbing the skin over the shinbone down from the knee), or the Gordon reflex (massaging the calf muscles) (Goetz, 2002). Both Oppenheim and Gordon reflex testing, when properly done, typically cause more discomfort to the patient than eliciting the Babinski response. Occasionally they may be useful if the Babinski response is in doubt or if the patient cannot tolerate the Babinski manoeuvre.

MOVIE 4.3.3 Chaddock reflex

FIGURE 4.3.7 Simultaneous stimulation of the Babinski (medial aspect) and the Chaddock reflex (lateral aspect) with broken spatulas





MOVIE 4.3.4 Simultaneously eliciting Babinski (medial) and Chaddock (lateral) reflex with broken spatulas

Because of the highly discriminating value of the plantar response for the diagnosis of upper motor neuron pathology only **unequivocal** responses should be accepted. Otherwise **re-testing** is needed, best with allowing a time interval of at least a **couple of minutes to reverse habituation** of the response. If in doubt the reflex is tested several times.

Note: the Babinski reflex group is prone to habituation. In clinical practice, a unilateral positive Babinski sign - up-going toes - most often indicates pathology of the CNS above the neck.

SECTION 5 – COORDINATION (MOTOR SYSTEM B)

General Terminology

The principal motor function of the cerebellum is the fine tuning of learned motor control. (Some aspects of coordination have already been displayed in **sections 2 and 3).**

Note: "coordination" skills are overlapping with "motor" skills. Abnormalities can only be interpreted as such if any of the manoeuvres can be performed, if the patient has no major motor impairment, and if vision is not impaired.

Dyssynergia

Dysmetria

Ataxia

Postural tremor

Kinetic tremor, action tremor, and **intention tremor**, as defined in section 3, can equally be considered as parts of the coordination realm (see section 3).

For testing coordination of eye movements, speech and other cranial nerve functions please refer to section 2.

Note: certain non-organic (functional) pathologies within the complex conversion disorders (section 9) will have inadvertent resumption of their strength when being tested for coordination, re-emphasizing the importance of not just the examination but observation thereof.

Finger-nose and finger-nose-finger tests

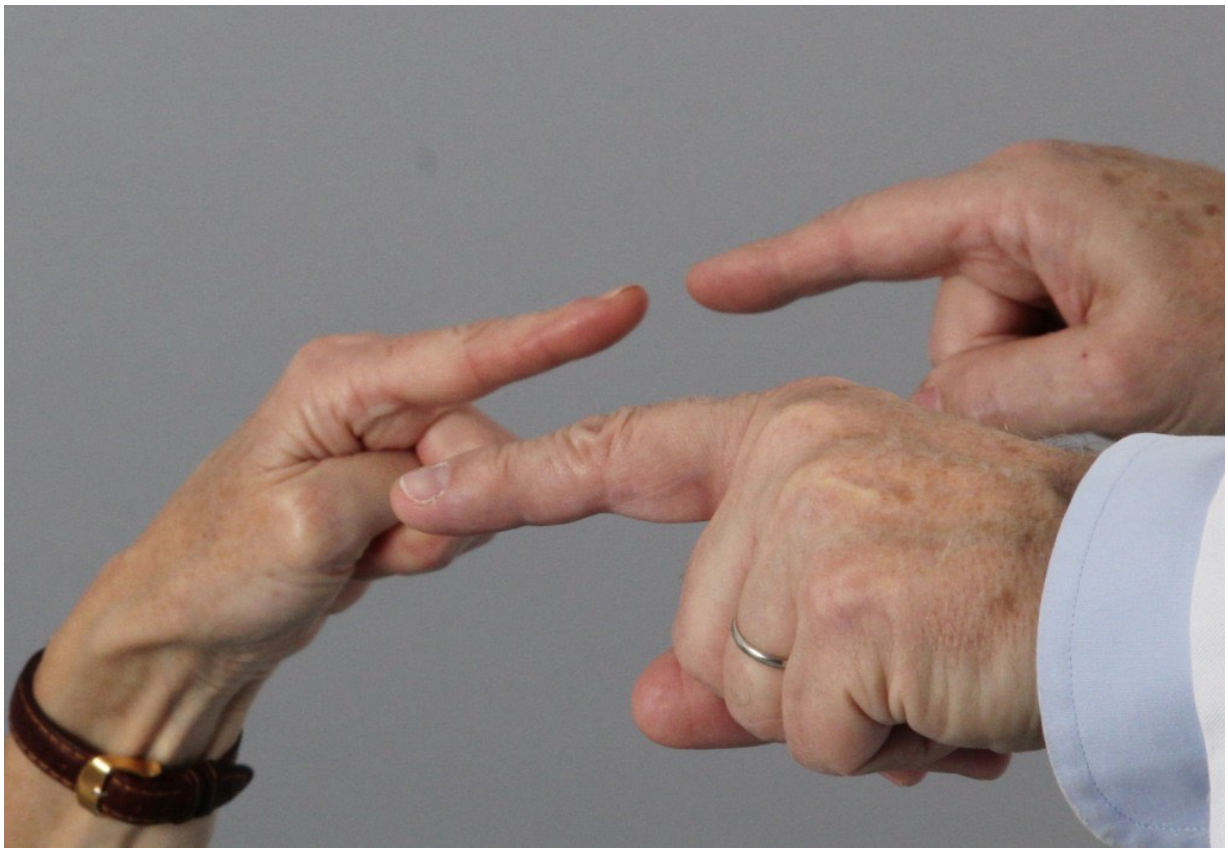
The patient is asked to perform an arm-and-hand movement with the aim of touching or almost touching his/her nose with his/her index finger; this is first performed with **open eyes** and then with **closed** eyes.

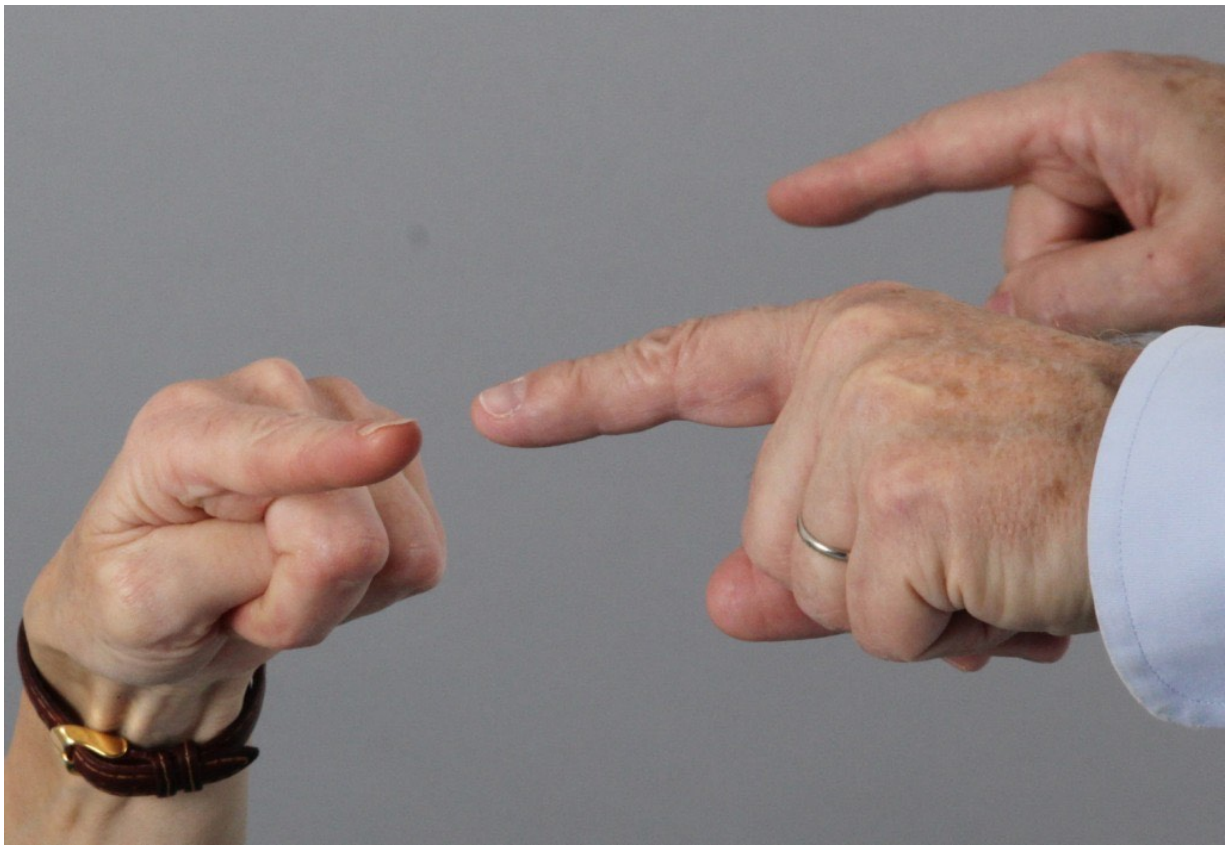
FIGURE 5.1.1 Finger-nose test

the patient reaches his/her nose precisely with open (a)

in (b) the patient's finger overshoots and misses the nose

FIGURE 5.1.2 Finger-finger test

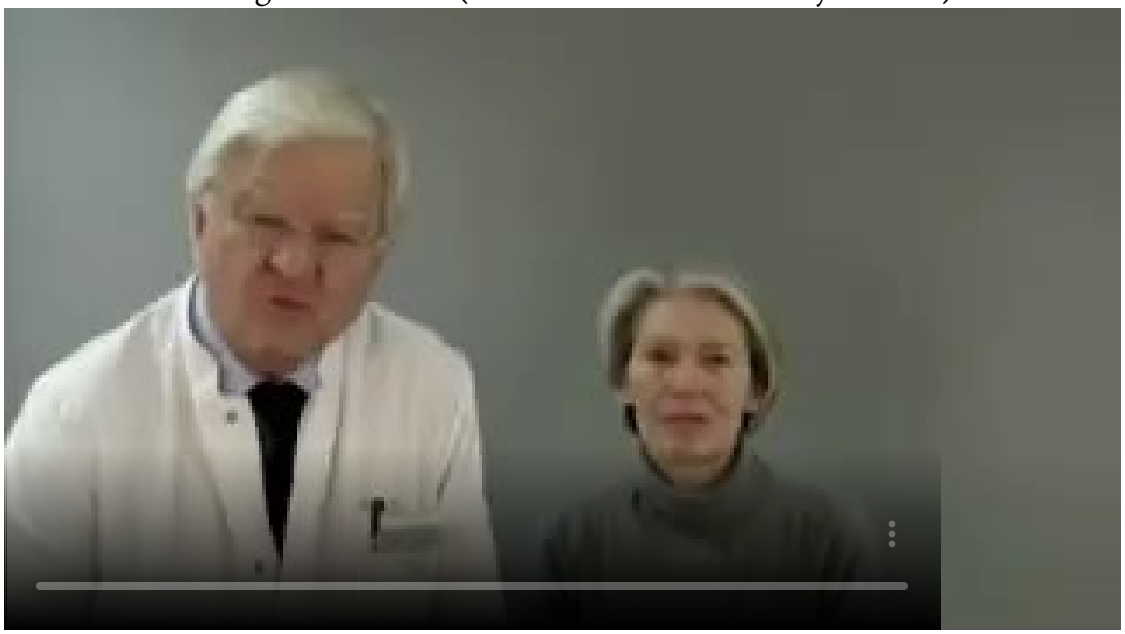




(a) and (b): in a normal test the patient (on left in the photographs) reaches either of the examiner's finger precisely

MOVIE 5.1.1 Finger-finger test – three modes (normal and abnormal dysmetria)

MOVIE 5.1.2 Finger-nose test (normal and abnormal dysmetria)



MOVIE 5.1.3 Dysmetria in cerebellar degeneration

The examiner observes the continuous movement for evidence of kinetic tremor and **precision** of reaching the target for evidence of dysmetria. With open eyes the patient is then asked to aim at the nose and back to the **finger** of the examiner (see Movie 5.1.2). Then, the examiner's finger may be moved to a **new position** while

the patient has to follow that new position. It is critical that the patient's arm is largely **extended** at the examiner's finger in order to properly assess for cerebellar dysfunction. If the patient's proximal strength is limited the patient may perform repeated target-to-target maneuvers at shorter distances as a surrogate for standard finger-nose-finger testing.

Even more sensitive and demanding is the **rapid finger-to-finger** ("mirror- movements") test: the patient is asked to reach out with one hand and then have the index finger in front of, **but not touching** the index finger of the examiner. The examiner moves his/her finger quickly to various positions and the patient is asked to follow precisely these movements without touching the examiner's finger at each of the positions. Patients with cerebellar dysfunction will overshoot (i.e. displaying **dysmetria**) when attempting to follow the rapid movements of the examiner's finger. In the second part of the exam the examiner has **two index fingers** in place (as in Figure 5.1.2 and Movie 5.1.1) and the patient points with his/her one index finger to each of the examiner's index fingers in **alternation**, while the examiner varies the distance between his/her own index fingers.

Note: major central motor weakness from hemispheric disease will also interfere with these tests and may preclude proper identification of cerebellar dysfunction.

SUBSECTION 2

Heel-knee-shin and toe-to-finger tests

In the **supine position** the patient is asked to lift either leg and **touch** the contralateral knee with the heel of the other foot and then **glide down** the shin and **back up**. Noticing how accurately the heel makes it to the knee and the straightness of the path along the shin are key aspects of the test.

FIGURE 5.2.1 Heel-knee-shin test

(a) the heel touches the knee precisely;

(b) the heel glides down and back up.

More sensitive is the **big toe-to-finger** test: the patient lifts either leg and aims at touching the examiner's finger in different positions (**similar** to the finger-nose-finger test). Both tests may be difficult to perform in elderly patients or in those in whom proximal weakness of a lower limb or arthralgia of the hip is present.

FIGURE 5.2.2 Toe-finger test (normal and abnormal)

in (a) the left big toe reached the finger target

(b) on quickly moving towards the other finger there is an overshoot

SUBSECTION 3

Alternating movements (diadochokinesia)

The patient is asked to turn the hands in and out (**supination – pronation**) as fast as possible and to open-close the fist. Diadochokinesia may be easily tested in the sitting patients by asking him/her to touch the thigh alternatingly with the dorsum or the palm of his/her hand. If movements are slow this is called **brady-** (= slow) diadochokinesia; if it is not properly coordinated (noted not only by overt incoordination and awkward hand positioning, but also with listening for poor rhythmicity and strength modulation) it is called **dysdiadochokinesia** (Movie 5.3.1). This problem can also be assessed through sequential finger tapping: index, middle, ring, pinkie, ring, middle, index as long as there is no **hemispheric** pathology (see also Movie 3.2.6 and Movie 3.2.7).

MOVIE 5.3.1 Alternating movements (normal)

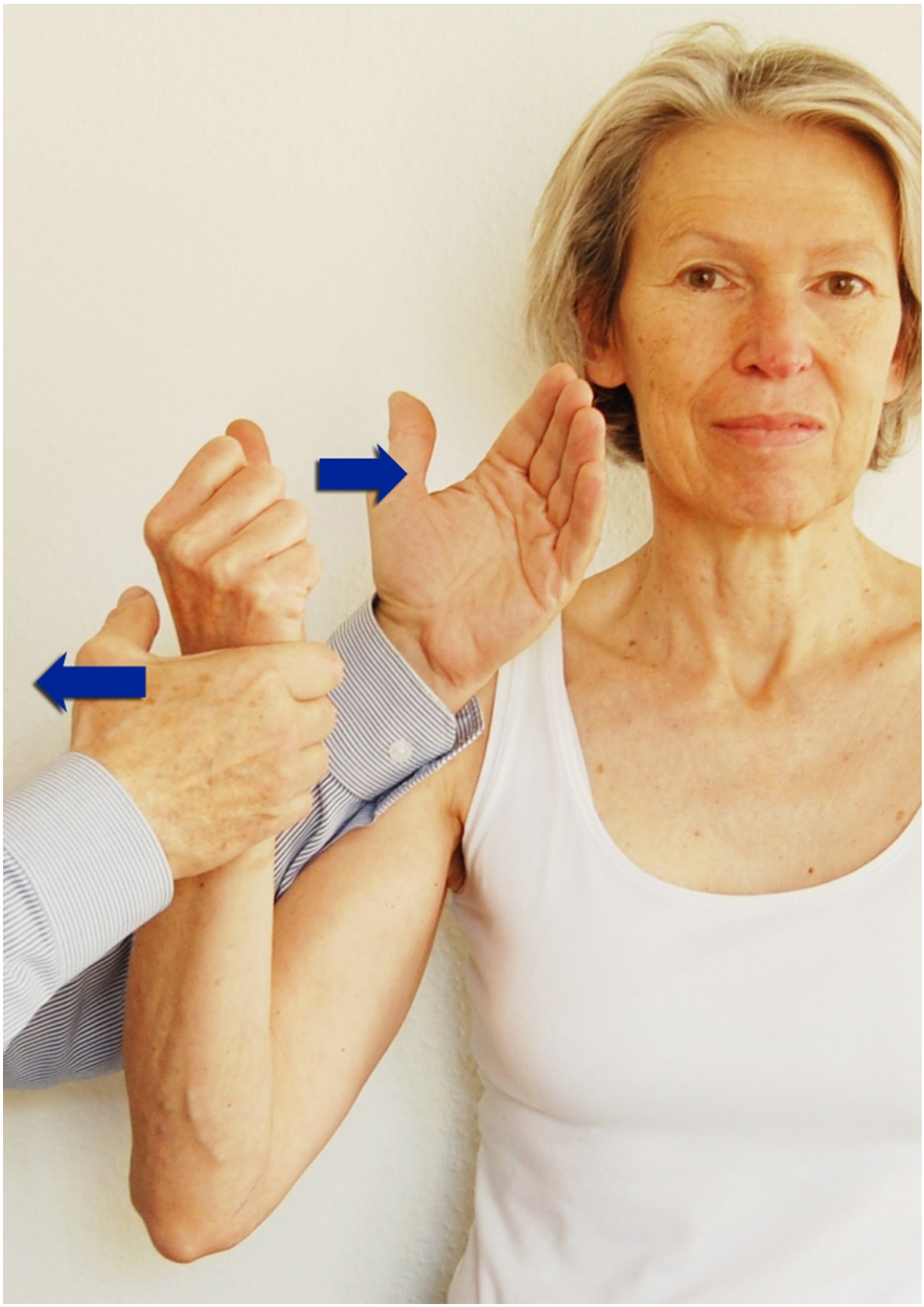
MOVIE 5.3.2 Alternating movements (abnormal, MS)

SUBSECTION 4

Arm rebound test

Cerebellar dysfunction leads to a loss of the fine tuning of a forceful movement. The patient is **unable** to break down an initiated bouncing movement. The patient is asked to pull the bent forearm towards the face against the hands of the examiner. The hand is then abruptly removed and releases the bent forearm which briskly moves towards the face. A healthy person would not hit his/her own face. To prevent a hit in an afflicted patient the examiner protects the patient's face with his other hand (Figure 5.4.1 and Movie 5.4.1). The test can also be performed with the patient pushing his/her **outstretched arms** upwards, against the examiners hands pushing downwards. When the examiner releases his/her hands the patient's arms will swing upwards.

FIGURE 5.4.1 The rebound test



MOVIE 5.4.1 Rebound test – two modes (normal and abnormal)

SUBSECTION 5

Control of posture ("pull test")

The patient stands upright and the examiner is **behind** the patient taking care to prevent the patient from falling. The patient is informed about the procedure. The examiner asks the patient to maintain his/her balance when pulled backwards. The examiner pulls the shoulder and trunk of the patient abruptly back assessing the patient's ability to recover. The patient is allowed to step back to regain postural stability. The test result is **abnormal** if the patient needs **more than 1 step** back to recover, or if the patient would have **fallen** had the examiner not provided immediate support (Figure 5.5.1 and Movie 5.5.1). One must avoid performing this test if the patient is too heavy to be held safely in any abnormal situation. Postural instability is a frequent finding in Parkinson's disease and related movement disorders.

FIGURE 5.5.1 The pull test

The patient is swiftly pulled backward and it is checked whether he or she make a counter-movement to regain balance and preventing a fall. The examiner is prepared to hold the patient. If pushed forward a second person should stand close by to prevent falling.

MOVIE 5.5.1 Pull test (normal and abnormal)

Note: while also part of this exam, please refer to Section 3 ("Motor exam") for descriptions of the Romberg manoeuvre, gait tests, and monopedal hopping/jumping.

SECTION 6 – SENSORY SYSTEM

General Terminology

Epicritical sensation

Position (proprioceptive)

Two-point discrimination

Pallesthesia

Allesthesia and Paraesthesia

Tuning fork

Hyp(o)esthesia

Dysesthesia

Allodynia

Protopathic (pain/temp)

Hyperalgesia

Hyp(o)algesia

Stereognosis

Graphesthesia

Kinesthesia

Extinction

When a patient does **not** complain about sensory symptoms and signs, it is recommended to **only** perform a few simple **screening**

tests before going into any detail:

Check sensation to light touch on upper and lower limbs and on the trunk with the examiner's hand by comparing proximal to distal and left to right, often ensuring that similar dermatomes are compared side-to-side. **Sensation in the face** is a good **reference** for assessing abnormalities below the neck.

Check pain sensation to pinprick (simple toothpick or single-use safety pin) and vibration sense on upper and lower limbs. ***(Note: people don't wince if they cannot feel pain.)***

When it turns out that sensation is abnormal a **detailed test** is needed. We tend to discount a minor deficiency or right-to-left difference, unless there is a striking pattern suggesting a need for further investigation. In areas of other overt abnormalities such as regional sweating abnormalities, redness of skin, purple discoloration of hands and feet, skin ulcerations, sensory testing has to differentiate between localized abnormalities at or near the skin alteration and those that may be part of a more widespread neuropathic disorder.

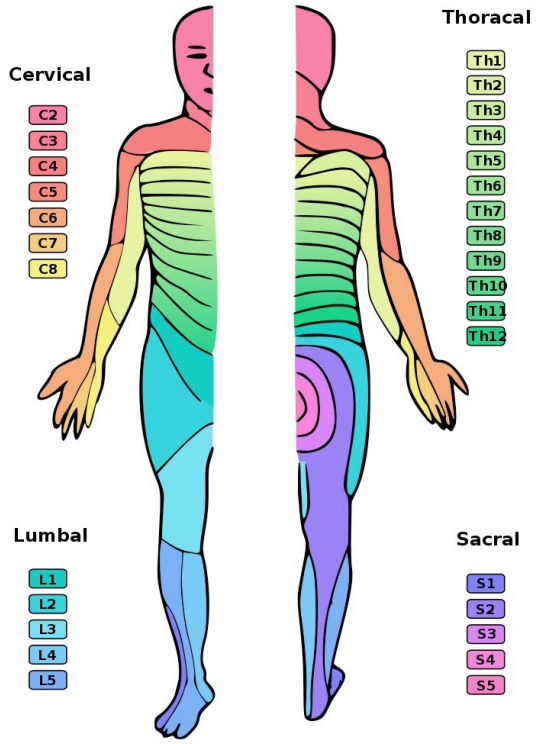
Sensory Examination

Note: any sensory testing is entirely subjective and fully depends on the cooperation of the patient. It becomes more convincing if findings are reproducible from hour to hour and day to day. There is never a sharp (knife-like) border of a sensory deficit because of the interdigitation of nerve endings at the border zones of their dermatomes. Similarly, with truncal sensory loss in hemispheric or spinal cord disease, the border is paramedian up to 3 cm away from the midline towards the abnormal side (see also Figure 2.4.2, section 2.4).

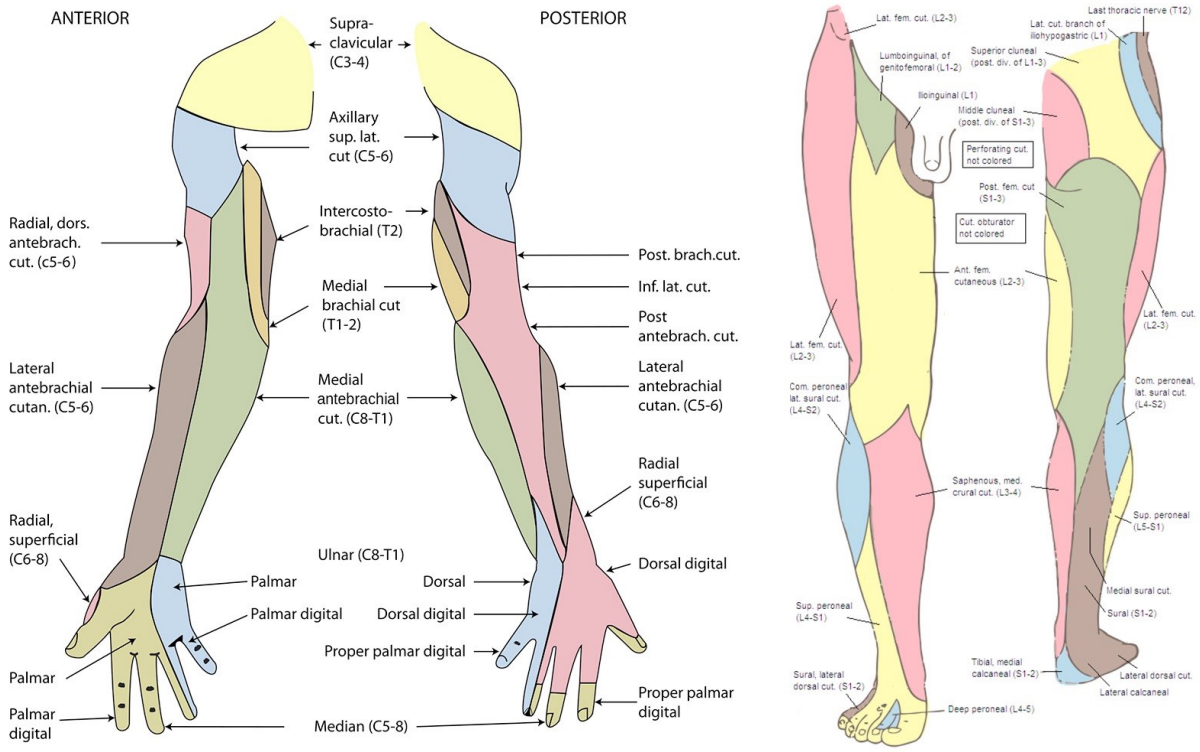
It helps **first asking** patients to delineate the distribution of their sensory loss or of any alteration. It is best to ask the patient to close the eyes while being tested. Test stimuli should cover **major** dermatomes and peripheral nerve innervation zones. These can be recapitulated by using an anatomical cartoon (Figure 6.1.1), where findings can also be documented.

In a given lesion of the PNS, complete loss of pain sensation has usually **narrower borders** than has sensation to touch (Figure 6.1.3). Yet there may be a radiating pain sensation with chronic pain syndromes. Pain syndromes from a lesion of the CNS are rare in clinical practice (dorsal horn of the spinal cord or as a delayed effect in thalamic lesions).

FIGURE 6.1.1 Cartoon illustrating innervation zones. Commonly used schemes indicating the sensory fields.



in the cartoons of Fig. 6.1.1a - the dermatomes - reflecting the radicular distribution;



in the cartoons of Fig. 6.1.1b - individual peripheral nerve innervation zones in the upper and lower extremities reflecting sensation transmitted by individual peripheral nerves..

Note: variations in the dominant segmental fields are common, i.e. in the cervical and lumbosacral regions by one or even two segment up or down as compared to the cartoon in Figure 6.1.1**.** The same applies to the territories of peripheral nerve innervation zones.

For the **epicritical sensory modalities** the encapsulated, differentiated **end organs** are probably mainly responsible for the mediation of such qualities as fine touch, discrimination, vibration, pressure, two-point discrimination and so forth, which travel in the **dorsal columns** and medial lemniscus to the posterior nuclei of the thalamus. The free nerve endings mediate protopathic **modalities** such as pain and temperature, which travel in the **spinothalamic tract**. The evidence for this functional distinction is incomplete, however. This suggests screening exams ought to contain elements from each of these 2 major domains:

Quick screen in each of the sensory domains: check whether the patient feels the **lightly touching finger** of the examin-

er on limbs and trunk (and face – see Section 2 "Cranial nerves") and compare the corresponding areas on both sides; occasionally it is important to also check the trunk, going from dorsal to ventral following the course of the intercostal nerves (may be affected, e.g., with severe diabetes mellitus, ganglionitis leading to pseudotabes).

Note: in clinical practice sensation at the forehead or cheek may serve as a reference area when trunk and periphery are tested.

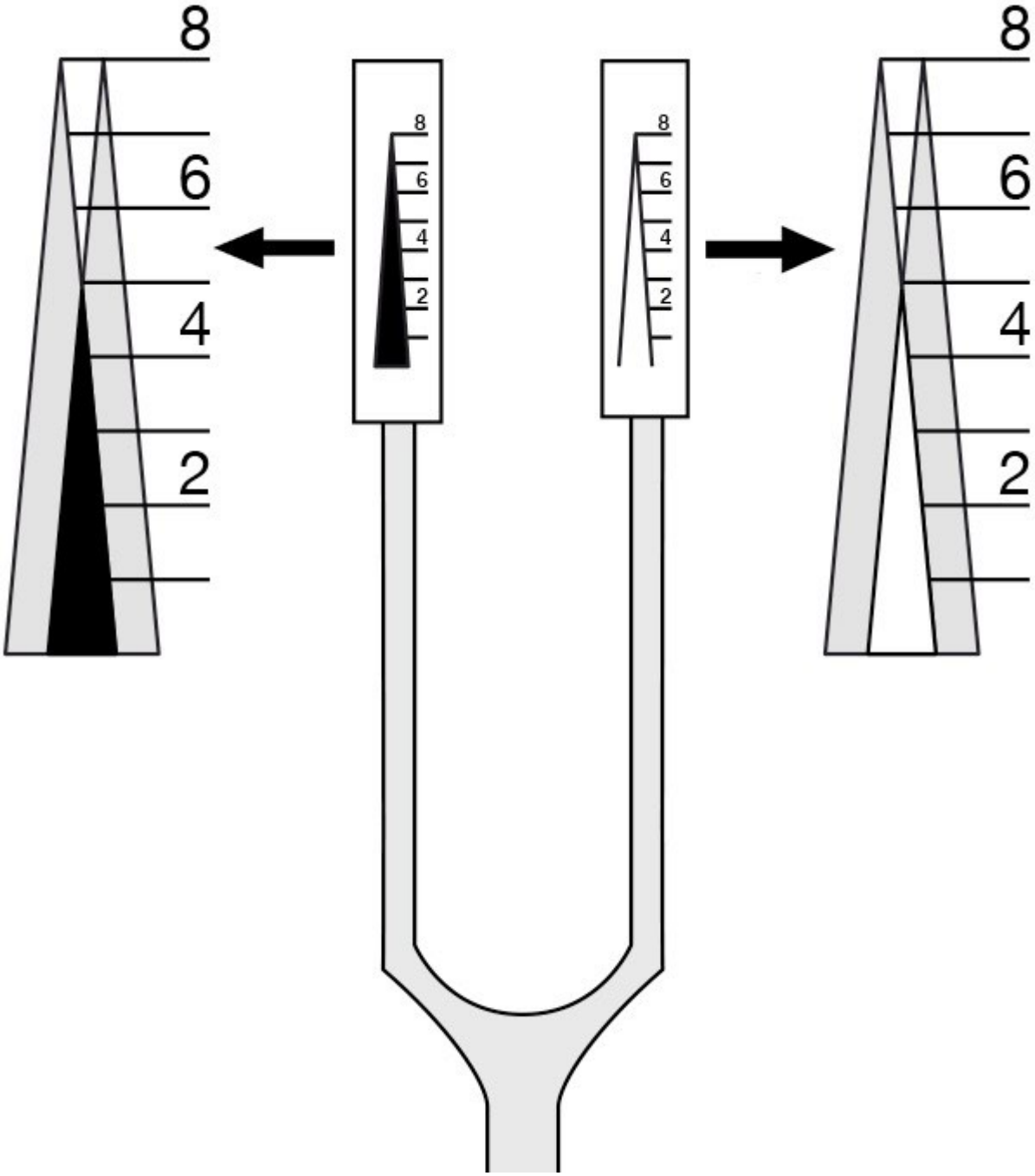
Check **pinprick** (sharp pain) by a **toothpick** (Movie 2.4.1) or a **single-use** sterile safety pin and **temperature sensation** by using a cool piece of metal (e.g. the **reflex hammer**) bilaterally and from distal to proximal at the limbs in the same way as above; this test is not feasible when the patient's hands and feet are cold because nerves only sense temperature relatively, not absolutely.

Vibration Sense: To expand on the basic screen one can test the vibration sense (**pallesthesia**) with a graded **tuning fork** (Type **Rydel-Seiffer**, 64 Hz with weights), which is preferred over a standard **tuning fork** with weights (128-256 Hz, Figure 6.1.2). Normal values have been published for the graded fork in different age groups (Martina et al, 1998). With the standard fork the reference is the forehead or the sternum in the patient and the hand of the examiner (provided he/she has no deficit him/herself). In order to properly use a standard **tuning fork**, one must ensure a standard mechanism of stimulation. In order to do this, bang the **tuning fork** hard enough that the heads "ding" together, this ensures that all stimuli are the same and the decay will occur in a predictable fashion: now you have a precision instrument. The accepted standards are that, from maximal stimulation, the interphalangeal joint of the thumb should perceive the vibration for > 15 seconds and the interphalangeal joint of the great toe should perceive the vibration for > 10 seconds.

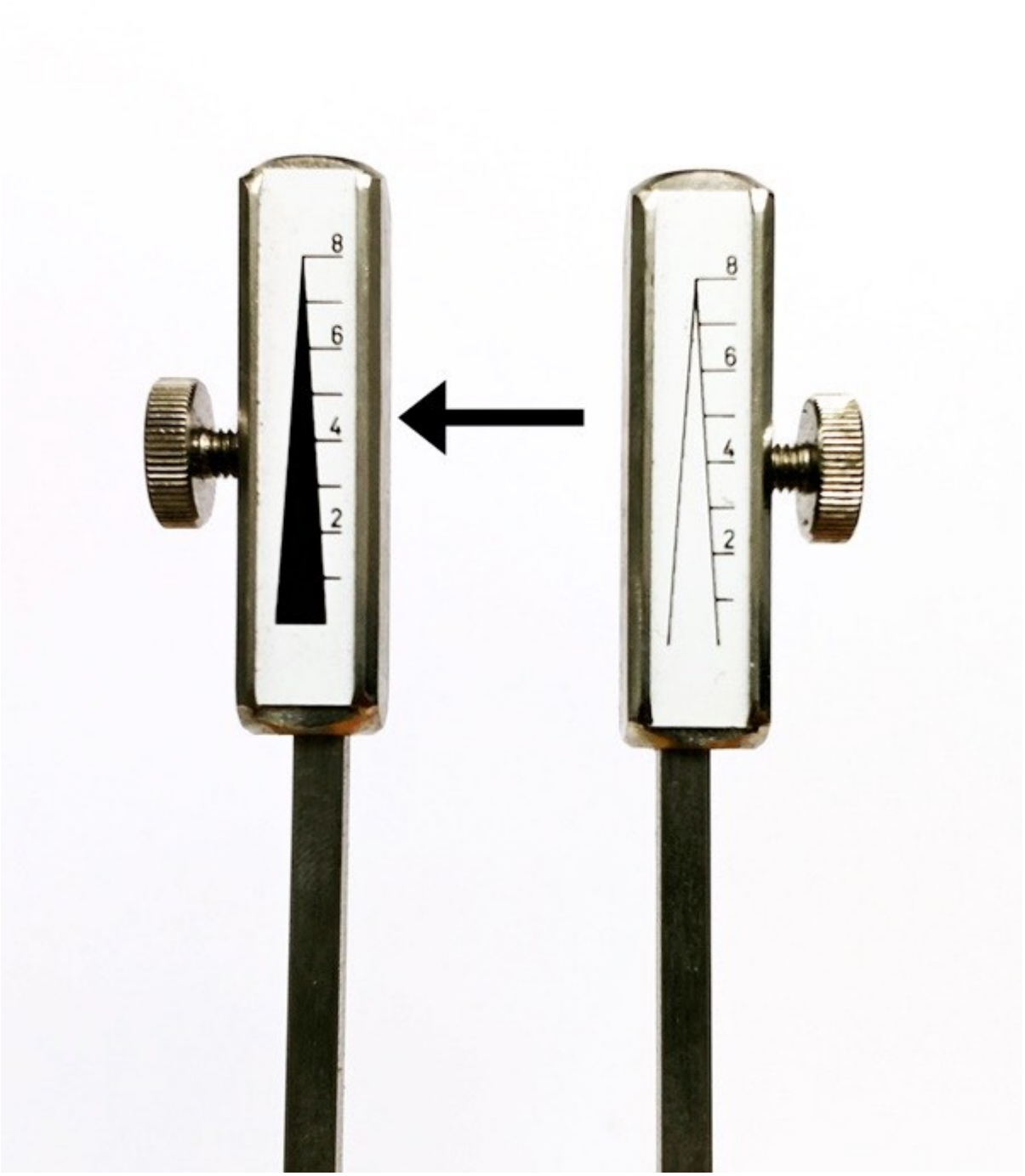
When using a graded Rydel fork the scale on the weight is observed. With decreasing vibration the blurred triangle (along with a scale showing numbers 0 to 8) will become sharp and the number

next to the sharp peak (Figure 6.1.2 and Movie 6.1.1) corresponds to the vibration sense threshold of the patient. If there are clear deficits this measure is reproducible. According to one study (Martina et al., 1998) a sensation of about 4/8 or better at the ankle and about 6/8 at the big toe is the normal lower limit for 40-80 year-old persons, but others set higher lower limits.

FIGURE 6.1.2 Tuning fork - Type Rydel-Seiffer, 64 Hz with weights



(a) The scale value at which the central black and white peaks are becoming sharp



(b) equals the mechanical intensity of the vibration at that moment in time (arrow points to about 4/8). The peak comes out better in reality than on the photograph and in the movie.

MOVIE 6.1.1 Rydel-Seiffer Tuning fork (in action)

Testing vibration sense: First, the distal end of the fingers and toes are tested (interphalangeal joints). If vibration sense is impaired, the distal radial bone and the epicondylus of the humerus bone are tested next, followed by ankle and epicondylus of the femur bone to check further proximal sites and thereby establish a distal to proximal **gradient** of pallhypesthesia, e.g. in a length-dependent type of polyneuropathy (see below). With the Rydell-fork the scale values at each bony point are documented.

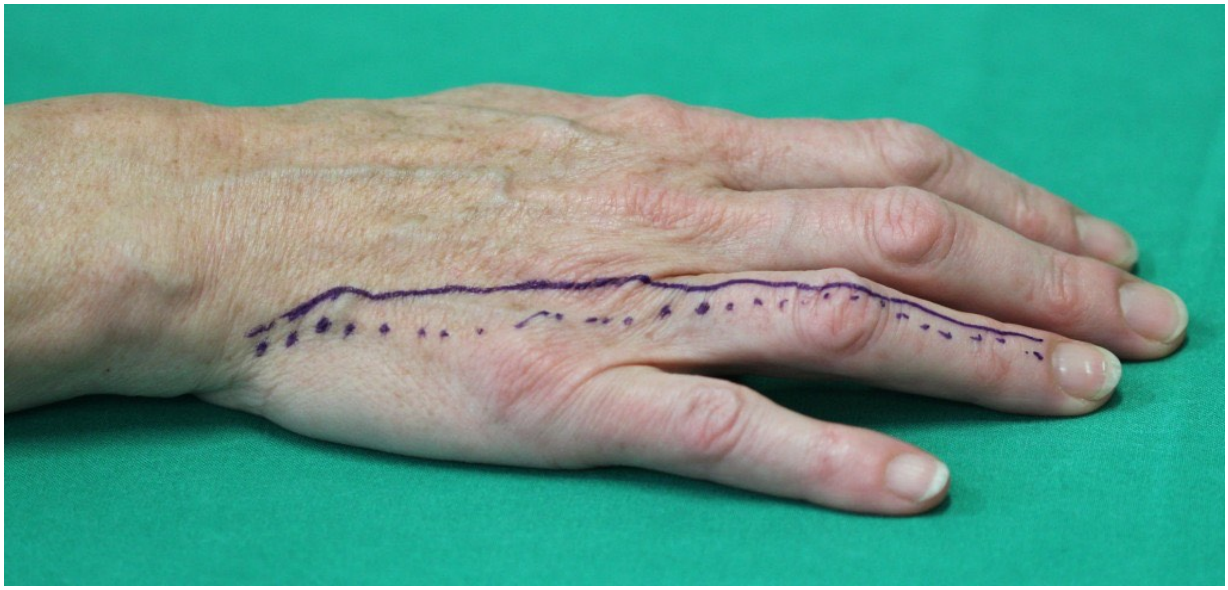
Note: the vibration sense is never affected by any lesion of one or even two spinal roots (e.g. with vertebral disc compression).

More thorough testing in each of the sensory domains

If any abnormality is found a more detailed testing is necessary, including evaluation of a **gradient** of sensory loss (by marching sensory stimuli from distal to proximal, looking for **length-dependent peripheral neuropathies**) and sensory levels (finding a **dermatomal level** at which sensation is lost or reduced, as with spinal root disease). Borders of the area of sensory impairment should be **checked at least twice**, from the **hypo**esthetic to the **normal** zone

and vice versa for touch and pain, and zones **marked** with a felt-tip pen. The examiner draws the findings in a cartoon (Figure 6.1.1) for sensory testing or may take a photo if needed. With a segmental or peripheral nerve field deficit the finding is also documented in a cartoon.

FIGURE 6.1.3 Borders of deficit in ulnar nerve lesion. Dashed line: pain; continuous line: touch sensation.



MOVIE 6.1.2 Pin prick testing for pain sensation

Note: conventionally, a dashed line denotes pain sensation and a continuous line touch sensation. The rule is that in a given lesion of the PNS, complete loss of pain sensation has usually narrower borders than has sensation to touch. This can also help to identify functional (conversion) disorder or malingering (see Section 9).

Then the patient is tested for the different qualities of sensation in more detail.

Touch sensation *(epicritical)*: the reference is the **finger** of the examiner and the **forehead** or cheek of the patient (Movie 6.1.3). If a deficit is present the sensation **threshold** should be examined by using a **paper tissue** (e.g. Kleenex tissue or the tip of a cotton-tipped applicator pulled to make a fine wisp of cotton). If this can be felt sensation is normal or close to normal. Then the pressure of the touching finger is increased until the patient starts feeling the stimulus. This should be repeated 2-3 times to check for reproducibility and to get convincing results. The rating scale is from "mild" to "severe" abnormality. While documenting the findings in a cartoon the anatomical distribution is defined.

Pain sensation *(protopathic)* is tested by pin pricking as follows: a clean, single-use toothpick is held in the pinch position with a gentle but constant pressure such that the pick may glide and slip through in case the pressure is getting too high. This procedure allows better comparison between left and right and between face and periphery (as shown in Movie 2.4.1). Then multiple stimuli are applied in any one region and the threshold estimated by altering the prick pressure. As with touch a **threshold** is being defined in qualitative terms. The rating is mild to severe abnormalities. While documenting the findings in a cartoon the anatomical distribution is defined. If a sterile safety pin is preferred equivalent measures should be taken to avoid injury and the ***pin must also be discarded*** after any one use.

Position sense *(proprioceptive)*: sensing position is a rather robust function provided by joint receptors **and** muscle spindles. One should **grab the sides** of the digits being examined (Figure 6.1.4 and Movie 6.1.4), because holding the digit from the top and bottom will provide **clues** by pressure-sensitive mechanoreceptors as to the direction of movement. The test is first explained to the patient in simple terms and by **demonstrating** what far up and far down movements feel like (a chance for the patient to correct any mistakes while they can see). Then, in a subsequent go, the test is performed with **eyes closed**: first testing larger and then smaller movements. A rough estimate of the angle detected is recorded and documented. In case of any major abnormality more proximal joints (hand and elbow, and ankle joint and knee) are tested. Normally, even small angles (15° degrees) are detected both at the big toe and in the **index finger**.

MOVIE 6.1.3 Touch sensation

FIGURE 6.1.4 Position sense testing at the index finger;

(a) middle position

(b) upper position. The finger is always grabbed at the sides.

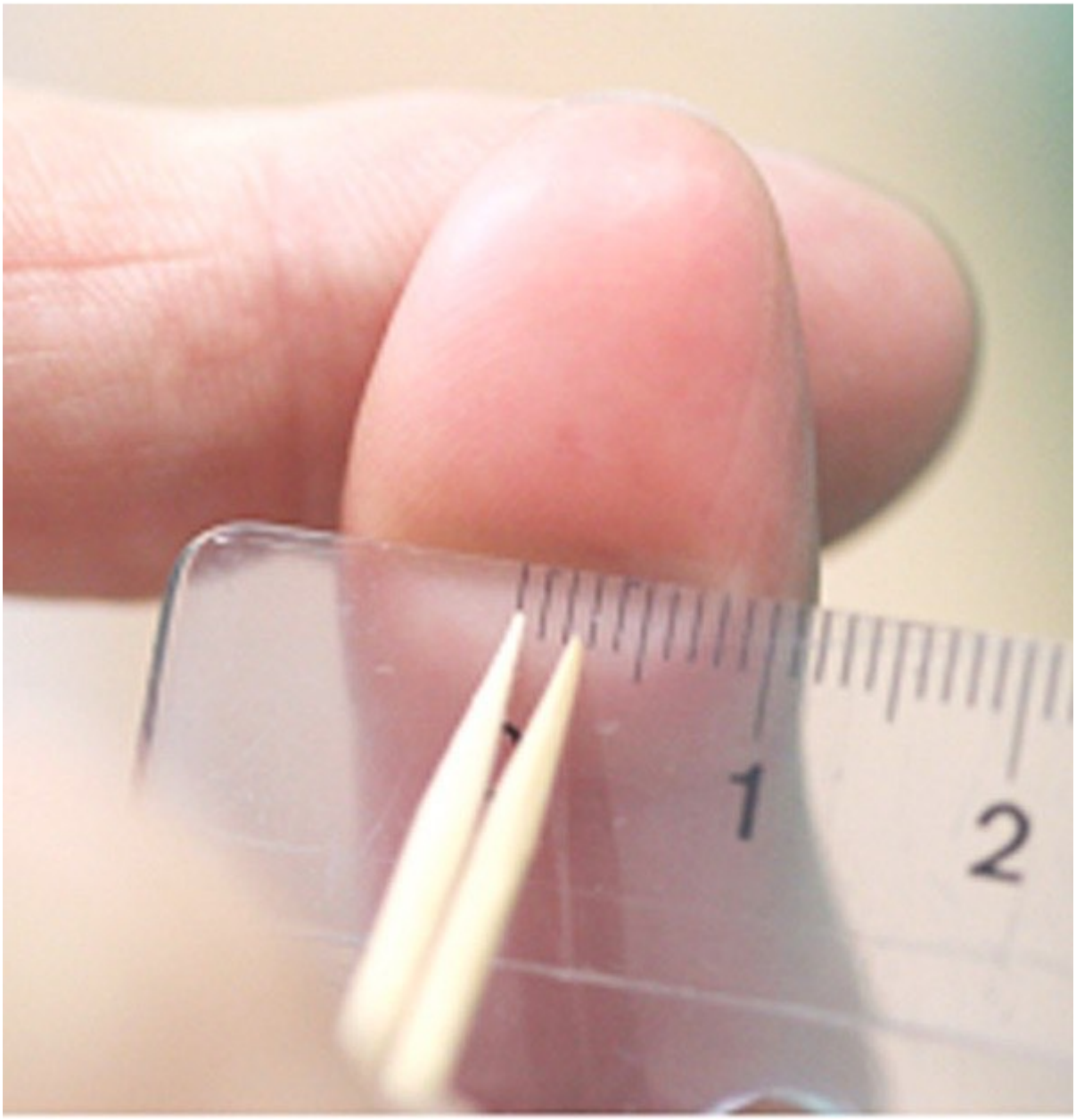
MOVIE 6.1.4 Position sense testing (normal and abnormal)

Note: proprioception is also tested to some extent while performing more complex tests like the Romberg stance test and the finger-nose test done with having the patient touch his/her nose with the eyes shut.

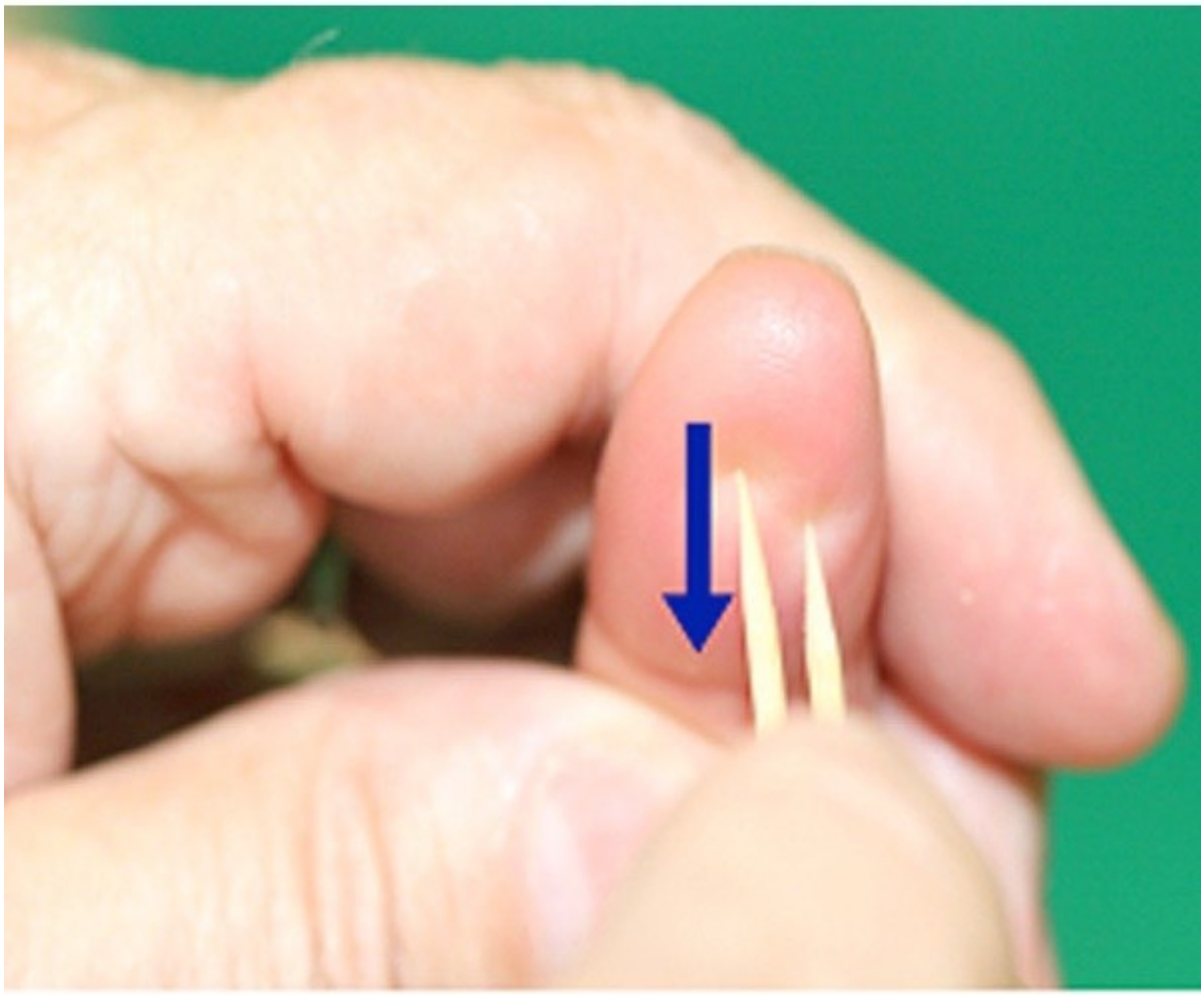
Two-point discrimination: two-point testing is best done with two **toothpicks** held in pinch position; pressure is adjusted to the level at which the patient can easily feel and discriminate a large distance. The distance of the two pricks at which it is just perceived is the **discrimination distance** in millimeters taken from a ruler. Technically, the two pricks are given simultaneously, first **"static"** (at one site at a time, and for **control** purposes also as **singlet** prick at any level), then **"dynamic"** by moving the two pricks along the skin over some 10 mm. As a rule, **two-point discrimination** is **better** and the discrimination distance is **smaller** with "dynamic" stimuli (Figure 6.1.5 b,c and Movie 6.1.5, Movie 6.1.6, Movie 6.1.7). Normal values are **age dependent**, about 4 mm for static testing on the finger tip and 5-6 mm on the big toe in young persons and up to 8 mm in the elderly in the fingers and over 10 mm on the big toe. On the trunk it can be 5 cm or more! (Nolan, 1982;1983; Van Nes et al., 2008).
Alternatively, a commercially available 2-point discrimination wheel (Dellon) with blunted pins may be used covering the range from 2 mm to 8 mm and from 9 to 20 mm.

Note: this test also may help to identify patients with a functional (conversion) disorder of sensation or with malingering that lack a reproducible difference between dynamic and static test results.

FIGURE 6.1.5 Two-point discrimination testing



a) static



b) dynamic

TABLE 6.1.1 Age-dependent normal values for 2-Point discrimination

Age Group	Static	Dynamic
20 – 40	4	3 – 3.5
40 – 50	4.5	3.5 – 4
50 – 60	5	4
60 – 70	6	5
70 – 80	7	6

2-Point discrimination at finger tip: Distance of stimuli in mm as measured with the Dellon Device (modified from van Nes et al, 2008)

MOVIE 6.1.5 Two point discrimination (static, normal)

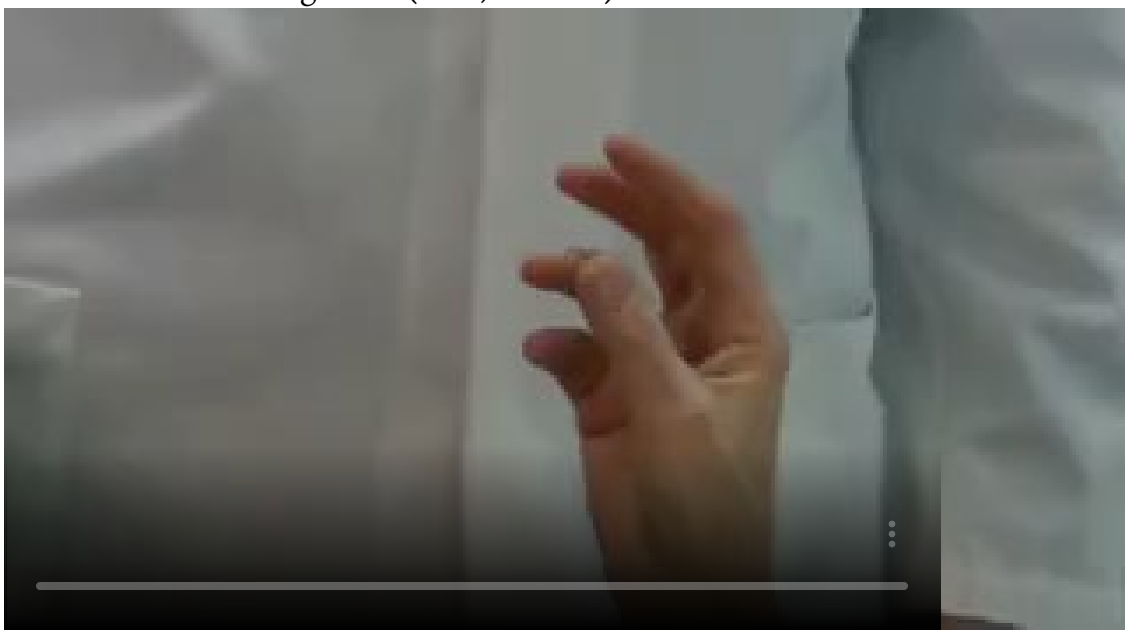
MOVIE 6.1.6 Two point discrimination (dynamic)

MOVIE 6.1.7 Two point discrimination (toe)

Stereognosis is a more sensitive but also **more complex** test, adding up two-point discrimination and higher cortical sensory functions. Test items are from **daily living experience** including paper clips, various coins, a key, a Kleenex tissue. Acoustic clues (e.g. with a bundle of keys) should be **avoided**. Normally, a person can

easily tell a car **key** apart from a house key and a Quarter from a 5 cent coin, and even may discern various **materials** like metal or wood. This test is also **dependant** on alertness and attention. An easy way is to have a number of simple items hidden in a bag to be discerned and named: key, coin, piece of fabric, cube of plastic and the like (Movie 6.1.8, Movie 6.1.9, Movie 6.1.10).
Note: this test also may help to identify patients with functional disorders of sensation (section 9).

MOVIE 6.1.8 Stereognosis (coin, normal)



MOVIE 6.1.9 Stereognosis (car key, normal)

MOVIE 6.1.10 Stereognosis (abnormal, sensory or motor problem)

Graphesthesia (tactile character perception) is another test for complex somatosensation; it can be assessed by "writing" numbers, letters, or symbols on the **distal skin** of limbs using a toothpick. The size should be about 2 cm on the back of the hand and 3-4 cm on the dorsal aspect of the foot. Wiping the examiner's hand

over the area being tested between successively presented characters ensures best test conditions by "resetting" the sensory area through transient overstimulation (like eating crackers between wine tastings). The test should be **repeated** with 3 different numbers if not correctly detected the first time.

Extinction: touch stimuli and pin prick are applied first on one side or at one point on either side of the body only and then simultaneously at the two **corresponding** sites bilaterally; the patient is asked on which side he/she is feeling the stimulus. With single stimuli **still appreciated**, bilateral simultaneous stimulation may lead to **extinction** of the perception on one side, contralateral to a hemispheric lesion (Movie 6.1.11). While this is most striking in the tactile domains, **extinction** and neglect can happen in multiple domains and should be verified in the visual realm as well (see section 2.2).

MOVIE 6.1.11 Right-left touch, serial and simultaneous, extinction

Note: sensation is also an important function of posterior thalamic and cortical integration mechanisms. In order to assess these functions, more complex testing such as described in paragraphs 4-8 may

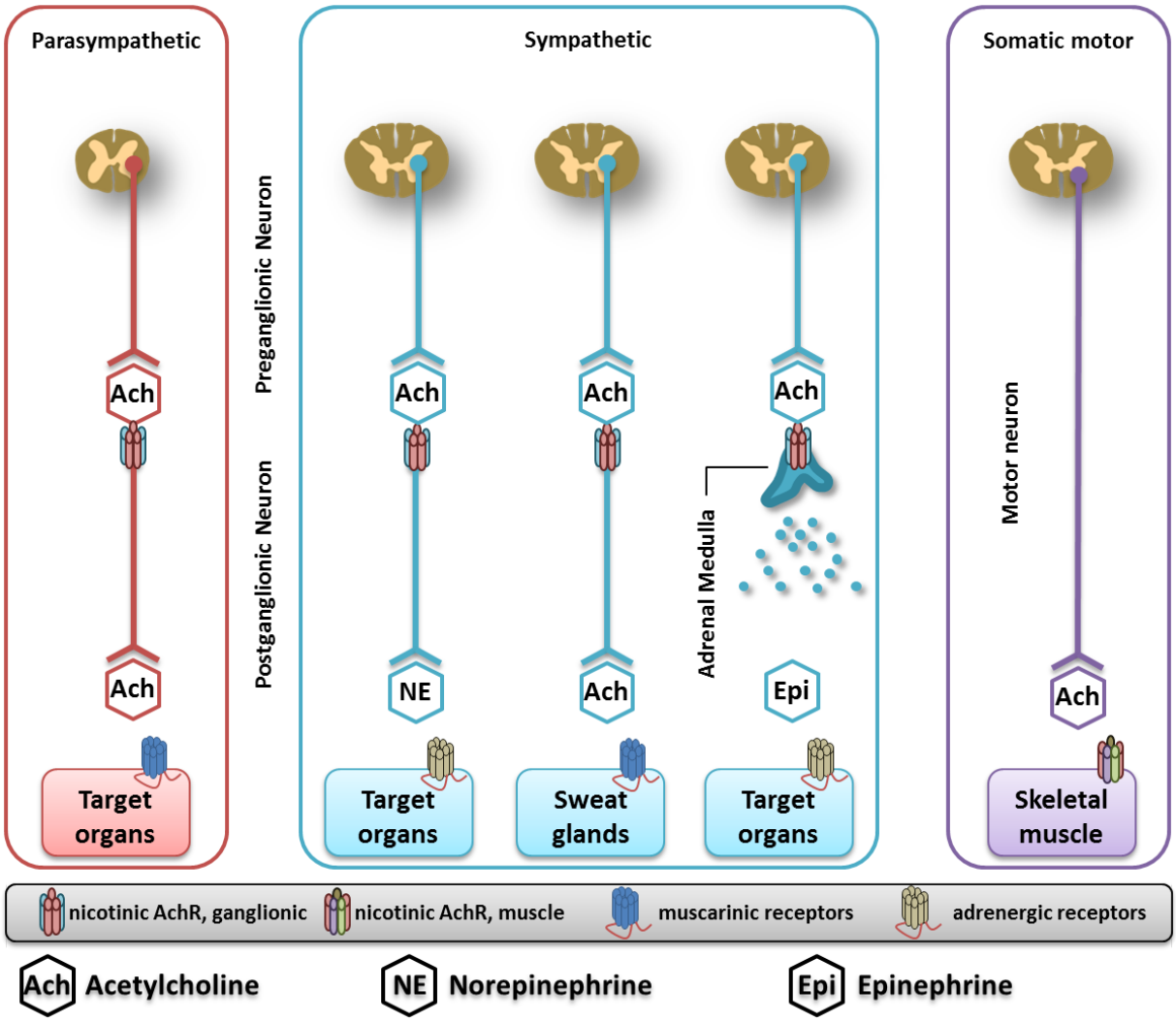
help localizing a lesion to the brain in the absence of (and occasionally with) peripheral sensory fiber abnormalities.

SECTION 7 – AUTONOMIC NERVOUS SYSTEM

The autonomic nervous system arises centrally from the insular cortices, which exert sympathetic and parasympathetic influence through output to the brainstem and the sympathetic and parasympathetic pathways including the thalamus and hypothalamus. Peripherally, the neurons originate in the sympathetic and parasympathetic ganglia of the spinal cord. A focus on **each of the main organ systems** with autonomic function is most helpful, and often relies upon direct questioning during system's overview while taking the history, with **only a few physical exam assessments** being possible. Thinking about the simple acronym **SLUDGE** (salivation, lacrimation, urination, defecation, GI distress, emesis) and perspiration will help to think of most of all these systems.

It is important to localize the problem to that of a division of the autonomic nervous system, certain nerves/regions, or neurotransmitters (Figure 7.7.1).

FIGURE 7.7.1 Autonomic systems



Bladder: Specific questions may be added during exam if they have been omitted during history taking: urgency, retention, incontinence, nocturnal polyuria with daytime oliguria (a position-dependent perfusion phenomenon), and frequent UTIs. During examination, abnormal smell of urea is indicative of urinary incontinence; it may be helpful to inspect the underwear. If retention is likely, the belly should be carefully palpated for a full bladder. Mild percussion of the lower abdomen while listening to the sound with auscultation using a stethoscope may give hints to a full or only partly emptied bladder. This can be confirmed at the bedside by a hand-held ultrasound device.

Sexual function: sometimes best assessed through colloquial questioning while taking the general history: changes in penile erection (spontaneous, present upon awakening, sexual intercourse), orgasm, ejaculation, desire, and lubrication.

Note: a proactive approach is needed because patients often do not spontaneously report symptoms of the urogenital system.

Alimentary and stools: questions during history should be directed at recent constipation or diarrhea, incontinence, **dysphagia** at the end of swallowing (distal 2/3 of esophagus is smooth muscle!), early satiety, and xerostomia. A rectal examination gives hints as to the resting sphincter tone, which is partly based on the internal (smooth) muscle sphincter (cf. section 4, anal sphincter reflex).

Eye and lacrimal glands (see also section 2): vision problems (may also arise from pupillary abnormalities) – specifically impaired **night** vision (sympathetic failure) or daylight intolerance (parasympathetic failure), dry eyes or the reverse, i.e. inappropriate tearing ("crocodile tears").

Partial (mild) **ptosis** indicates weakness of the non-striated component of the levator palpebrae superior (Müller's muscle). It is sympathetically innervated. Schirmer's test strips can assess for the degree of lacrimation. The most common pattern associated with an abnormal Schirmer's test is the **"sicca"** (dryness) syndrome.

Respiratory: involuntary sighing, stridor, snoring, and apneas are the historical elements, which may point to respiratory autonomic dysfunction, but may more frequently point to a motor disorder proper. On physical exam common disorders of respiratory rhythm are appreciated (drug intoxication, major alcohol intake before bedtime, stupor and light coma, brain stem stroke)

Cardiovascular: Patients with dysfunction of positional blood pressure and heart rate variation will have a variety of complaints

including orthostatic hypotension, palpitations or bradycardia, a "coat hanger" neck and shoulder pain with upright position.

Note: except for the inspiration-expiration test none of the following should be done at the bedside if there is already evidence of overt autonomic failure or when the diseases in question are prone to cause life-threatening autonomic dysfunction (e.g. in the Guillain- Barré syndrome).

In any of these situations, formal testing in the lab is advised with proper precautions for emergency intervention, and only if the risk-benefit ratio for any of these tests is acceptable.

At the bedside, a few simple tests are available:

- Check for **in**spiration-induced **in**crease and an **ex**piration-induced **de**crease in heartbeat frequency, which indicates normal autonomic control. Its absence suggests cardiac autonomic dysfunction. An ominous sign is fixed tachycardia, being an indicator of lost vagal and sympathetic control of the sinoatrial node (inherent pace of the node is 110 beats per minute). If there is no change or if paradoxical slowing is seen this is also an **ominous sign for a life threatening autonomic neuropathy**. In addition, major blood pressure changes without any adequate demand are suggestive of cardiovascular dysregulation.
- Another bedside test is simple **orthostatic vital sign** testing (particularly in the morning, following a meal, or following exertion) with blood pressure taken while lying supine followed by a blood pressure taken after standing for 5 minutes, **with no sitting transition in between**.
- Sympathetic heart rate increases can be induced with sustained hand grip (3 minutes), immersion of the hand in ice water for 90 seconds, or simple mental arithmetic (serial 7s).
- Similarly, parasympathetic heart rate decreases should be inducible with carotid massage with patient tilted or upright (<u>contraindicated</u> in patients with a hypersensitive carotid si-

nus, or with the possibility of carotid plaques), as well as with deep breathing/hyperventilation.

- Capillary refilling upon applying pressure onto the distal parts of the fingers may be delayed in disorders with autonomic failure such as multisystem atrophy.
- Another parasympathetic (vagal) stimulus decreasing pulse rate and blood pressure is the **Valsalva test** (Figure 7.7.2): have the patient close the mouth and the nose with his/her pinch and then try to breathe out against resistance for 2-3 seconds while checking the pulse rate.

FIGURE 7.7.2 Valsalva manoeuvre

Sudomotor and trophic alterations: One should specifically ask for increased or decreased **sweating**, temperature **dysregulation**, skin **changes** (dryness, change in texture, color, etc.). The examiner

can assess sweating by touching the skin at the face, trunk, thighs, and palms with the finger tips and the **back** of his/her hand. A very dry or a sweaty and moist skin can easily be discerned on exam, but a simple test with caking of sprinkled talcum powder may help reveal **hyperhydrotic** skin and can easily be documented by a photograph. The anatomical sites are compared right to left and proximal to distal.

Any longstanding autonomic dysfunction associated with a degenerative disorder may show **pathognomonic** signs: finger- or toenails may show lines or dystrophic changes; the skin may become thinner and shiny, loosing its hairy cover, wounds may not heal well. Particularly in the lower limbs ulcerations may indicate trophic changes, and the bones and joints may develop degenerative changes (arthrosis, osteoporosis). This can all be confirmed by bone densitometry, x-ray, CT, and MRI. A well known prototypic entity in this domain is the "**diabetic foot**" with large and small-fiber neuropathy and autonomic neuropathy making the patient prone to minor injury or ulcers with superinfection sometimes leading to **sepsis and death** if remaining unnoticed.

SECTION 8 – NEUROPSYCHOLOGICAL FUNCTIONS

A first impression of a patient's neuropsychological function is usually generated during history taking and when doing the standard neurological exam. Much of what one needs to know is brought up by simple, **colloquial-style** questions, e.g. "How did you get here today?" "Who took care of you this morning?" "What did you see on TV last night?" "Have your kids visited you today?"

If next of kin are in the examining room they tend to **help** the diseased relative if he/she does not properly **respond or perform** when asked. This may be an **indicator** of neuropsychological abnormalities in the patient, mainly in memory and orientation.

Note: proper appreciation of neuropsychological malfunction may have important implications for the lack of or still-existing capacity to provide informed consent. This is also important due to the common question of capacity. Capacity assessments can be performed by anyone; the only key aspect is that of proper documentation. Capacity can vary from decision to decision, and must be clarified as to the patient's current state of mind, his/her understanding of what is to be done, what the risks and benefits of that action are, what the alternatives are, and, most importantly, that he/she expressly understands all of these factors and has demonstrated the ability to provide a logical rationale for his/her decision.

Introduction to the examination of neuropsychological and higher cortical functions

All screening tests are complex in nature, testing more than one cognitive function at a time. Whenever abnormalities or deficits are found a more focused exam is needed, including quantitative tests that are validated for different age groups and populations. In disordered language, specific tests for aphasia are available, in apraxia various other tests can be done to delineate the deficits and classify the disorders.

The **Movie 8.8.1** illustrates how to approach a patient with cognitive decline. The movie shows part of a longer interview with a patient. The impairment could have been overlooked easily if the patient had not been questioned and examined carefully. In the movie, the former engineer is attentive, shows a normal body language and replies swiftly to questions posed by the doctor. An unexperienced observer might find nothing wrong. Only here and there, he uses wrong words or hesitates a bit before answering the posed question. When formally asked to tell a ladder apart from stairs he fails to explain the difference between the two tools - not as one would expect from a healthy person at age 68 years. Upon history taking from the patient's wife she explained that her husband cannot remember where he places things, gets lost when shopping alone, and that he frequently complains about funny sensations in his head. It is obvious that the patient is unaware of these memory problems nor is he aware of the mild anomia.

Note: a colloquial and empathic style of the interview is essential if the doctor wishes being accepted and appreciated. Specific testing questions should not be stressed too early.

MOVIE 8.8.1 How to interview a patient with early dementia

Evolutionary (ontogenic) reflexes

In patients that appear with obvious dementia or any other advanced degenerative disorder simple bedside tests can be done collectively named evolutionary or ontogenic reflexes. These reflexes (or better reactions) are characteristic for advanced degenerative disorders of the brain. None of them are disease-specific. During postnatal maturation of the CNS these reflexes are normal phenomena and gradually disappear after the first half year of life. Perhaps the most famous of these reflexes is the Babinski reflex discussed Section 4.

Palmar grasp reflex: upon touching the palms, the patient will grasp the hands of the examiner. The movie is showing the palmar grasp reflex (M 8.8.2). The patient in the video is demonstrating disinhibition of an ontogenic reflex that is usually absent after 5-6 months of age. Despite the examiner providing no grip force the reflexive grip of the patient is stimulated by the gentle pressure and traction of the examiner's fingers on the patient's palm.

MOVIE 8.8.2 Palmar grasp reflex

Rooting reflex: well known in babies, the lips extend slightly along with a head turn towards the side of stimulation. This reflex is being elicited by gently touching a spatula to the cheeks, next to the angle of the lips (Movie 8.8.3). The first part shows the normal situation with no response, the second part mimics an abnormal rooting reflex.

MOVIE 8.8.3 Rooting reflex

Palmomental reflex: upon gently scratching the palms, the mentalis muscle at the chin will briefly contract uni- or bilaterally (Movie 8.8.4). The movie shows how to elicit the reflex. The abnormal response cannot be mimicked in a healthy person.

MOVIE 8.8.4 Palmomental reflex

Snout reflex: Upon gently tapping the upper lip, the mouth will make a brisk or soft snout movement (Movie 8.8.5). The first part shows the normal situation with no response, the second part mimics an abnormal snout reflex.

MOVIE 8.8.5 Snout reflex

Glabellar reflex: with repeated, gentle tapping on the glabella from a position out of the view of the patient, there is a habituation response, and the patient eventually stops blinking in response to taps as shown in the first part of the movie (Movie 8.8.6). In pathologically disinhibited individuals the blinking continues with each tap, indefinitely. This is mimicked in the second part of the movie.

MOVIE 8.8.6 Glabellar reflex

Orientation

The patient is asked about 1- him-/herself (autopersonal orientation), about 2- orientation in time (temporal orientation), 3- location (spatial orientation) and 4- why he/she is hospitalized (situational orientation).

A simple sequence is as follows:

- Situation: Why are you here?
- Person (self): What is your date of birth? How old are you right now?
- Place: Where are we now? (current location)
- Date: day, month, year, season, day of week

SUBSECTION 2

Attention

Simple tests include the following:

- Name months of year forward
- If this is fine: "Can you do the months backward, too?"
- Name days of week forward and backward
- Counting or subtracting backwards from 100 (see section 8.3)

Note: producing backward serial subtractions also significantly challenges working memory capacity (see below).

SUBSECTION 3

Short-term and long-term memory

It is not recommended to start formal testing before the test has been performed in simple terms in a colloquial way. **Most of the items** can already be appreciated while the history is taken and during the formal neurological examination: "What is the name of your roommate?" or "Do you recall the names of your doctors and/or your nursing team?"

A very useful test, in case of any perceived dysfunction, is **subtracting** 1, 3 or 7 (7 being the most difficult of the three) from 100 sequentially. This tests **attention span and immediate memory**, in addition to the direct testing of the abilities in **arithmetic**. If the patient stops, this is always **abnormal**, and if he/she does not recall what the task was going to be, this indicates an attention and/or **working memory** problem.

Digit span is an easy way to assess **short-term memory**. The patient is asked to repeat a series of **digits** (like phone numbers). Each time the task is mastered the length of the digit series is increased by 1 (e.g, 2; 4-7; 3-5-9; 1-5-7-8), until the patient fails to repeat the digits correctly. Repeating the digits in **reversed** order represents a bed-side test for working memory since information stored in the short-term memory (e.g. digits) has to be worked on when putting the digits in reversed order.

A simple version is as follows: ask the patient to repeat the names of 3 common **objects**. You have to **ensure that the information is registered** by having the patient repeat the names immediately after giving the words, until they are correctly repeated. Objects may be pencil – watch – automobile, or more difficult objects like: penny or cent (chosen for category of currency and commonality), hippopotamus (chosen because it's organic but uncommon and multisyllabic), faith (chosen because it's abstract, and cannot easily be catalogued with a set of mental images). Normally, patients are

able to recall at least 2 of the 3 names of objects after an interval of about 5 to 10 minutes (termed **delayed recall**). Note that the delayed recall, even with this short time interval, already involves the **long-term memory**. It is more sensitive to choose unrelated objects, some of which are concrete (purple, apple, etc.) and some of which are abstract (happiness, faith, etc.). Others use short stories or phrases in their **native tongue** like "John Brown, 42 Market Street, London" or similar ones to assess this. If the patient cannot speak (anarthria, expressive aphasia) one may first show and then hide particular items around the room, and ask him/her to indicate the hiding places after a similar 5-10 minute delay.

A simple way to test **short- and long-term memory** in a **single sequence** is described in the following procedure: First one asks the patient to immediately repeat the names of 3 common objects. Then the test of digit span is performed as indicated above challenging the short-term memory store, so that the names of the three objects given before need to be encoded in the long-term memory stores. Finally, we ask the patient again for the names of the three objects (delayed recall for testing long-term memory).

The **long-term memory** is further screened by asking about school education and other milestones of life (**biographic** long-term memory) or events that happened in the past, e.g. what did you do last weekend? (**episodic** long-term memory). If a next of kin is present you may arrange that a short nodding indicates that the information is correct and not confabulated (invented).

Simple tests include the following: "Are you interested in politics? Who is the current president; chancellor, prime minister - or the like?" (depending on home country); "Who was the last one before the present one?" Naming other generally well-known persons is a simple alternative for this test of **semantic** memory.

Note: it is important that the questioning is tailored to the educational level, ethnicity, and native tongue of the individual, as one may miss out on cognitive deficits in the lawyer who can't explain the

American toxic mortgage crisis, but still is able to recall any simple factual recitation. Too simple a test may trigger aversive reactions in a patient with a minor deficit. In most memory disorders e.g. Alzheimer's dementia, there is a strong gradient of time in long-term memory functioning: the longer ago something was encoded the better the recall.

SUBSECTION 4

Testing for Language and Calculus

Glossary

Spontaneous speech and repetition

- Prosody
- Paraphasias
- Neologisms
- Paragrammatism
- Agrammatism
- Speech automatisms
- Broca's aphasia
- Anomic aphasia

Understanding language

- Wernicke's aphasia
- Dyslexia or Alexia
- Dyscalculia or Acalculia

Note: often one sees language abnormalities to be associated with deficits in other higher cortical functions.

Testing for Aphasia

Movies of 3 patients with different types of aphasia are provided for better illustration. All patients were awake and attention was full (see Movie 8.4.1, Movie 8.4.2, Movie 8.4.3).

The free speech during colloquial exchange may already tell the examiner what is wrong. A **systematic** assessment of language function requires testing of all relevant items is following:

Spontaneous speech / language production

- Ask open-ended questions: "What seems to be the problem?" "What do you do for a living?"
  If there is **no** spontaneous speech, ask the patient to count or to enumerate the names of the months. By analyzing spontaneous speech, try to answer the following questions:
  - Does the patient show **high effort** to speak?
  - Is the speech **fluent** or **non-fluent**? This is the most distinctive feature for the classification of the aphasia (Movie 8.4.1).
  - Is the "melody" of speech (prosody) normal or is it poorly modulated (reminiscent of an old-fashioned computer speaking)?

- Does the patient show paraphasias or **neologisms** of any kind?
- Does the patient present **word-finding difficulties**, i.e. he/she fails in finding the right noun and circumscribe words that he/she cannot precisely recall (instead of saying pen, they may say "the thing used for writing")?
- Does the patient not understand what the examiner asks him/her (typical of Wernicke's aphasia, not infrequently combined

with a typical irritable and questioning **mimic** expression indicating that the patient feels lost; however, unawareness of a language problem may occur, especially in Wernicke's aphasia)?

Note: analyzing spontaneous speech also allows for diagnosing dysarthria, i.e. poor articulation. Dysarthria is a motor speech disorder and must be distinguished from language disorders (aphasia).

Repetition

- This is done in ascending difficulty: letter *(„a“)*, words *(„house“)*, long words *(“disproportionality”)*, foreign words *(„orchestra“)*, or sentence *(„the dog barks“)*.
  Repetition of the following phrases tests mainly **short-term** memory:
- “No ifs, ands, or buts” (or equivalent conjunctions in the mother tongue of the patient if he/she is not fluent in English). This sentence is virtually meaningless, unless understood to be an epithet comprised of a conjunction of conjunctions that means “no excuses”.
- “If I am here, then you are there” (doesn’t require familiarity with English grammar, but does have a confusing referential

architecture that makes it difficult to repeat without higher level processing)

- Finally, simplest may be required: "The weather outside is bright and sunny"

Naming

- Show simple objects from your coat pocket or desk: the items are presented to the patient. Common and uncommon objects should be included:
    - Hand, then the fingers (extended test: patient names the presented fingers of the examiner followed by showing the same ones on his/her own hand)
    - Necklace, Necktie, then the knot
    - Watch, then the face or band/clasp
    - Ballpoint pen, then the clip
    - Safety key, and its parts

○ Glasses, then the lens

Speech comprehension

- Following of commands: „Touch your right ear with your left hand." Or, more difficult: "Touch your left ear with your right thumb, *after* touching your nose."

Reading (Alexia)

- Reading comprehension. „Close your eyes tightly."
- Reading aloud a simple text, e.g., from a newspaper or magazine (with reading glasses if needed).
- A multi-step written command for redundancy "Fold this paper, then put it on the ground").

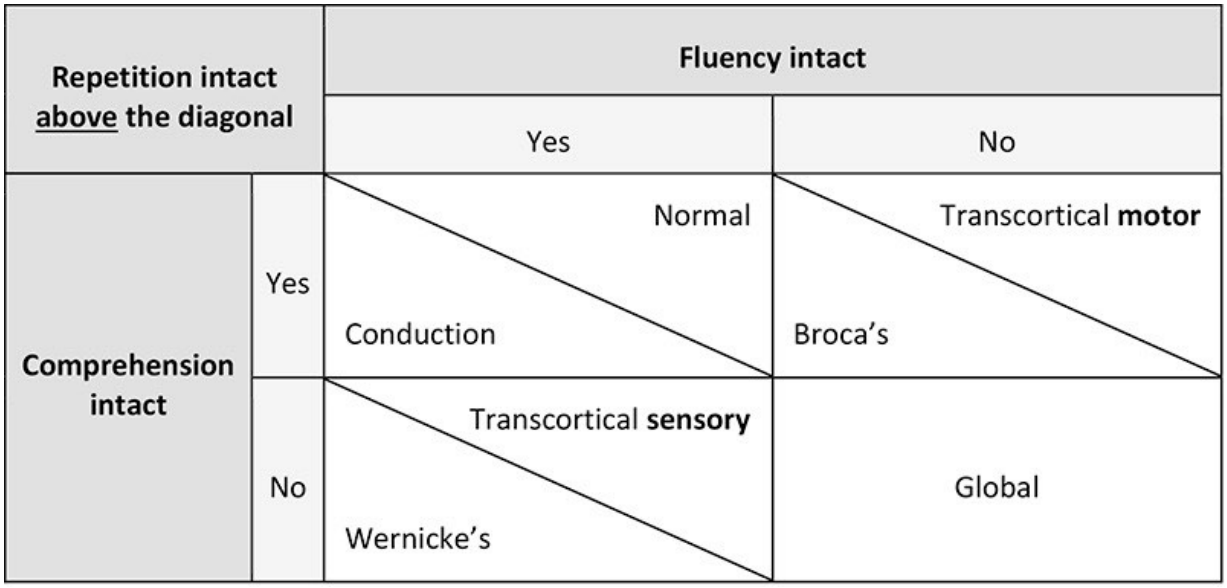
Writing (Agraphia)

- Ask the patient to write a simple sentence from dictation ("Today is a nice day") or to copy a short text.

Note: the mistakes observed in written language are typically the same observed in spoken language

A simple scheme (Figure 8.4.1) may help to classify the most common forms of aphasia:
Examples: a patient with disordered fluency, normal comprehension and still able to repeat is classified as transcortical motor, with inability repeating he/she is classified as Broca's type; a patient with intact fluency, disordered comprehension and intact repetition is classified as transcortical sensory, with inability repeating he/she is classified as Wernicke's type.

FIGURE 8.4.1 Simple Bedside Classification of Aphasia.



These types may match with typical lesions found on MRI/CT scans.

Whenever deficits are noted a more **formal testing** may be done in the respective realm:

repeat text read by the examine

name more complex objects that are presented to the patient

read a more difficult text

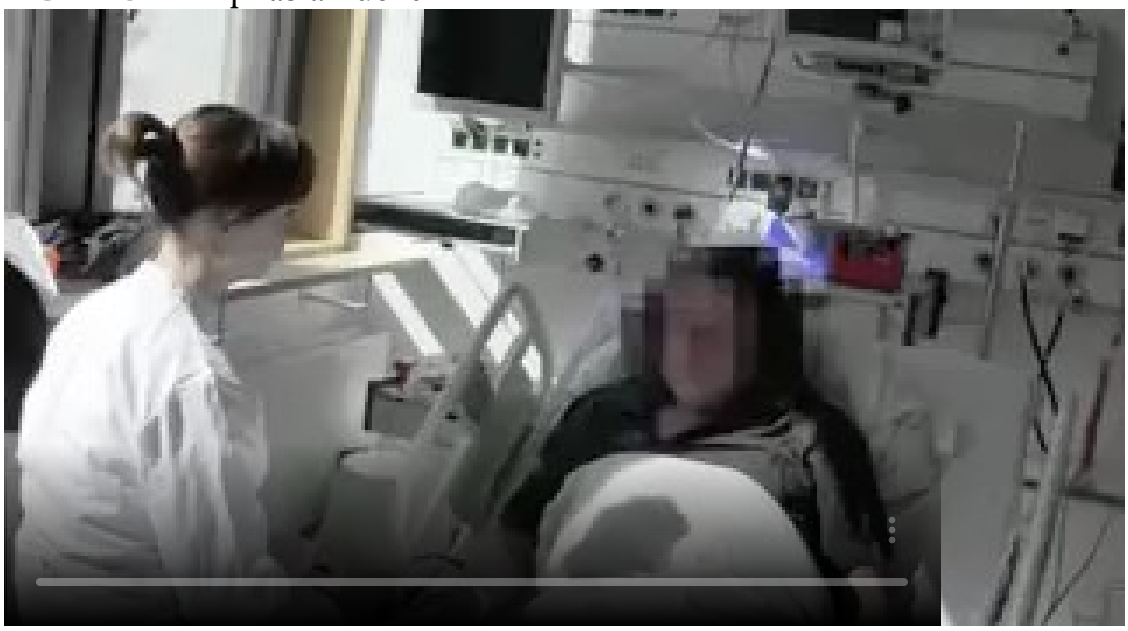
write a more complex statement from dictation

- perform complex syntax such as "before doing B, do A", follow multistep commands

Note: even if not formally tested, by partaking in the neurological exam, aphasia can often be assessed and can be commented on.

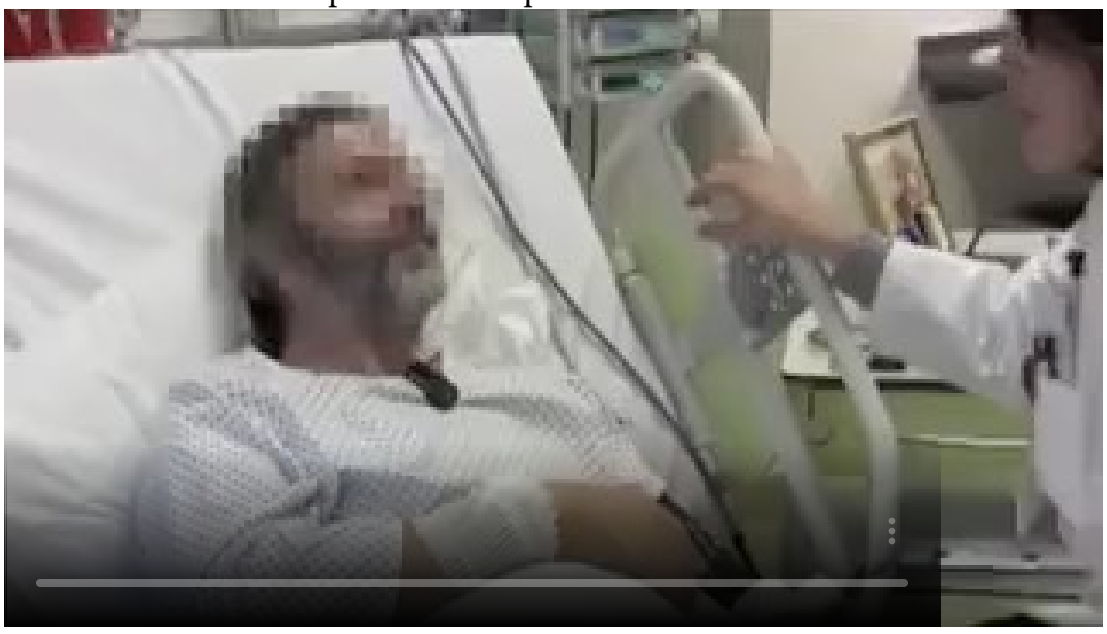
Movies of patients with aphasias

MOVIE 8.4.1 Aphasia fluent



Movie 8.4.1: 49 year old pharmacist who presented with fluent aphasia due to ischemic stroke of the left posterior temporal cortex. Testing was performed 2 days after symptom onset. Spoken language is characterized by many semantic (e.g. “boy” instead of “doctor” or “answers” instead of “questions”) and phonematic (e.g. during repetition “lottir-litor-lolitor” instead of “lottery”) paraphasias.

MOVIE 8.4.2 Global aphasia with speech automatisms



Movie 8.4.2: 57 year old artist who presented with severe global aphasia. His spoken speech is characterized by prominent speech automatisms (e.g. “only for you – understand you only”) as inappropriate utterances that did not fit into the context and which were repeated by the patients many times (not shown). In addition, language comprehension was also severely affected. He had a haemorrhage of the left temporo-parietal cortex due to cerebral venous thrombosis.

MOVIE 8.4.3 Aphasia non-fluent

Movie 8.4.3: 69 year old engineer who presented with severe non-fluent aphasia, caused by an ischemic stroke of the left frontal cortex. In addition to his non-fluent aphasia, speech production is affected by apraxia of speech, a motor speech disorder which often accompanies non-fluent aphasia in Broca’s aphasia.

Testing for acalculia

After making sure that the patient has his/her **reading glasses**, test for problems with numbers and simple calculations (dyscalculia or acalculia), by first having the patient, in a colloquial way, **read 2-5 digit** numbers (“What is that phone number on your phone?”). If unable to do so, dyscalculia, dyslexia, or dysphasia may alone or in

combination be the underlying deficit. Then ask the patient to do **simple arithmetic**. One can have the patient perform **subtracting** serial 3s and 7s (100-93-86-79-72) which is part of an evaluation of attention span as well. Some patients find word problems easier to work with so it may help to have the patient perform more functional math problems, such as telling you how much change is in a nickel, dime, and quarter or how many apples can be purchased with $1.75 if apples cost $0.50 each. Acalculia is often associated with other deficits like aphasia, and **dyslexia**.

Note: acalculia is often not noticed by the patient and may be overlooked by the examiner if not properly tested.

Praxis and spatial and bodily attention

Testing for limb apraxia

Before testing for apraxia there should be some indication of abnormality from observing the patient throughout the previous steps of the examination. Since aphasia and apraxia may co-occur in some major hemispheric disorders (e.g. in stroke) it is not always possible to test the isolated neuropsychological pathology in early stages of CNS disease.

Apraxias are disorders of higher motor function. They may show as disorders of planning or performing certain actions such as gestures, and object or tool use. They can only be diagnosed with certainty, if primary sensorimotor deficits are absent. Although they mostly affect both sides of the body, in rare cases, they may affect only one (in this case, mostly the left) side. Therefore, **all tests should be done on either side.**

Note: Here we focus on limb apraxia, although apraxia may affect many other parts of the body, including face (buccofacial apraxia) or speech (apraxia of speech).

Imitation

Ask the patient to **imitate** meaningless hand-head **gestures** as shown in Figure 8.5.1.

Ask the patient to imitate a symbolic gesture, using common gestures, such as:

- Threatening someone.
- Waving one's hand to stop.

- Wave the hands good-bye.
- Saluting like a soldier.

Pantomime or tool use

Ask the patient to use a common tool, such as:

- Lighting a match.
- Drinking from a cup.
- Stirring a cup.
- Unlocking a door.
- Combing the hair.
- Drilling a hole.
- Sewing or knitting movements.
- Hammering a nail.

Note: In contrast to imitation, where the patient can copy the movement from the examiner, pantomime requires the internal retrieval of a motor program.

Real tool use can be tested with readily available tools:

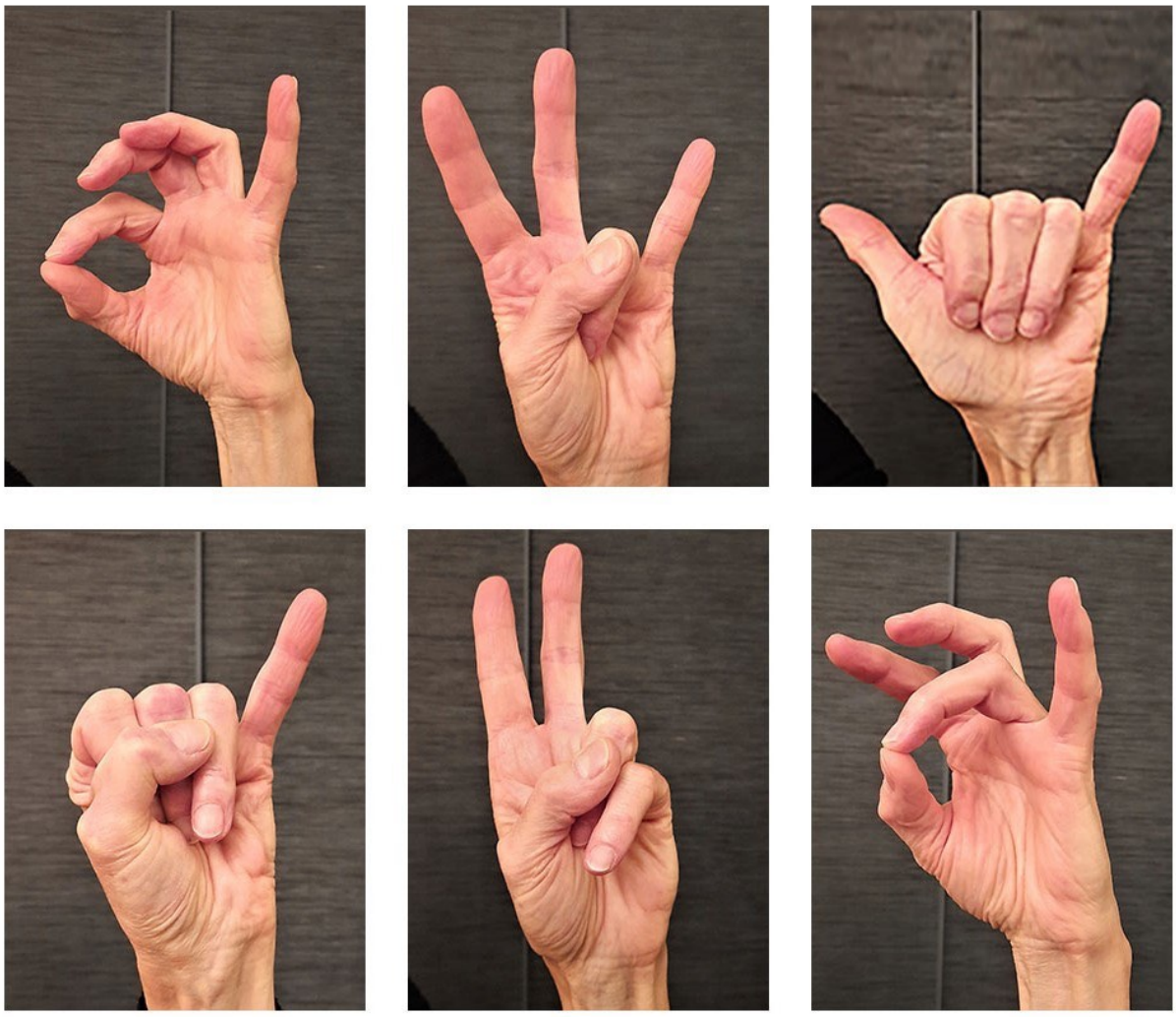
Using a hammer or a pair of scissors.

Using a hole punch on a piece of paper

Putting on and taking off eye glasses.

Examples for **imitation** of abstract gestures are shown in Figure 8.5.1:

FIGURE 8.5.1 Imitational praxis testing with hand positions and forms



A collection of arbitrary hand positions (a) and forms (b) that are presented by the examiner and should be copied by the patient as part of imitational praxis testing. Additional forms may be presented.

Sequential actions may be tested by asking the patient to pretend he/she is taking out a can of milk, pouring it into a mug, and starting the microwave, or any similar series of actions

Note: inability to perform a sequence of actions, in the absence of more elementary deficits is classically termed ideational apraxia, in contrast to the inability to imitate or pantomime which is classically termed ideomotor apraxia).

Movies of 2 patients with different types of apraxia are demonstrated for better illustration. All patients were awake and attention was full.

MOVIE 8.5.1 71 y old right-handed lady with a right sided brachiofacial hemiparesis of sudden onset.

About 3 weeks after the stroke, the video showing apraxia (and aphasia) was taken while weakness had already remitted. MR imaging revealed a left temporoparietal infarction. The first segment (Movie 8.5.1) shows the attempt of the patient to imitate symbolic gestures presented to her by the examiner; first, a threatening gesture is presented by the hand, then followed by a gesture indicating "don't worry, it does not matter".

MOVIE 8.5.2 71 y old right-handed lady with a right sided brachiofacial hemiparesis of sudden onset.

In the second movie segment (Movie 8.5.2) the patient is asked to imitate two consecutive meaningless gestures (cf. Figure 8.5.1).

MOVIE 8.5.3 71 y old right-handed lady with a right sided brachiofacial hemiparesis of sudden onset.

In the third segment (Movie 8.5.3) the patient is asked to pantomimically demonstrate how to brush her teeth which she fails to accomplish. However, the movement that she does produce has certain repetitive elements resembling elements of the target movement.

MOVIE 8.5.4 71 y old right-handed lady with a right sided brachiofacial hemiparesis of sudden onset.

In the fourth segment (Movie 8.5.4) tool use is tested. The patient is unable to demonstrate how to use a hole puncher.

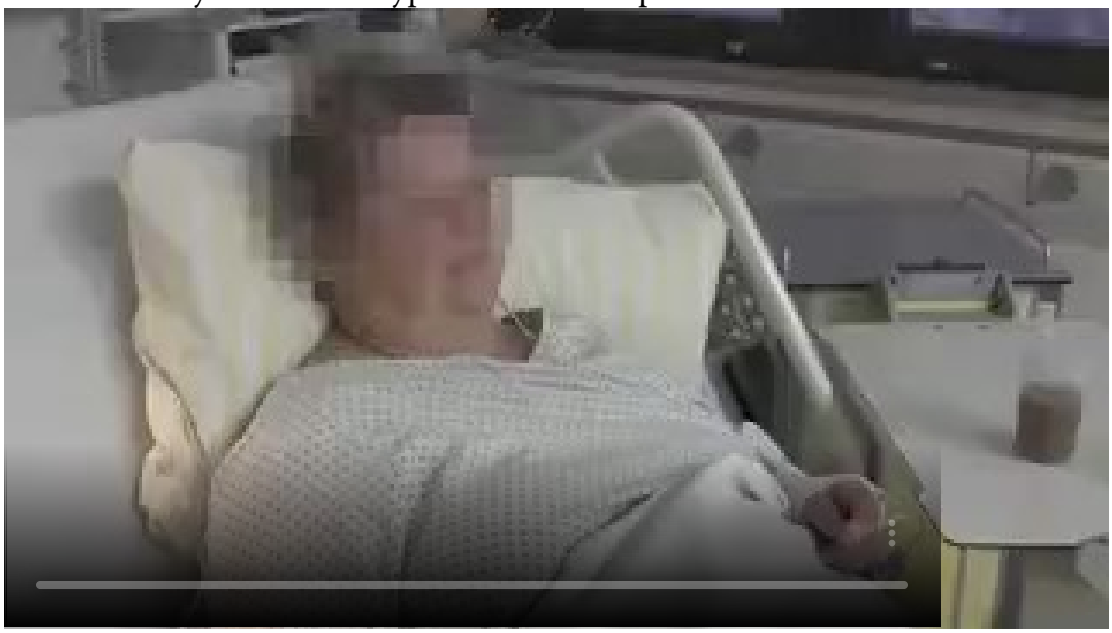
MOVIE 8.5.5 60 y old right-handed lady with speech difficulties of sudden onset with a history of arterial hypertension and prediabetes.

CT scan revealed left parietal haemorrhage. The first segment (Movie 8.5.5) shows the attempt to imitate meaningless gestures (cf. Figure 8.5.1) presented to her by the examiner.

MOVIE 8.5.6 60 y old right-handed lady with speech difficulties of sudden onset with a history of arterial hypertension and prediabetes.

In the second movie segment (Movie 8.5.6) the patient is given a hammer, but has no idea how to use it when asked to drive a nail into the wooden board.

MOVIE 8.5.7 60 y old right-handed lady with speech difficulties of sudden onset with a history of arterial hypertension and prediabetes.



In the third segment (Movie 8.5.7) the patient is asked to pantomimically demonstrate how to screw in a light bulb and then to show how to brush her teeth. Providing her with a small wooden stick (pencil) does not cue the action of brushing.

SUBSECTION 6

Executive function

Executive functions involve the following steps: analyze a task; plan how to address the task; organize the steps needed to carry out the task; develop a timeline for completing the task (scheduling); complete the task in a timely way; ignoring inappropriate distractions; and appropriately adapting to changing circumstances. There may be overlap with apraxia.

While there are many tests of executive function, the applause sign (M 8.6.1) and the Luria sequence (M 8.6.2) both represent useful bedside tests to assess a patient's ability to execute a simple task.

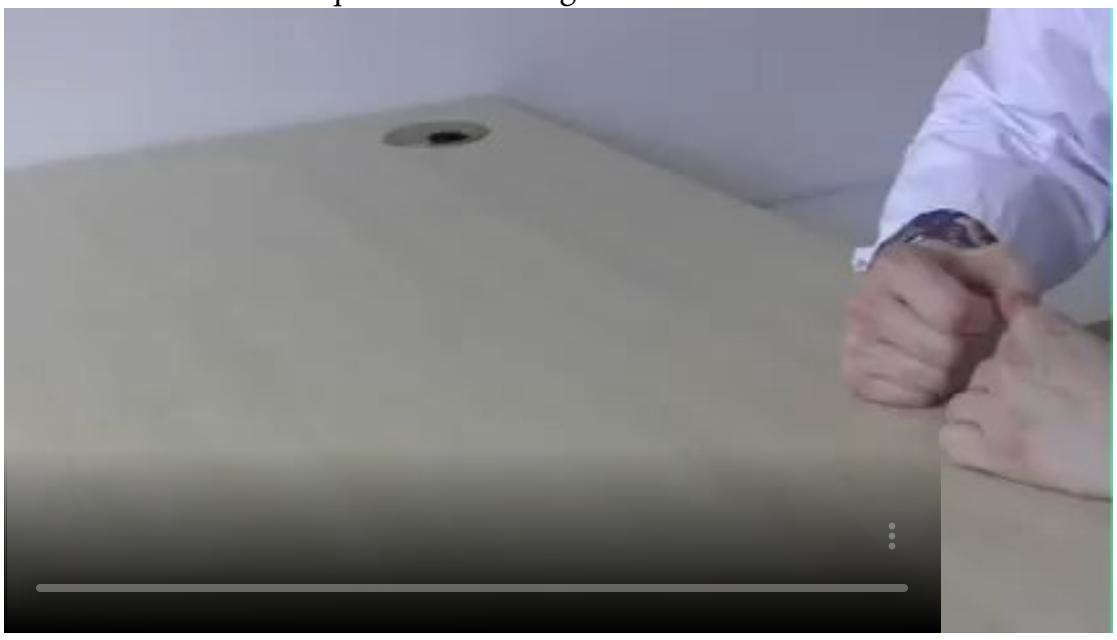
Note that none of these tests can be done or interpreted with confidence in an intoxicated or stuporous patient, or in case of a lesion-associated attention deficit.

Movie showing the applause sign in a patient with dementia (M 8.6.1). The patient is asked to clap three times as quickly as possible just repeating the same three claps as demonstrated by the examiner. The performance of the subject is normal when he or she claps only three times and then stops. The sign is considered as marginally abnormal if the patient repeats the task with a few more than three claps; the sign is clearly abnormal if the patient repeats the task with (almost) endless clapping. A positive applause sign is indicative of frontal disinhibition.

MOVIE 8.6.1 Applause sign in a patient with dementia

Movie showing the Luria sequence (M 8.6.2). The patient is asked to repeat the gestures shown by the examiner in the correct way and including the correct sequence. Wrong order or incomplete repetition of the sequence is indicative of executive dysfunction. Another simple bedside test is to ask the patient to perform a sequential gesture task producing hand positions as in "rock", "paper", and "scissors".

MOVIE 8.6.2 Luria sequence of hand gestures



Visuo-spatial orientation, testing for neglect, anosognosia

Visual-spatial abilities: The patient is asked and observed for a potential disorder of spatial organization.

- can the patient **read** the clock?
- is the patient able to **draw** a clock? (Ask the patient to draw the numbers inside a circle have him/her placing hands so as to indicate "10 past 10 o' clock"). Any abnormality in this test is noted.
- can the patient draw a bicycle or a plane?
- can the patient copy certain graphic elements?
- can the patient find his/her way back to his/her room?

Neglect: lateralized attention deficit (hemi-neglect) for tactile stimuli on the side contralateral to a hemispheric brain lesion (more often the right hemisphere) while the perception itself is not disrupted. Equivalent syndromes exist for the visual and auditory system. In practice neglect is often confounded by hemiparesis (usually on left) or hemianopia. In sensory terms, this is not identical to the **extinction** of simultaneously applied tactile stimuli (see Section 6). Neglect can be observed in spontaneous behavior such when the patient misplaces his/her glasses by missing the ear on the affected side, or when he/she fails to clean one angle of the mouth after eating.

Simple bedside tests are as follows:

The patient is approached from the left and from the right side and confronted with a simple verbal command or question. There will be a deficit (**slowed or missing reaction**) on the side with ne-

glect. Therefore one should **avoid** examining the neglecting patient from the impaired side as it may affect the quality of your exam.

Asking the patient how many people or how many males and females are in the room, and then asking to point out the people mentioned, may also suggest a **neglect**.

Further bedside tests, in case of any abnormality:

Note: some of these tests may provide similar information

- Reduced or absent perception of a stimulus on the affected side as compared to the healthy side.
- When reading a full-size text, the patient will **omit** part of the text on the **affected** side while **visual** fields may be **largely normal**. In contrast, a patient with hemifield visual deficits, will likely **search** for the missing text.
- The patient may not perceive the **full shape** of items in the room by neglecting parts on the affected side.
- A quick screen can also be performed by asking the patient to grab the **middle** of the length of the stethoscope and any gross deviation should be documented.

Paper-Pencil-Tests:

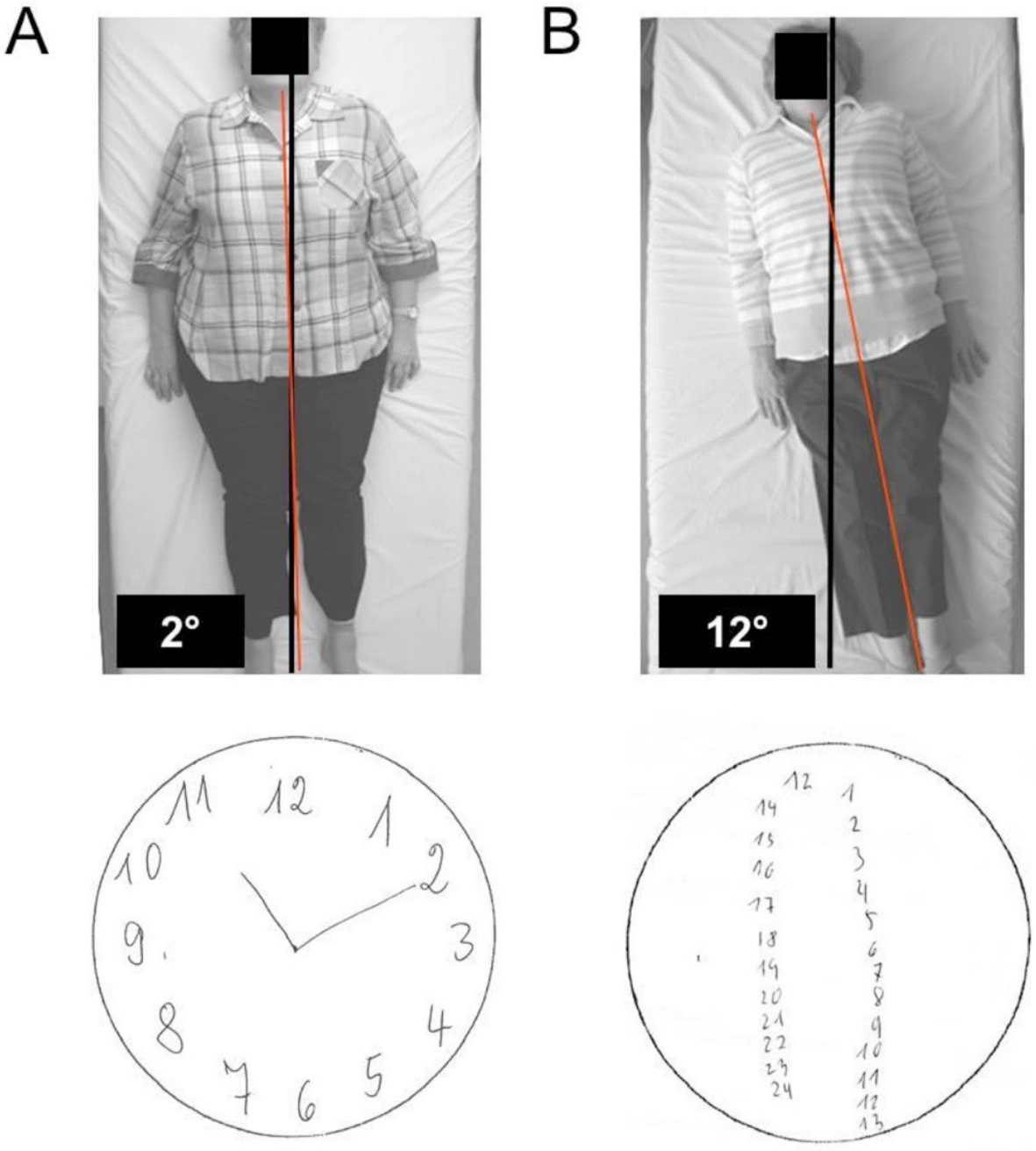
- Simple drawing of flowers may reveal **missing leaflets** on one side (similar with copying).
- A group of straight lines with different lengths are drawn on a simple pad. The patient is asked to mark the **midpoint of each line**. With hemi-neglect the apparent midline is shifted away from the afflicted side of the visual field because that part of the line is perceived shorter than real. This **line bisection test** allows quantitation of the deviation of the attended field from the midline.

- An even more sensitive test is the **letter cancellation test** which involves cancelling our target letters placed amidst of distracting letters.

Associated disorders include **anosognosia**, i.e. the patient does not realize the deficit, and **anosodiaphoria**, i.e. the patient is aware of the deficit, but seems unbothered by its presence (*"la belle indifference"*).

Some patients may display a peculiar abnormality when lying in bed or when asked to lie down in bed from a sitting position at the edge of the bed. Healthy people spontaneously orient their body in parallel with the longitudinal axis of the bed. Affected patients lie **obliquely in bed**, i.e., the longitudinal axis of the body deviates from the longitudinal axis of the bed (see Figure 8.7.1). This behaviour can easily be recognized at the bedside by nurses or doctors alike and may be termed **body axis orientation failure** (Kraft et al., 2009). This has been demonstrated to be associated with cognitive impairment (Kraft et al., 2009) and may be associated with neuropsychological deficits such as visuo-spatial disorientation as seen in drawing of the right clock by the lady on the right photograph.

FIGURE 8.7.1 Illustration of two patients demonstrating intact (left panel) and failure (right panel) of body axis orientation. Angles of body axis orientation are indicated in the figure. Corresponding clock–drawing tests are shown below showing a profound constructional apraxia on the right.



Judgement, abstraction

These are simple screening tests evaluating intellectual functions. Validated tests like the Mini Mental Status Exam and various other psychological tests should follow if indicators of dementia become evident.

Abstraction

This task addresses the understanding of complex sayings and proverbs. One should use well known sayings and proverbs that are being used in the home country and are familiar to the ethnic background of the patient.

- “A golden hammer can break down an iron door” → Money is power
- “The tongue is the enemy of the neck” → What you say can get you into a lot of trouble
- “The grass is always greener on the other side of the fence” → Other people’s circumstances often appear better than your own
- Comparisons (tests for understanding categories - start simple so the person understands):
    - How are an apple and an orange the same → they’re both fruit (also they are both round, but this is

where you can clarify you mean fruit)

- How are a watch and a ruler the same → they are both systems of measurement
- How are a chair and table the same → they are both furniture (being made of wood is too concrete and not always correct)

Judgement

- What do you do if you find an addressed, stamped envelope lying on the ground? → put it in the mailbox, or the like
- What do you do if you're in a crowded theater and someone yells fire? → head to the nearest exit, provide help for your next neighbor if needed, or the like
- What would you do if you see a bad traffic accident → call the police and the ambulance, or the like

SECTION 9 – FUNCTIONAL NEUROLOGICAL DISORDERS - CONVERSION SYNDROMES WITH CLINICAL EXAMPLES

Note: this section only provides an overview for neurologists in training. Much of what is laid down needs extended training with case presentations and videos that illustrate the various patterns that one may encounter. In practice, functional disorders need to be considered if history and findings appear in themselves inconsistent or incongruent. Only then the examiner is alerted to be looking for actual proof of the presumed inconsistencies and paradoxical findings needed to make a positive diagnosis.

Introduction

Conversion syndromes are listed with psychiatric disorders but patients present frequently to the neurologist first. All patients depicted with this manual were seen by the first authors between 1986 and 2015 and have given consent to be filmed for neurological educational purposes.

An experienced neurologist is alerted when signs and symptoms do not fit with the history of present and past illnesses and with well-defined neurological disorders. For the unexperienced this group of conditions is very difficult to diagnose. Functional and somatic disorders may be present simultaneously complicating the matter further. There are some potential clues when attempting to diagnose functional disorders. The various signs are named after the most closely matching organic neurological disorder with the prefix

“pseudo” indicating that the specific neurological sign is only mimicked

An approach to the diagnosis of functional neurological symptoms

The diagnosis should **not** be based solely on the presence or absence of psychiatric illness or on traumatic life events. Co-morbidity with anxiety disorders may be up to 30%, indicating that **many** patients with anxiety have **no** conversion disorder. Conversely, in a sizeable portion of patients with functional disorders, no overt psychiatric disease may be found.

The following common neurologic conditions may be mimicked as part of the functional (conversion) disorder:

- **Hyperkinesias**
  Tremor, Involuntary Movements, Tics, Blepharospasm, Torticollis, Opithotonus
- **Gait disorders**
  Astasia/abasia, Falls (without major injuries), Hemiparesis
- **Others**
  Seizures, Complete Paralysis, Weakness of any part, Stiffness, Spasticity
- **Disorders of sensation**
  Deficits not following anatomical rules, non-reproducible deficits with incongruence of various findings in the sensory realm, deficits in special senses (hearing, vision) incompatible with behavorial observations in the test situation
- **Speech disorders**
  Stuttering, trembling voice, aphonia

Functional Motor Symptoms

Note: clues evolve by systematic neurological examination. Because functional disorders are a diagnosis of exclusion, and because they can coexist with concomitant organic neurologic disorders (e.g. when patients embellish clear-cut neurological symptoms and signs), a positive diagnosis needs confirmation by follow-up and reappraisal of the differential diagnosis.

Weakness

The examination of the patient requires systematic testing of the neurologic functions, taking advantage of redundant/parallel pathways of assessment. If incongruences and inconsistencies become apparent, various test are at hand to help proving that the apparent weakness is rather pseudo-paresis. In general, the suspicion rises when upon diversion of attention away from the apparently abnormal movement, one can demonstrate that normal movement is possible.

A classic example of a test indicating functional muscle weakness is Hoover's sign for functional weakness. This is most easy to demonstrate in people with unilateral leg weakness. The patient is asked to perform hip extension with the weak leg and the examiner assesses power. The patient is then asked to perform a strong hip flexion with the good leg while the examiner's hand is kept underneath the **weak** leg. When the patient performs hip flexion, the previously weak leg can be felt to extend **strongly** at the hip. This test provides a clear dissociation between the patient's inability to move the leg when directly trying and the ability for this same movement to happen when the movement is triggered by an automatic postural response. Another iteration of the **Hoover's sign 2** (named reverse Hoover's sign in Tab. 9.4.1) is performed when the weak leg is being lifted while the examiner's hand is kept under the good leg.

When the apparently weak hip flexion is ongoing, if the good leg is able to be lifted off of the bed (after confirming normal extensor power), then the patient is presumed to not be providing real effort to lift the "weak" limb off of the bed.

Additionally, a recently validated **abductor sign** has better sensitivity and specificity for functional leg weakness (http://jnnp.bmj.com/content/75/1/121.full).

First, as a **preparatory** test, the examiner tells the patient to **ABduct both legs** simultaneously as hard as possible. **Weak abduction of the supposedly paretic leg** is noted: the good leg remains fixed in an abducted position fully opposing the examiner's hand, whereas the paretic leg moves towards the midline. Next, as the **main test**, have the patient **ABduct each leg in turn**, while the **examiner observes the behaviour of the unABducted leg**, which should initially be placed in an ADducted position along the midline. The examiner directs the patient's attention solely to the ABducted leg, so the patient thinks this is the leg being tested (Figure 9.2.1).

- In **organic** paresis the weak paretic leg is always mobile, while the good leg is not:
    - When the patient is told to ABduct the good leg, the good leg remained fixed in an abducted position, whereas the paretic leg, which must normally exert a synergic ABducting force

to oppose the examiner's hand, was overpowered and moved in the hyperad-ducting direction.

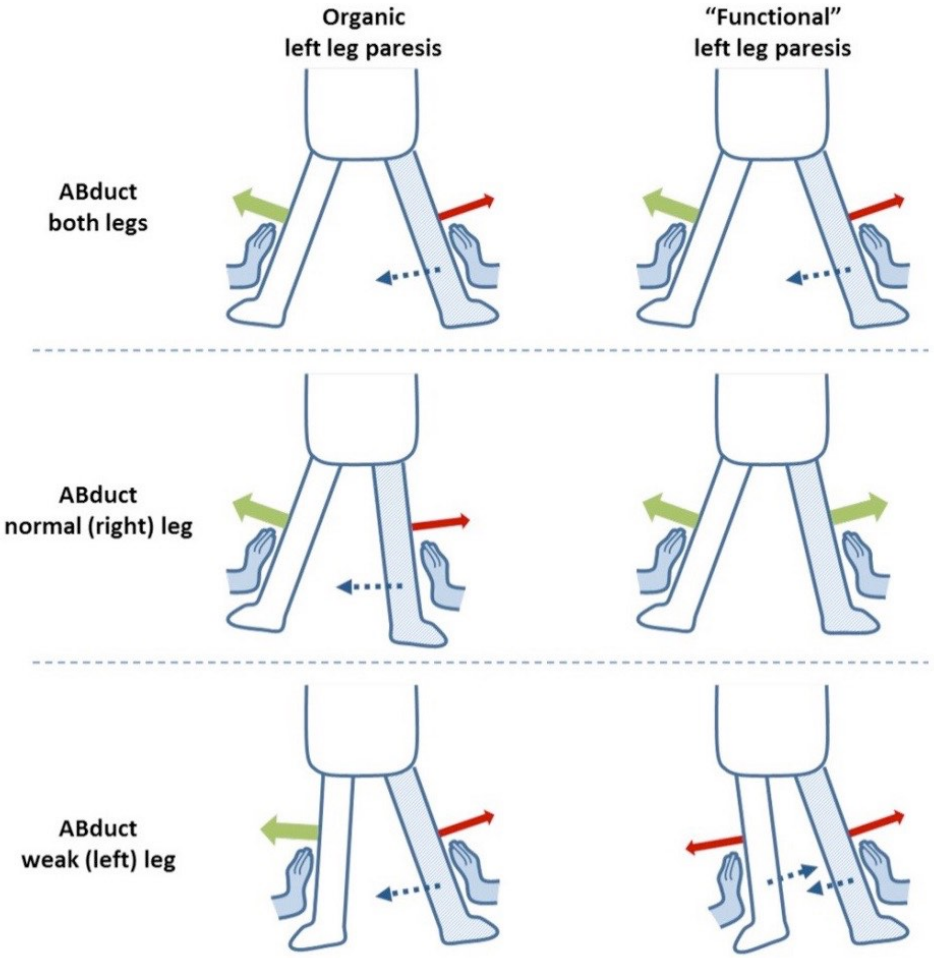
    - When the patient is told to ABduct the paretic leg, it is overpowered by the exam-iner's hand, whereas the good leg exerts a strong synergistic force and re-mains fixed in its original position.

- In **non-organic** paresis the "paretic" leg is strong when not tested, and both legs are weak when the "paretic" leg is tested:
    - When the patient ABduct the good leg, not only is abduction of the good leg strong, but the synergic

opposition of the "paretic" leg is also strong, and the latter remains fixed in its original position.

- When the patient is told to ABduct the "paretic" leg, the entire movement set becomes weak (i.e. ABduction of both legs).

FIGURE 9.2.1 Comparison of results for ABductor sign test in organic and "functional" weakness of the left (shaded) leg



The patient's focus must be directed to the ABducted leg being tested. When both legs are ABducted and an ADducting force is applied by the examiner to both legs, the weak leg drifts inward in both cases. When the normal leg is ABducted and an ADducting force is applied by the examiner to both legs, an organic paresis will have the weak leg drift in, while the "functional" patient will have strength in both legs. Conversely, when the paretic leg is ABducted and an AD-

ducting force is applied by the examiner to both legs, an organic paresis will still have the weak leg drift in, while the "functional" patient will demonstrate weakness of both legs, letting them drift inward. See (http://jnnp.bmj.com/content/75/1/121.full).

It is possible to perform a similar manoeuvre to the Hoover's sign in the upper limbs: something called the "flex-ex" test. This is based on the normal phenomenon that when someone performs a **strong** elbow flexion on one side, there is an automatic extension that occurs at the opposite elbow. This test has **less clear** sensitivity and specificity compared with Hoover's sign, but can be useful to demonstrate normal power is possible in an apparently weak upper limb.

Another common manifestation of functional weakness is a high degree of variability of the muscle weakness during strength testing. Here, with encouragement, patients are sometimes able to generate near full power for a second or so, before the power suddenly stops, causing the limb to **"give way"**. This sign is not definitively proving pseudo-**paresis** because it is important to note that people with severe pain or fatigue from any cause can have give-way weakness, and so this sign needs to be considered in the clinical context of the patient. Often the patient demonstrates via gestures and facial expression that he or she is trying really hard. Importantly, patients are not able to accurately determine the degree of force they apply. So, having the patient initiate the effort, while the examiner progressively increases resistance until maximal force, may assist in eliciting the maximum possible force of the patient. If the patient doesn't exert force at the outset or gives way at negligible resistance, then cooperation of the patient is limited. Pain associated with the task should be excluded.

Tremor

Tremor is a very common form of hyperkinesia. Functional **tremor** may be present at rest and exaggerate with arms outstretched or bended and may become a bizarre **postural tremor**. Indicators of its

possible functional nature are the following (see Movie 9.6.3 and Movie 9.6.5):

1. Distractibility.
    a. Usually, the tremor gets slower, is less intense, will become arrhythmic, or will have pauses when the patient is asked to perform another task needing attention.
    b. Useful distracting tasks are involving the patient in a discussion, counting numbers backwards with eyes closed.
    c. Additionally, by having the patients perform skilled movements with the non-

tremoring hand, two effects can be seen:

i. Changes in the "tremoring" limb.

ii. Inability to perform the task in the normal limb, due to too much effort being dedicated to maintain the "tremor".

1. Entrainability.

a. Because there is only one volitional, central pattern generator, by having the patient perform tapping/moving tasks in other body parts (specifi-

cally the contralateral unaffected limb), the functional tremor will take on the frequency of the volitional tapping.

b. Useful tasks include asking for rhythmic squeezing of the examiner's hand or tapping with different frequencies.

1. Ballistic pause.

a. When performing a ballistic movement, the tremor will briefly pause for 1-2 seconds.

b. Have the patient use his/her non-tremoring

pointer finger to touch the examiner's fingertip as quickly as possible after it appears.

1. Multiaxial or axis variability.
    a. Tremors tend to affect a muscle group that performs a specific function (e.g. flexion of the fingers), and almost never intermittently activate antagonistic groups (i.e. flapping, head shaking, etc.). If the "tremor" affects multiple different actions (e.g. up/down and lateral/medial wrist deviations) at the

same time, be suspect, and describe what you see.

b. Tremors consistently affect the same muscle groups, so they should not vary with activities, just think of the pill rolling tremor of PD happening no matter how the limb is held.

c. The "tremor" may change its direction and pattern with varying co-activation of hand, arm, and shoulder muscles spontaneously or upon applying a distracting task.

(Pseudo-) Dystonic Movements

Historically, most of the dystonias now accepted as organic brain disorders have long been considered being of a functional nature. With this in mind the classification of abnormal motor patterns as being functional (pseudo-) dystonia should be made only if the pattern is bizarre, variable over time, and beyond any familiar pattern in organic dystonia. Moreover, distractibility should be obvious. The best way to document changes in pattern is video filming at serial examinations (see Movie 9.6.1, Movie 9.6.4 and Movie 9.6.6).

One particularly challenging aspect in the evaluation of functional dystonias is the existence of "sensory tricks" that aid patients with organic dystonia. Some patients with organic dystonia (particularly cervical dystonia/torticollis) are able to alleviate the dystonia with touching certain areas. Don't discount the organic origin of a dystonia, solely if it can be eased with "gentle assistance".

Myoclonic, Ballistic, and Choreatic Movements

Several organic movement disorders may produce mixed hyperkinesias of the myoclonic, ballistic, or choreatic type. If continuously active, functional (pseudo-) myoclonic jerks show distractibility as with tremor. Variations in pattern upon distracting manoeuvres are common and this may also happen spontaneously over time. While intermittent, appendicular myoclonus is often difficult to discern as organic from the exam alone, "propriospinal myoclonus" characterized by flexion and/or extension jerks of the abdominal/axial muscles is commonly recognized to be a functional disorder.

A good way to document changes in pattern is video filming at serial examinations.

The classification as a functional disorder is more difficult in episodic cases. Long-term EEG recordings should be done to rule out epileptic myoclonus and referral to a functional neurophysiolo-

gist can help assess for volitional potentials in specialized testing (see Movie 9.6.1 and Movie 9.6.3).

Gait and Stance

Functional gait disorders may often have bizarre patterns with the patient dragging one limb behind them as if they had a **spastic** hemiparesis, often combined with apparent **ataxia** (pseudo-ataxia) and gross jerky or throwing movements of the trunk to either side. This contrasts to the fact that injury from falls are are uncommon. While being examined they may initiate a fall only if a supporting person is nearby preventing injury.

With the pull test (cf. Figure 5.5.1 and Movie 5.5.1) the patient shows similarly gross movements of the trunk but with efficient body control and again no fall unless support is readily available.

When the patient's gait is widely abnormal (often likened to the individual "walking on ice"), consider the effort required to produce such a gait without falling. Patients will be seen to walk with legs slightly bent and violently shifting gravity while transitioning from leg to leg. This requires more strength than standard walking, and despite this the patient will usually not fall, unless able avoid injury with a proximate examiner or bed. The usage of the term **astasia-abasia**, has come into popularity to describe this classically functional gait.

The patient with a paralytic variant often comes to clinic in a wheel chair and appears unable to stand upright (pseudo-astasia). Once lifted with bilateral support the examiner will note that despite changing his/her supporting force the patient does not fall. If the patient is accepting the use of a walker, gait will be with short and slow shuffling steps. Asked to move 3 steps backward, with protective support being present, the patient may indeed walk backwards and sometimes better than forward which is incompatible with true leg weakness (see Movie 9.6.2).

In milder cases, variants of the Romberg test can be done while the patients is distracted by counting backward with eyes closed showing a much better stability than with open eyes, again incongruent with an ataxic gait.

SUBSECTION 3

Functional Disorders of Vision, Hearing, and Cutaneous Sensation

Sensory perception is by definition not objective. Focal **paresthesia**, **allesthesia**, and **dysesthesia** are very common symptoms but do not in themselves always reflect disease. They become potentially meaningful if they are severe and fit a classical organic syndrome, e.g. hemispheric disease with contralateral sensory deficits in stroke or brain tumors. By means of careful examination incongruences of the findings can be defined including too much variability with repeated testing or patterns contradicting the basics of anatomy and neurophysiology (see sensory examination in sections 2 and 6).

Functional blindness or visual impairment, are often the most amenable to evaluation at the bedside. In patients who are reporting a visual field defect, "tunnel vision" is the most common functional variant. A good way to assess this is to have the patient read sections of text at different distances, if the same section of text is always read regardless of distance, then suspect a functional disorder. This is because the field of view broadens the further away an object is, based on how light transmits through a lens.

In the assessment of hearing abnormalities, performance of the Rinne and Weber tests (section 2) can aid in assessment of hearing loss, but ultimate confirmation is often dependent upon the usage of evoked potentials.

Functional abnormalities in cutaneous sensation are common in neurology clinics including the symptoms numbness, tingling, and pain. The most common findings are **hypoesthesia** or even anesthesia/analgesia in limbs, trunk, or genitals. At the bedside great care should be taken to standardize the cutaneous stimulation techniques, in particular with pin-prick and temperature sensation. Mild differences left-to-right or proximal-to-distal may be appreciat-

ed as a threatening symptom by some patients and do not in themselves define a disease. There are simple bedside tests defining cutaneous sensation abnormalities as functional:

- Pseudo-hemispheric syndromes with left-right dividing borders in the body midline ("midline split"; see Figure 2.4.2)
- Identical, i.e. overlapping sharp-edged borders in pain and touch perception (see Figure 6.1.3)
- Identical **two-point discrimination** borders upon static vs. dynamic two-point stimulation (see Figure 6.1.5)
- Incongruence between tests, such as **two-point discrimination** and stereognosis
- Focal reductions in vibration sense
- Right-left differences in vibration perception at the skull and sternum (because these structures vibrate as a whole, bilaterally)

Abnormalities in one or several of these tests raise suspicion as to a functional disorder (see sections 2 and 6 for more detail).

SUBSECTION 4

Functional Neuropsychological Disorders, Pseudo-dementia

Functional disorders of this category are common. The examiner aims at finding the core symptoms and the perceived deficits by engaging in a colloquial history taking. Start by looking for familiar patterns in *sensu strictu* organic brain disorders like monopolar depression, e.g. expressed as a midlife crisis, dementia, **hallucinations**, phobias or early stages of schizophrenia and of narcotic drug abuse. In talking with the patient, one must assess what type of memory loss is reported and systematically test these domains (Section 8). Anything that does not plausibly link the perceived deficits together or does not allow finding a likely unifying clinical diagnosis should be carefully explored for a conversion disorder. Of note, the loss of identity (i.e. a patient forgetting his/her name) is almost certain to be psychogenic in origin, because this declarative memory is the most fundamentally learned feature in life. Isolated memory loss is not uncommon as a functional disorder following major threatening life events. In addition, concomitant somatoform disorders may be found helping in finding the correct disease pattern. Formal neuropsychiatric batteries often include measures to assess effort/malingering, and thus this must be specified in consultation.

TABLE 9.4.1 Characteristic "functional" signs and mechanisms in conversion disorders

Sign	Purpose	How to Perform	Interpretation / Reasoning
Hoover's Sign	Assess for non-organic leg weakness	**1.** Have pt lay on his/her back w/ legs extended. **2.** Start by testing the strong leg. Place your hand under the heel of the "weak" leg, and place your other hand on the thigh of the strong leg. Ask pt to lift the strong leg into the air against your hand. **3.** Pay attention to the "weak leg".	•A normal response is to press the heel of the contralateral leg downwards while elevating the leg. •If pt depresses heel w/ good strength while elevating the contralateral leg, there may be functional weakness.
Reverse Hoover's Sign	Assess for non-organic leg weakness	Use the same technique as the Hoover's Sign except place your hand under the heel of the strong leg and ask pt to lift the "weak" leg against resistance.	•If the pt does not press down the heel of the strong leg, then they are not trying to elevate the weak leg.
Abductor Sign	Assess for non-organic leg weakness	**1.** Have pt lay on his/her back w/ legs extended. **2.** Place your hand on the lateral sides of his/her thighs. **3.** Ask pt to push their strong leg out (i.e. ABduct his/her strong leg) while you push inwards against BOTH legs w/ equal force. **4.** Repeat above but ask pt to ABduct his/her "weak" leg.	•Step 3 - normal ► weak leg is pushed inwards while strong leg ABducts. •Step 4 – normal ► weak leg is pushed inwards but strong leg remains in place •Suspect functional when strong leg is pushed inwards while assessing ABduction of the "weak" leg.
Sternocleidomastoid Weakness	Assess for non-organic weakness	•Test SCM strength as usual. •Remember that SCM turns the head towards the *contralateral* side of the body	•If weakness is present, it should be present when the head is turned towards the STRONG side of the body.
Tongue Protrusion	Assess for non-organic weakness	•Ask pt to stick tongue out. •Supranuclear innervations of hypoglossal nucleus is bilateral and symmetrical	•In organic weakness, tongue should protrude to the *SAME* side as limb weakness (excluding brainstem lesions)
Giveway Weakness	Assess for non-organic weakness	Test strength as you normally would. Put pt's limb in position to be tested, and ask him/her to give you all of their effort in a single push on the count of three.	•Limb falls rapidly in functional weakness, sometimes before pushed. •Can occur in true disease, especially w/ pain/neglect.
Astasia Abasia	Functional gait disturbance	**1.** Ask pt to walk w/ forced uneconomic posture (w/ knees & hips flexed, in a duck-like posture, or backward) **2.** Chair test – sit pt in rolling chair and ask him/her to pedal the chair forward	•If gait improves w/ either of these maneuvers, suspect a functional gait disorder
Dragging, Monoplegic Gait	Functional gait disturbance	Whole leg is dragged as a single unit behind the pt w/ hip either externally or internally rotated and ankle either inverted or everted	•Hip circumduction found in pyramidal hemiparesis is absent •Classic functional gait
Midline Splitting	Functional hemisensory loss	**1.** Test temp or pinprick sensation by marching laterally across chest/face from abnormal to normal sensation, assessing where perception changes **2.** Test vibration across the forehead or sternum	•Intercostal nerves overlap 1-2 cm so sensory loss from organic lesion is usually paramedian (MANY ORGANIC EXCEPTIONS) •Vibration felt at joints so should not be sharp change in the middle of a bone

SUBSECTION 5

Conclusions

The key to a successful approach to diagnosis of patients with functional neurological symptoms is to concentrate on signs which positively discriminate between functional and organic neurological disorders. It is very important to avoid making diagnoses based on the presence or absence of psychopathology, or simply because the symptoms manifested by the patient seem odd or bizarre. It is important also to remember that functional symptoms are sometimes overlaid on top of other neurological (or general medical) disorders and therefore there may be a dual diagnosis. Positive physical signs of functional symptoms, especially motor symptoms, can be a good starting point for explaining how the diagnosis has been reached. A successful diagnostic explanation is the most important step in achieving a good outcome with treatment.

SUBSECTION 6

Movies of patients with complex conversion syndromes

For better illustration a few telling movies are shown. All patients were awake and attention was full.

MOVIE 9.6.1 Gait disorder, pseudo-weakness, postural tremor

67 y old former salesman. The movie was done 6 months after hip replacement. An acute stroke-like disease had occurred in the rehab institution. Note the changing patterns of left vs. right side weakness depending on direction of gait and on which side the rail is positioned. The pattern is only in part mimicking a hemiparesis after hemispheric stroke; moreover, an incongruent pattern is seen when supported by a left-sided cane vs. being supported by aids bilateral-

ly. The patient can stand upright for 12 seconds until jerky truncal movements appear. The right arm is outstretched towards the stretcher but is not actually using it for support. This is followed by a truncal retropulsion after 6 and a second time after 12 seconds, again without support through the arm stretched out backwards towards the stretcher. When arms are outstretched in the standing position a postural tremor is seen. When lifting the legs in supine position an asymmetrical postural tremor with myoclonus-like jerks is seen with apparent weakness in the hip flexor muscles. Along with the examination the patient also shows deep breathing as if being exhausted. The patient was referred to a psychosomatic rehab institution. Outcome unknown.

MOVIE 9.6.2 Paraparesis – referral diagnosis chronic demyelination inflammatory neuropathy with acute onset

22 y female student, referred for escalating immunotherapy with a diagnosis of inflammatory polyneuropathy. The movie was taken on

admission 12 months later. The patient communicated in a set-back, emotionless way, not revealing any details about potentially stressing life events linked to disease onset. No family members were available for taking the past history. On examination no muscle atrophy, no sensory changes and no reflex loss was found. On nerve conduction studies and EMG no abnormalities were identified. Clues for making the diagnosis of a functional disorder are the following *(note that some sections of the movie are shown twice to better illustrate the core incongruences on examination)*: 1. she walks slowly with a walker with variably strong support from her arms; 2. she is able to walk backwards with the walker, incompatible with severe leg weakness; 3. while sitting she does not lift either leg when asked to and has symmetrical 1 of 4 grade patellar reflex responses. When getting up from the chair she stands on her legs for just one moment before grasping the handles of the walker. The patient denied follow-up. Outcome unknown.

MOVIE 9.6.3 Tremor, gait disorder, bizarre stomping movements

74 y old with hyperkinesias starting at age 62 y while in a rehab institution after hip replacement. The patient shows a left-more-than-right gross tremor in arms and feet, with stomping on left while sitting. After rising up, a bizarre stomping is seen on left. When asked to lie down on the stretcher (a distracting action) the tremor stops almost completely while the patient walks with a pseudo-hemiparetic left arm posture. Once in the supine position the bizarre tremor-like stomping movements restart but only in the left leg. It appears that the patient suffers from pain when stomping down at the edge of the stretcher. The patient denied follow-up. Outcome unknown.

MOVIE 9.6.4 Fixed abnormal positioning of the left arm with stiffness, pseudo-ballistic movements in the same arm

26 y old male student with disabling arm movements for 8 months. He did not report stressful life events linked to disease onset. Initially the left arm is fixed to the trunk and cannot be moved by the

PT. Suddenly a pseudo-ballistic movement occurs with the left arm outstretched and again being fixed in a stiff position. When lying down the arm snaps back into the initial fixed position. While the PT tried to lift the left arm up again, the patient resisted the movement but then suddenly moves the arm up again into a fixed and stiff position but now changing to a flexion-extension pattern. When kneeling down on the stretcher, first only the right arm is used for support when bending the trunk, followed suddenly by a rapid extension of the left arm now supporting the trunk symmetrically. On playing with a ball the patient is able to catch it with the right arm while the left is first fixed over the trunk but then is suddenly extended. Finally, he briefly grasps the ball with left hand before the left arm is falling back into a fixed position. An inquiry at another university medical center revealed that upon neurological examination his symptoms and signs had been completely different from those shown in this movie. The patient denied follow-up. Outcome unknown.

MOVIE 9.6.5 Bizarre staggering gait, increased sway and throwing trunk movements

78 y old lady who lost her husband and lives in social deprivation with broken family contacts. Two different ways of distraction abolish most of the pathology, firstly by shifting attention to a coordination test and secondly by having her walk faster. Note that support is always nearby and available for safety reasons. Good outcome with organized family contacts and physical therapy.

MOVIE 9.6.6 Pseudo-myoclonic jerks, pseudo-epilepsy, tremor, opsthotonus (Charcot's trias)

29 y old female student, 6 years before this movie was done, a diagnosis of "unusual epilepsy" was made. Note that the pattern and frequency of the tremor intermixed with pseudo-myoclonic jerks changes fundamentally during the examination. On the stretcher an opisthotonus is seen and myoclonic jerks in all four limbs are the dominant sign. When turned to a supine position opisthotonus is maintained but only the arms now show a rapid tremor. When lifted up to a sitting position the tremor becomes slower and jerky including the trunk. Upon standing with support, the pattern changes again with an abduction-adduction type tremor in the legs followed by yet another type of alternating tremor-like flexion-extension movement. When asked about the history of her symptoms as a distracting maneuver the shaking is much diminished for a few seconds until again a fast arm tremor is restarting while the legs are no longer shaking for a while. During the walking tests she responds coherently to the doctors' questions. When asked to stand on one

leg, almost no tremor is seen in the legs while there is profound postural tremor in both arms. No signs or history of falls, tongue bites; normal prolactin levels, no EEG abnormality. The patient denied follow-up. Outcome unknown.

MOVIE 9.6.7 Caveat: incorrect diagnosis of a conversion disorder

Waddling walking pattern, with mild "spinal" ataxia, pseudo-dystonic hand position, pseudo-tabes; on sensory testing complete loss of position sense up to the proximal joints.

54 y old male salesman with waddling walking pattern, with mild "spinal" ataxia, pseudo-dystonic hand position, pseudo-tabes; on sensory testing complete loss of position sense up to the proximal joints suggesting paraneoplastic ganglionitis (pseudo-tabes) due to small lung cell cancer. Despite a working diagnosis of a functional disorder a complete neurological examination revealed de-afferentiation with complete loss of position sense. With eyes closed

the patient was unable to stand and walk (not shown). No evidence of a conversion disorder.

SECTION 10 – SUMMARIZING, CATEGORIZING, AND STRUCTURING PATHOLOGIC FINDINGS

Deducing possible Etiologies, Pathogeneses, Differential Diagnoses; and considering appropriate ancillary Tests.

All abnormal findings are collected (problem list)

The pathology is grouped in syndromic terms by anatomical or functional localization
e.g. symmetric flaccid weakness of the lower limbs; **spastic** hemiparesis; paraplegic **spastic gait** disorder

Try to define a unifying lesion or disease that allows including most of the pathology and make a tentative diagnosis

Consider other possibilities in the differential diagnoses

What is the most likely diagnosis?

What will endanger or even kill the patient if missed?

What can be treated?

What really bizarre/interesting thing can do this?

- Mnemonics such as VITAMIN C&D (V – vascular, I - infectious/inflammatory, T – trauma, A – autoimmune, M - meta-

bolic/toxic, I - inherited/iatrogenic, N – neoplastic, C – congenital, & - other/unknown/idiopathic, D - degenerative)

Discuss possible etiologies based on findings and history
e.g. a patient with a likely major ischemic hemispheric stroke and a history of myocardial infarctions, untreated diabetes mellitus type II, and smoking habit; a patient with fluctuating and progressive muscle weakness suggestive of myasthenia gravis.

Plan appropriate tests based on clinical considerations

- **Lab analyses** from serum, and in some instances from cerebrospinal fluid (CSF), are done on a routine basis to which special tests are added (e.g. specific autoantibodies, serology for infectious agents, paraneoplastic biomarkers, etc.). In some diseases genetic testing is indicated, e.g. in myopathies, some neuropathies, storage diseases, PRION diseases. It is important to differentiate the lab work in urgently needed tests (e.g. electrolytes, markers for inflammatory disease) and those that need more time (e.g. genetics, PRIONs) or are less urgent (e.g. autoantibodies).
- **Neuroimaging** is now a standard test procedure: e.g. a patient with a suspected stroke needs an emergency cranial CT scan or MRI, the patient with acute spastic paraparesis needs urgent MR imaging of the spinal cord and brainstem, the patient with a slowly progressive spastic tetraplegia needs less urgent testing.

Clinical neurophysiology

Nerve and muscle **electrophysiology** can help as well. Special scheduled tests need a clear indication, e.g. work-up of the patient with fluctuating weakness includes repetitive nerve stimulation; in the patient with progressive neuropathy nerve conduction studies (NCS) and electromyography (EMG) need to be done.

Evoked potential studies can help in finding long tract conduction abnormalities in evaluation of multiple sclerosis, myelopa-

thy, coma, and others.

Electroencephalography (EEG) is also extremely helpful in the evaluation of epilepsy and of value in certain neurological problems, particularly in the investigation of altered mental status (e.g Creutzfeld-Jakob's disease).

Tissue pathology

Some neurological diseases require a tissue diagnosis on which appropriate treatments need to be based. These include muscle biopsies in most muscle disorders (e.g. dystrophies, metabolic myopathies, inflammatory myopathies, including vasculitis etc.); sural nerve biopsies in some peripheral nerve disorders (e.g. chronic inflammatory neuropathies including vasculitis); temporal artery and/or muscle biopsy in giant cell arteritis; brain biopsy in brain tumors and PRION diseases; intestinal or other tissue biopsies in storage diseases.

SECTION 11 – THE EXAMINATION OF THE UNCONSCIOUS PATIENT

The determination of the cause of unconsciousness

Staging pre-coma and coma and classifying its potential etiology is an essential task in the emergency room and in the neurocritical care unit because the necessary tests will depend on the differential diagnosis. The main point is differentiating coma due to focal brain disease (e.g. massive hemorrhage with midline shift, brain abscess with cerebritis, brainstem injury or compression, structural injury of the hemispheres), from **coma due to global or diffuse brain disease** (e.g. encephalitis, seizures, hypothermia, neuroleptic malignant syndrome, metabolic derangement, etc.). With severe head injury, focal and diffuse damage may coincide, and peripheral nerve or cranial nerve lesions may be superimposed in polytraumatized patients. A well-based clinical impression at the bedside may help to focus on the most needed tests, to avoid any unnecessary (and potentially painful or even risky) movement of the critically ill patient.

Special attention to the presence or absence of **wakefulness** and **awareness** as well as focality of neurological deficits will help in the classification of the patient's condition. **Encephalopathic** (delirious) patients will often have waxing and waning of arousability, but most prominently manifest impairments of attention (e.g. distractability, inability to perform complex tasks) but will not have any focal neurologic impairments. Abulic patients (often due to prefrontal and/or anterior cingulate lesions) may require time to respond, but the neurologic function is otherwise intact. Whereas coma often results from damage to the ascending arousal systems (reticular activating system) which courses from the dorsal midbrain to the bilateral centromedian nuclei of the thalamus and then up to the bilateral frontal cortices.

Two common pitfalls of the comatose patient evaluation include the locked-in syndrome and psychogenic unresponsiveness. The **locked-in** patient (usually from ventral pontine damage) is both awake and aware but has limited ability to interact (often just vertical eye movements of one or both eyes, and blinking). A diagnosis of **psychogenic** unresponsiveness is often a diagnosis of exclusion, but particular attention to key physical exam findings will help to avoid unnecessary and expensive workup.

In the long-term determination of persistent vegetative states (syn. Vegetative State/Unresponsive Wakefulness Syndrome) and minimally conscious states can be made with the aid of advanced diagnostic tests, but discussion of such topics is beyond the scope of this text.

Staging the level of unconsciousness

Using terms to describe the conscious state has resulted in inconsistency and confusion. Toward this end, the level of consciousness is best described in **simple language**, e.g.

"the patient required continual verbal and tactile stimulation to maintain alertness" or "the patient was unarousable to even vigorous painful stimulation". Nonetheless, some general categories of arousal and other associated patterns of abnormal findings are described below.

- **Somnolence**: The patient resembles a sleeping person. The patient can be awakened by calling him up or gently clapping a hand on the cheeks or trunk. While briefly awake, the patient is able to fixate the examiner and to perform simple tasks such as squeezing the examiners hand on either side or close and reopen the eyes on demand. There may be a short sentence. Each test should be repeated at least twice if there is any doubt.
- **Deep somnolence or stupor**: The patient needs repeated and stronger stimuli to briefly wake up and to open the eyes, such as mildly to moderately painful stimuli (pinching the skin). The patient may not follow any command specifically but may groan or grunt and will not speak. Wakefulness does not persist in the absence of continuous stimulation.
- **Coma**: The patient cannot be awakened. The following patterns of features can be encountered:
    - **Light coma**: There is a local withdrawal reaction to painful stimuli. The vestibuloocular reflex (VOR) is still present upon passively turning the head of the patient. The eyes will show a deviation away from the direction of the turning. (**Do not do this test** in patients with possible cervical spine lesions). The pupillary reflex to light is present and at resting position the pupils are rather small (around 2-3 mm). One should take care that other reasons for a

small pupil (cholinergic drugs or local medication) are excluded.

- **Light to moderately deep coma**: Local painful stimuli induce more general movements; the VOR is still present (Do not test in patients with possible cervical spine lesions). The pupil will react to light and may widen upon a painful stimulus (sympathetic reaction).
- **Markedly deep coma**: no reaction to normally very painful stimuli applied bilaterally in sequence (like squeezing the thumb nail with the bar of the reflex hammer; applying firm pressure on the glabella above the nose, or at the septum of the nose); the patient may show "decorticate" posture with both arms going into flexion: the VOR is either absent or at most rudimentary. Pupils are small and the light reaction is weak or may

be absent (a bright light is needed and an ophthalmoscope or a magnifying lens may help to see minor contractions of the pupil). Some tendon reflexes may still be obtained, and the corneal reflex can be elicited. While the VOR is negative, cold water irrigation to the middle ear may lead to a slow deviation of the eyes, often not conjugated. Breathing is abnormal with deeper breaths and higher frequency. Babinski reflexes are often positive.
Note: here meningismus may be weak or absent even in patients with meningeal disease proper (e.g. subarachnoid hemorrhage or meningitis).

- **Deep coma**: no motor reaction to any stimulus or a decerebrate posture is invoked, also asymmetrical posturing is possible ("decorticate" flexion on one side and "decerebrate" stretch-

ing on the other). Corneal and other brain stem reflexes are absent. Pupillary light reaction is absent and pupils are wide and no more round. Usually there is no spontaneous breathing.

Note that meningismus is usually missing even in patients with meningeal disease.

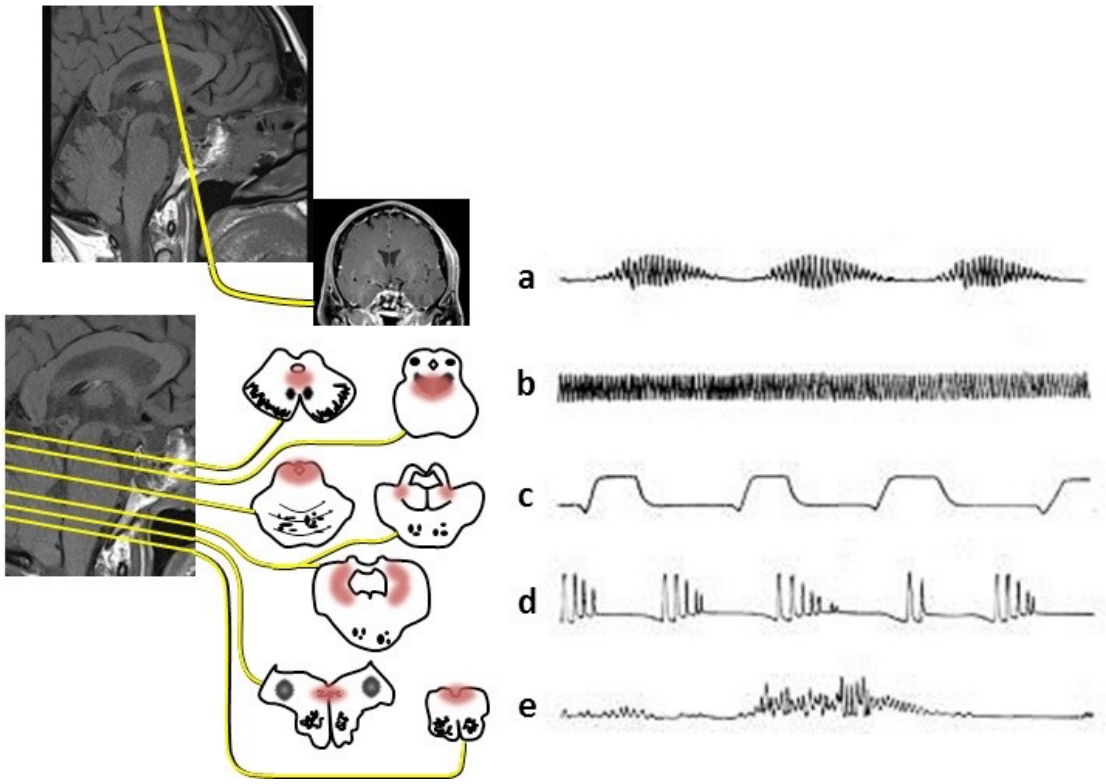
Note: any deviation from these patterns of abnormality suggests a more complicated and even a multicausal lesion, or superimposed metabolic factors. The most telling (and ominous) finding is the loss of brainstem reflexes.

The examination of the unconscious patient includes the following essential points

The very first consideration in the unresponsive patient is ensuring stabilization and management of **C**irculation, **A**irway, and **B**reathing, Not only will these assessments and interventions avoid further decompensation, but their evaluation may also help point to an underlying etiology. Standard laboratory investigations are critical aspects of the initial investigation: arterial blood gas and lactic acid, complete metabolic profile, complete blood count with differential, ammonia level, thyroid stimulating hormone level, urinalysis and culture, blood cultures (if infection suspected), urine and serum volatile toxicologies, bedside glucose assessment, electrocardiogram and cardiac troponins, and chest roentgenogram. A non-contrasted head CT may be of additional help if an intracranial pathology is suspected, as well as a lumbar puncture if a central nervous system infection is a possibility or no other cause for coma can be identified. Eventual MRI and EEG may be of further assistance if the coma persists without an etiology being defined.

Vital signs: not only the steady state of the blood pressure, heart rate, and breathing pattern (often confounded by ventilator), but their response to stimuli should be noted. Don't forget the Cushing triad: hypertension, consequent bradycardia, and irregular breathing pattern. Temperature extremes also may indicate an underlying cause such as infection, alcohol intoxication, etc.

FIGURE 11.11.1 Respiratory patterns based on location of damage (indicated by red shading)



a) Cheyne-Stokes respiratory pattern (oscillation in breathing depth and frequency) in diffuse, bihemispheric damage. b) Central neurogenic hyperventilation. c) Apneustic breathing. d) Cluster breathing. e) Ataxic breathing (irregular)

Meningismus: Check in the neck region and in the lumbar region by passively bending the head and the legs.

Speech: If there is any verbal expression check for aphasia, and any meaningful content.

Motor activity: Observe and document spontaneous motor activity (symmetrical or asymmetrical), myoclonus, spontaneous decerebrate (Adduction and internal rotation of the shoulder, arm extension, and wrist pronation and fisting) or decorticate (slow flexion of the elbow, wrist, and fingers) posturing. Assess response to painful stimuli by performing central (supraorbital nerve, temporomandibular joint, trapezius, and sternum) stimulation. Discriminating between a withdrawal and flexion responses (such as the spinally-mediated triple flexion in the LE) is helped through proximal (arm or thigh pinch) and distal (nailbed pressure with a blunt object) noxious stimulation.

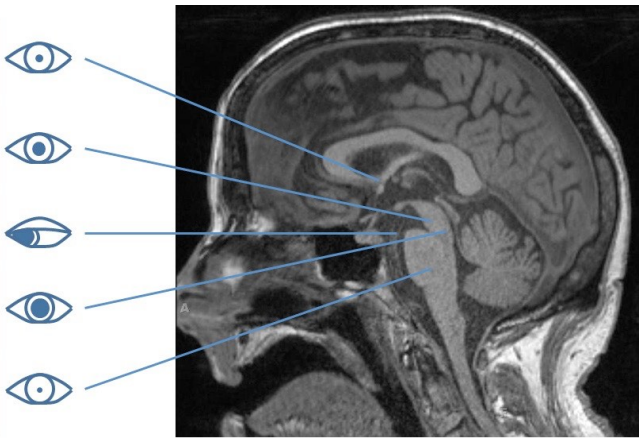
Response to threat and visual responsiveness: To determine if the patient is processing visual stimuli in any visual field. See if the patient will track objects: a monetary note (e.g. $20) or a mirror provides the best assessment of tracking.

Fundoscopy: Papilledema from increased intracranial pressure (ICP) or subhyaloid haemorrhage following aneurysmal subarachnoid haemorrhage can often be found.

Pupils: Take a measure of the size in millimetres on either side, check for direct and consensual light reaction; observe the pupil's size and shape upon painful stimuli (see section 3). Pupillary dilation along with hypertension may be the only signs of seizure in a patient following intubation with paralytic agents (as the vasculature and pupillary sphincter muscles have muscarinic receptors).

FIGURE 11.11.2 Pupillary abnormalities based on location of damage.

Diencephalon	Small <2 mm	Reactive
Midbrain	Mid-position	Fixed
CN III	Dilated >8 mm	Fixed Ptotic, "down and out"
Tectum	Dilated >8 mm	Fixed
Pons	Pinpoint <2 mm	Minimal to no reaction

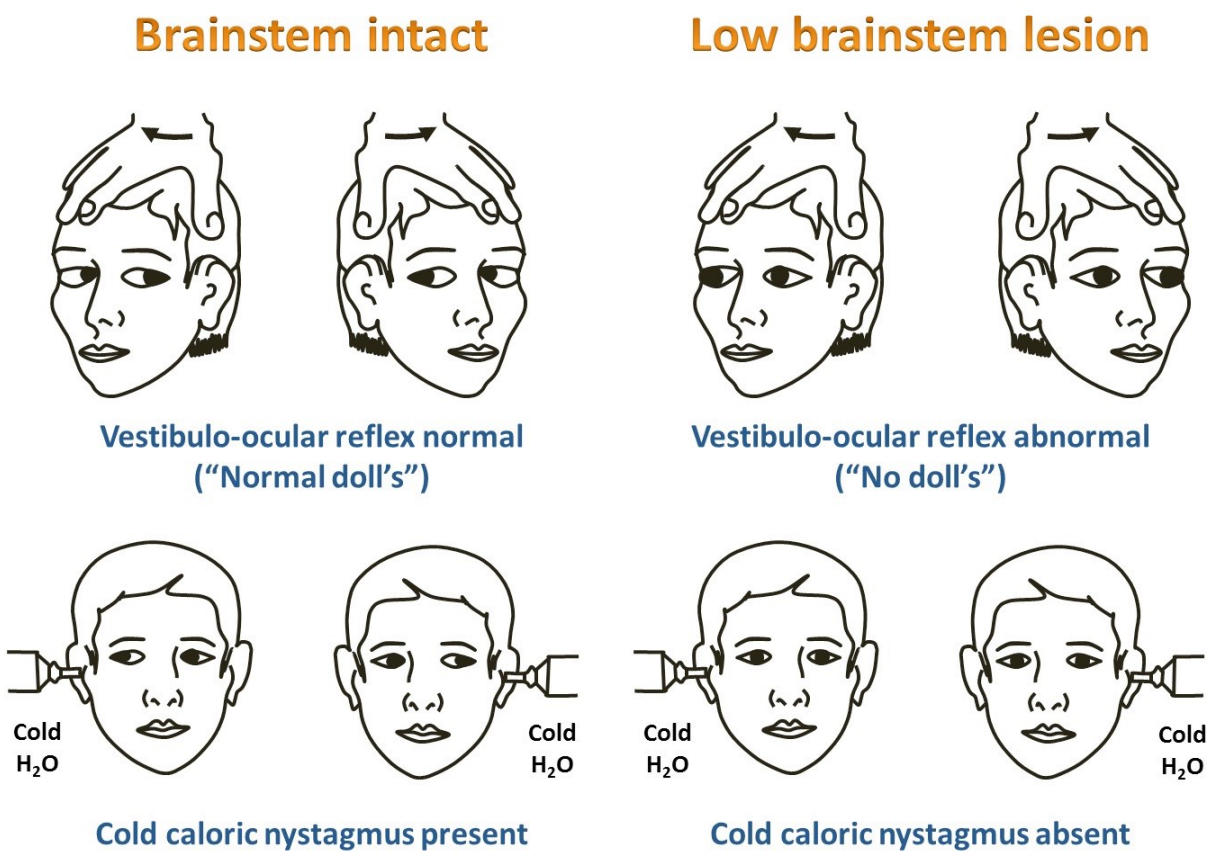


Note that patients with metabolic/toxic/infectious etiologies to their altered mental status will often have small, but reactive pupils.

Eye position: Is it conjugated (suggesting seizure if forced and unable to be **overcome** with VOR/doll's maneuver or unilateral frontal eye field pathway damage if VOR/doll's maneuver remains intact) or is there a discordant deviation vertically or horizontally (suggesting brainstem pathology or possibly a baseline exo/eso-tropia)? (see section 3). Are there spontaneous, slow movements (roving eye movements, ping-pong-like lateral eye movements), which are typical in bihemispheric dysfunction, but suggest an intact brainstem? Are the eyes **bobbing** (rapid downgaze, with slow upward drift) to suggest a pontine lesion? Very mild nystagmoid (jerky) eye movements can be appreciated in subtle status following a seizure.

Vestibulo-ocular reflex (VOR) and oculocephalic reflex (doll's eyes): associated with the cold caloric testing to look for **nystagmus**. In moderately deep coma there will only be a deviation, no **nystagmus**. In deep coma there is no reaction at all. In the "doll's eye" test, with an intact brainstem the eyes should loll in a direction that is opposite the rotation of the head (see section 2).

FIGURE 11.11.3 Vestibulo-ocular reflex / doll's eye reflex



Demonstration of the VOR/doll's reflex and cold calorics in patients with intact brainstem function vs those with a low brainstem lesion.

Cold calorics: Performed with a 1-minute instillation of ice-cold water (< 30°C, or < 86°F) while looking for a slow **drift toward** the cold ear and a **saccade away** from the cold ear. Testing on the contralateral ear is performed after a 7-minute temperature normalization period. The mnemonic "jerking COWS" (cold-opposite, warm-same) can help to remember which direction the fast phase is supposed to go.

Corneal reflex: (see section 2): it will be lost on both sides in deep coma. If lost only on one side it indicates focal ipsilateral brainstem pathology on the ipsilateral side (e.g. in brainstem stroke).

Gag reflex (see section 2) **and coughing reflex**: these are best observed during suction of the saliva from the throat or with a cotton-tipped applicator placed in the oropharynx. The cough reflex is also assessed during deep suctioning at the carina from the endotracheal tube. It is absent in markedly deep coma.

Muscle tone: flaccid or spastic, rigid, Gegenhalten possible only in light coma, symmetric or asymmetric tone may point to a focal lesion of the upper motor neuron.

Reflexes: focal alterations, asymmetry. The presence or absence of Babinski's sign, unilaterally or bilaterally, will help to identify focal pathology.

At the end of the examination a summarizing syndromic statement is needed:

- **Without known trauma**
  Coma without focal pathology often points to a diffuse CNS disease, e.g. intoxication, metabolic derangements, non-focal inflammatory disorders;
  Coma with focal or one-sided pathology often points to major hemorrhage, focal inflammation, advanced brain tumours.
  Coma with meningism points to a disease involving the meninges, e.g. subarachnoid hemorrhages, bacterial meningitis. Note that meningism may disappear in deep coma.
- **With known head trauma**
  The Glasgow Coma Scale has been developed as a simple and quick way to assess the severity of brain trauma (though there are many other scales such as the FOUR Score: http://www.ncbi.nlm.nih.gov/pubmed/16178024). It can also be used for other brain disorders causing coma as long as there are no discrepancies between the three major test items, e.g. in intoxications with narcotics.

TABLE 11.11.1 FOUR Score and Glasgow Coma Scale

FOUR Score	Glasgow Coma Scale
Eye response	**Eye response**
4 = eyelids open or opened, tracking, or blinking to command **3** = eyelids open but not tracking **2** = eyelids closed but open to loud voice **1** = eyelids closed but open to pain **0** = eyelids remain closed with pain	**4** = eyes open spontaneously **3** = eye opening to verbal command **2** = eye opening to pain **1** = no eye opening
Motor response	**Motor response**
4 = thumbs-up, fist, or peace sign **3** = localizing to pain **2** = flexion response to pain **1** = extension response to pain **0** = no response to pain or generalized myoclonus status	**6** = obeys commands **5** = localizing pain **4** = withdrawal from pain **3** = flexion response to pain **2** = extension response to pain **1** = no motor response
Brainstem reflexes	**Verbal response**
4 = pupil and corneal reflexes present **3** = one pupil wide and fixed **2** = pupil or corneal reflexes absent **1** = pupil and corneal reflexes absent **0** = absent pupil, corneal, and cough reflex	**5** = oriented **4** = confused **3** = inappropriate words **2** = incomprehensible sounds **1** = no verbal response
Respiration	
4 = not intubated, regular breathing pattern **3** = not intubated, Cheyne-Stokes breathing pattern **2** = not intubated, irregular breathing **1** = breathes above ventilator rate **0** = breathes at ventilator rate or apnea	

Comparison of the FOUR Score (Wijdicks et al, 2005) to the Glasgow Coma Scale

Note: the comatose patients with a flexion response to pain will bend both arms except when there is an additional peripheral lesion, as with brachial plexus trauma in a patient with multiple injuries; in coma with an extension response the patient usually will extend both arms and pronate (invert) both hands except when there is an additional peripheral lesion (see Section 2).

Categorization: Coma: No eye opening, no ability to follow commands, no word verbalizations (Scale value 3-8).

Head Injury Classification:

Severe Head Injury	GCS score of 8 or less
Moderate Head Injury	GCS score of 9 to 12
Mild Head Injury	GCS score of 13 to 15

(Adapted from: Advanced Trauma Life Support: Course for Physicians, American College of Surgeons, 1993).

The Glasgow Coma Scale (GCS) does not allow a clearly operationalized distinction between the syndrome of unresponsive wakefulness and the minimally conscious state. For this purpose, the revised Coma Recovery Scale (CRS-R) has been established internationally. It is mainly used for chronically persistent disturbances of consciousness and follow-up examinations. The application of this scale is considerably more time-consuming than the Glasgow Coma Scale or FOUR Score which have been developed for use at the scene of an accident, in the emergency room or intensive care unit, and requires relevant experience. The CRS-R allows not only an assessment of the course of the disease using an ordinal scale from zero (deepest coma) to a maximum of 23 points (awake and fully conscious), but also a differentiation between the syndrome of non-responsive wakefulness and the minimally conscious state. The patient's reactions are graduated in the 6 different domains according to the following functional scales:

Auditory function scale point designations

- 4 - Consistent Movement to Command
- 3 - Reproducible Movement to Command
- 2 - Localization to Sound
- 1 - Auditory Startle

- 0 - None

Visual function scale point designations

- 5 - Object Recognition
- 4 - Object Localization: Reaching
- 3 - Visual Pursuit
- 2 – Fixation
- 1 - Visual Startle
- 0 - None

Motor function scale point designations

- 6 - Functional Object Use
- 5 - Automatic Motor Response
- 4 - Object Manipulation
- 3 - Localization to Noxious Stimulation
- 2 - Flexion Withdrawal
- 1 - Abnormal Posturing
- 0 - None/Flaccid

Oral movement/speech function scale point designations

- 3 - Intelligible Verbalization
- 2 - Vocalization/Oral Movement
- 1 - Oral Reflexive Movement

- 0 - None

Communication scale point designations

- 2 - Functional: Accurate
- 1 - Non-Functional: Intentional
- 0 - None

Arousal scale point designations

- 3 – Attention
- 2 - Eye Opening w/o Stimulation
- 1 - Eye Opening with Stimulation
- 0 - Unarousable

Note: Coma recovery scale – revised – download scoring guidelines at: https://www.tbims.org/combi/crs/CRS%20Syllabus.pdf

SECTION 12 – THE PSYCHIATRIC EXAMINATION IN THE EMERGENCY ROOM

General Note: Section 12 is meant to be an aid for non-psychiatry trained physicians once confronted with psychiatric patients. Patients with acute psychiatric manifestation may be directed to various hospital departments depending on the local medical system and the availability of resources. In many countries, on call emergency medics or emergency physicians decide about where the patient is being admitted: a general emergency room, a neurology-neurosurgery ER, or a psychiatry ER. Only upon careful history taking and examining the patient, it may turn out what type of underlying disorder is likely causing the psychiatric symptoms. This may then allow transferring the patient to the appropriate medical specialty.

General Terminology

Anhedonia

Avolition

Cluster B personality disorder

Delusion

Echolalia

Echopraxia

Flight of ideas

Hallucination

Legal highs

Mutism

Rumination

OTC

Stupor

Waxy flexibility

A first impression of a patient's mental state is usually established during history taking and when doing the standard neurological exam. Much of what one needs to know at this stage is brought up by observation and simple **colloquial-style** questions, e.g. "What seems to be the problem?" "Did anybody accompany you?", "How can I help?", or others as outlined in this section. (cf. Movie 8.8.1)

If next of kin are in the examining room they may not wish to disclose relevant information in front of the patient. In this case, it is important to take a focused history without the patient present. If the patient opposes his/her accompanying person to leave the examining room, they may search for an excuse, and, for instance, claim a need to use the bathroom. This may provide a first **hint** about the psychiatric problem of the patient. Psychiatric disorders manifest themselves in various ways. These are illustrated below and, in Section 9 (conversion disorders). In principle, comorbidities of various disorders are a common feature with advanced age and this warrants proper consideration. Common examples are delirium within the realm of severe medical diseases, polypharmacy, or chronic substance abuse.

For illustration of how a real patient may present in the emergency room, links are provided (YouTube) to actually see patient interviews. In addition, a few case histories are presented at the end of this section.

Note: Appreciation of psychiatric conditions may have important implications for the capacity to provide informed consent. Capacity assessments can be performed by any physician who is familiar with symptoms and signs: the key aspect is that of proper documentation. Capacity can vary from decision to decision, and must be clarified as to the patient's current state of mind, his/her understanding of recommended treatment options, what the risks and benefits of alternative procedures are, or the objection of them. Most importantly, it needs to be clarified that he/she does understand all of these factors and has demonstrated the ability to provide a logical rationale for his/her decision. If any one of these points is not fulfilled further measures and treatments may be limited unless there is a legal power of attorney or there is an emergency, like severe self-harming behavior, including suicide attempts or harm to others, as in the case of severe psychomotor agitation or violent behavior.

A psychiatric emergency examination starts as any other exam with first assessing the patient's complaints, signs and symptoms, and asking for information about the patient's medical and psychiatric history. Then follows the psychiatric mental status examination (see below). Even when in an emergency room setting, it is essential to perform this psychiatric examination if the patient presents with psychological complaints or evidently shows psychiatric abnormalities. Only then, the need for transferring the patient to a psychiatric unit can be appreciated. However, some psychiatric syndromes, like a highly agitated patient, lack of cooperation, or potential violence toward others, do not allow for an in-depth examination but rather, first, protective measures are needed.

Note: Psychiatric patients are more stigmatized than other groups of patients - and health professionals are not excluded from a biased view -, so remember to be non-judgmental, objective, and non-condescending.

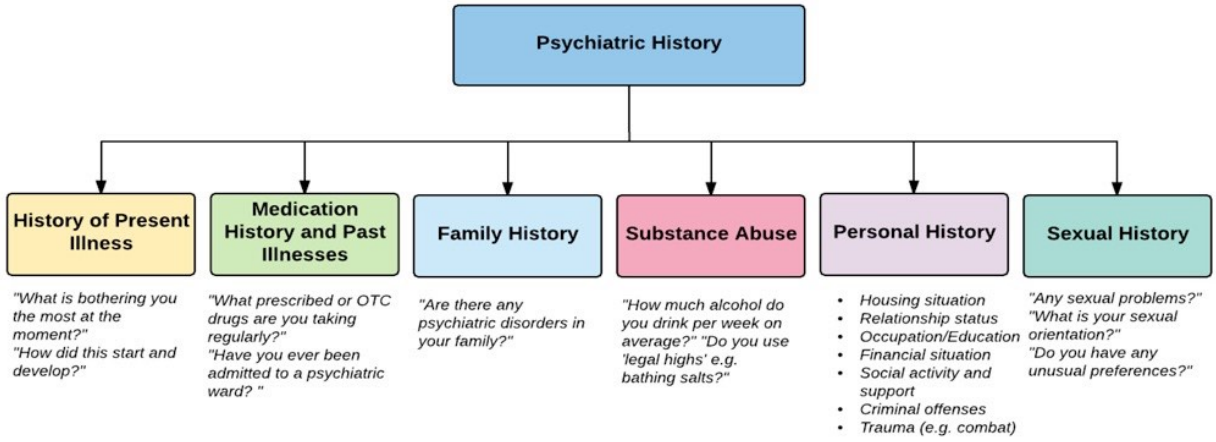
SUBSECTION 1

Psychiatric History

In the following paragraphs, we give an overview of how to approach and examine a psychiatric patient in the emergency room.

Note: Often a full history cannot be taken in the General Emergency room but has to be taken later by a psychiatry consultant. The order of exploration may vary depending on the specific symptoms the patient presents.

FIGURE 12.1.1 Psychiatric History



Summary of the most important categories of psychiatric history taking and examples of specific questions to ask the patient.

SUBSECTION 2

Mental Status Examination

The Mental Status Examination in psychiatry is about the **examiner's observations and impressions** of a patient at the time of the interview. It is not only about **what** the patient says but also **how** he/she says it and how **plausible** it seems to the examiner. This definition **differs** from the term as used in the Neurological Examination.

Whenever the patient's view differs from the doctor's observation, you may mark it as "subjective" (e.g., "attention subjectively impaired", i.e. the patient feels this way while it seems "objectively intact" to the examining physician).

The order of the exam and the various items to be addressed area listed below and its completeness should vary according to the patient's most pressing complaints and signs. Even when not immediately obvious, **always ask about suicidal ideation and impulses**.

TABLE 12.1 **Mental State Examination**

Appearance and Behavior

Inappropriate or filthy clothing

Insufficient personal hygiene

Agitation

Patient can't sit still

Subdued, motor retardation

Flirtatious, ecstatic, laughing

"Has anyone commented on how you look/smell?"

Speech

Talkative

Reduced drive to speak

Speech too rapid or slow

Dysprosody

Foreign accent despite being a native speaker

Alertness, Orientation, Attention, Memory, Judgement, Abstraction

See section 8, subsections 1-3 and 8

Formal Thought

Loose or inappropriate associations

Flight of ideas

Neologism

Perseveration

Rapid/slow thought

Prolonged response latency

Rumination

"Do you think about mistakes of the past over and over again?"

"Do you feel your thoughts are racing?"

Thought Content

Delusions

Ideas of reference, ideas of influence

"Do you feel followed by anyone?"

"Does the TV send secret messages to you?"

"Is anyone controlling your behavior/ movements/thoughts?"

"Am I able to read your mind? Are you able to read mine?"

Phobias, Obsessions, Compulsions

Very tidy patient

May disinfect hands during interview

"Are you afraid of anything in particular?"

"Do you feel you must count/control things/wash your hands or otherwise something bad will happen?"

"Are you afraid of being in a crowd or in an elevator?"

Perception

Auditory, visual, or olfactory **hallucinations** indicated by the patient turning his/her head in a certain direction several times as if seeing or hearing something

"Is there anyone else in this room except you and me?"

"Do you hear someone else's voice except mine?"

Mood and Affect

Looks depressed

Laughs a lot/cries during interview

Mood swings

Anxious

Emotional responsiveness impaired

Affect inappropriate

"How do you currently feel?"

"On a scale of 1 to 10, how anxious/depressed/elated do you feel?"

"Do you feel you have lost emotional contact to your loved ones?"

Volition, Level of Energy

"Are you able to easily get up in the morning?"

"Are you easily able to do your everyday chores?"

"Does your job seem too demanding lately?"

Psychomotor Activity

Agitation

Restlessness

Hyperactivity

Twitching

Slowing of body movements

Tics

Stupor

Sleep, Appetite, Libido

"How many hours of sleep per night did you get last week on average?"

"Have you lost your appetite/ lost weight?"

"Has your interest in sex recently changed?"

Self-mutilation and Suicidal Ideation

Incised wounds around wrist/ forearm (fresh/old/scarred/deep/superficial)

Picked skin

"Do you cut yourself?"

Also see subsection 12.3

Insight

- "Why do you think you are in the emergency room?"
- "Do you believe you have a psychiatric disorder or a psychological problem?" Cf. Section 8

SUBSECTION 3

Potentially Suicidal Patients

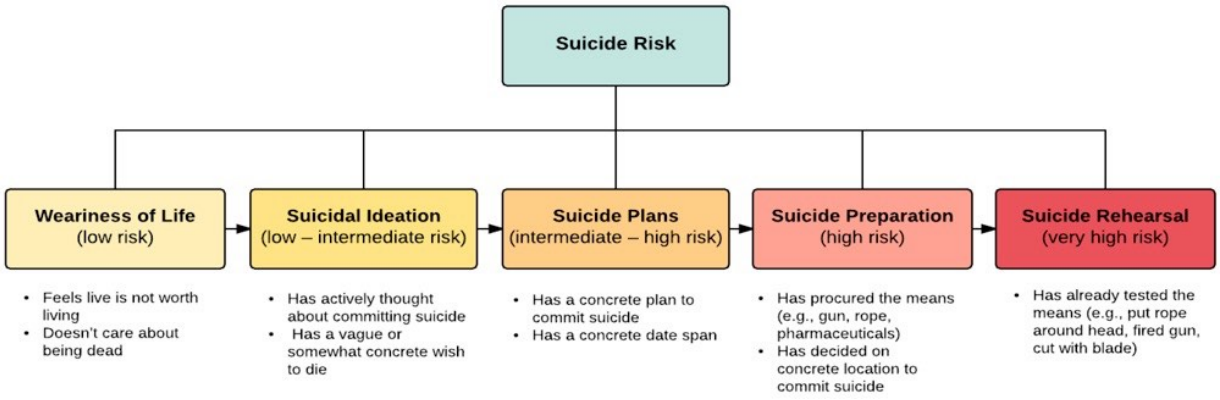
If you get the impression that a patient might be suicidal, you need to check for the following risk factors:

TABLE 12.2 **Risk Factors for Suicide**

Previous suicide attempt/s (strongest predictor!)
Family history of suicide
Comorbid psychiatric disorders
Hallucinations
Substance abuse/dependency
Low social support
Male sex (suicide ratio 3:1 males vs. females)
Expressing feelings of hopelessness
Lack of insight in his/her illness

Note: Asking about suicidal ideation does not increase the risk for suicide although this may sound counter-intuitive! Some suicidal patients at very high risk may be misjudged as non-suicidal as they may feel relieved and appear calm or even joyful because they believe that they have found the perfect "way out". In a first encounter just ask as much as is needed for a good judgement as to immediate psychiatric consultation.

FIGURE 12.3.1 Assessment of Suicide Risk



SUBSECTION 4

Agitation

When confronted with an agitated patient presenting with pacing, shouting, psychomotor arousal, high irritability, threatening gestures, and aggressiveness, talk him/her down by introducing yourself as a "helper" in a **calm and friendly** tone. If you suspect potential violent behavior, conduct the examination in company of a nurse or security staff. Leave **enough space** between the patient and yourself, **do not stare** at him/her, do not stand over him/her, and **avoid physical contact** so that the patient does not feel threatened. For any necessary diagnostic work-up or for starting treatments, it is legally correct to restrain a patient on a bed temporarily with the help of at least four experienced staff members if needed.

Note: Dependent upon the federal or state legislation, sometimes physicians and medics need to install preventive measures including calling the police for help if needed, even against the patient's will.

Many patients who present with psychiatric symptoms in the emergency room have an underlying substance-use or medical etiology, especially when patients present with psychiatric symptoms for the first time and are over the age of 40 (Testa et al., 2013). Acute onset of agitation and fluctuation of symptoms with symptom-free intervals of hours to days are indicators of a substance-use or medical etiology.

When patients present with agitated behavior in the emergency room, **ethanol**, any other legal or illicit **drug intoxication** or **withdrawal** are the most common causes; further psychiatric diagnoses to be considered in addition to the above **include (hypo)mania, schizophrenia, and** **cluster B personality disorders**.

Acute onset of **agitation** and/or severely psychotic symptoms in a patient without any psychiatric history or complaints so far, may also indicate acute-onset autoimmune encephalitis caused by

autoantibodies directed at NMDA (or other) receptors or at other brain target molecules and must be worked-up accordingly (Graus et al., 2016). Often these patients also suffer from epileptic seizures.

Video of an agitated patient:

https://www.youtube.com/watch?v= 7cZDfWcIitg

SUBSECTION 5

Delirium

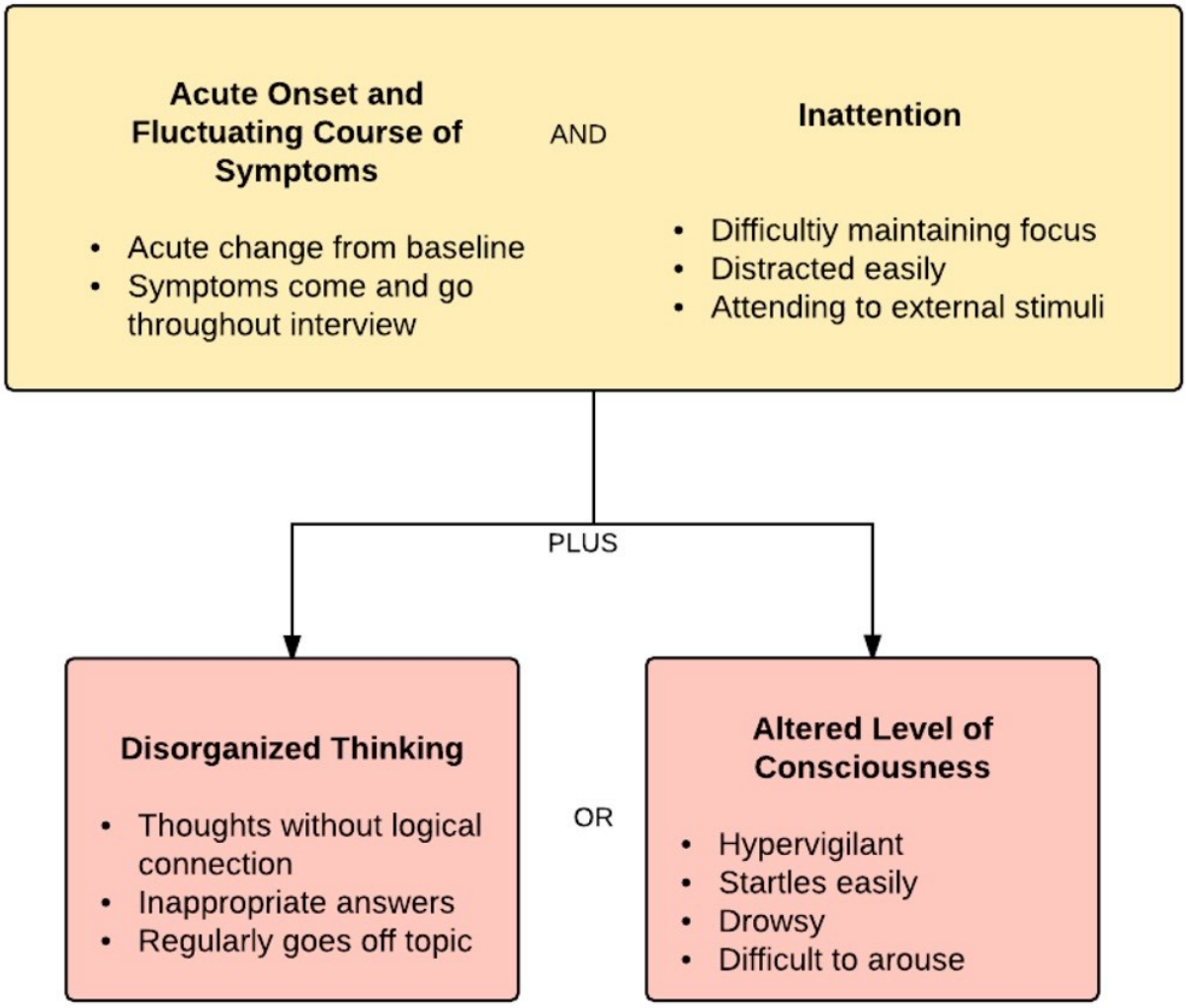
In the **elderly**, the most common cause of agitation is delirium due to a **medical condition** such as metabolic or electrolyte derangement as seen with systemic infections, dehydration, hypoxia, head trauma, drug intoxication/withdrawal, polypharmacy, and any type of encephalitis including septic forms.

There are two **different types** of delirium: (1) **hyperactive** (agitated) and the (2) **hypoactive** (lethargic). Especially when elderly patients seem "odd", delirium needs to be considered. Many elderly patients experience a delirium immediately prior to or during a hospital stay (Hein et al., 2014). This type of delirium has a poor prognosis if untreated. Here, a thorough listing and evaluation of all drugs currently taken by the patient is essential.

Note: Especially in the elderly, almost any drug can trigger delirium, often combined with metabolic derangement due to dehydration.

The best-established screening instrument for delirium is the **Confusion Assessment Method** (CAM) with a sensitivity of 94 % - 100 % and a specificity of 90 % - 95 % (Wong, Holroyd-Leduc, Simel, & Straus, 2010).

FIGURE 12.5.1 Diagnosis of Delirium using the CAM



Videos of Delirium in elderly patients:
https://www.youtube.com/watch?v= lJH1AoVuVS0
https://www.youtube.com/watch?v= 2mCfFmNmJGY&list= PLLbB-ZOIyRri2hHWX5xg2i_mUB7qIa9D8L&index= 3
https://www.youtube.com/watch?v= 9QURzexhWP4

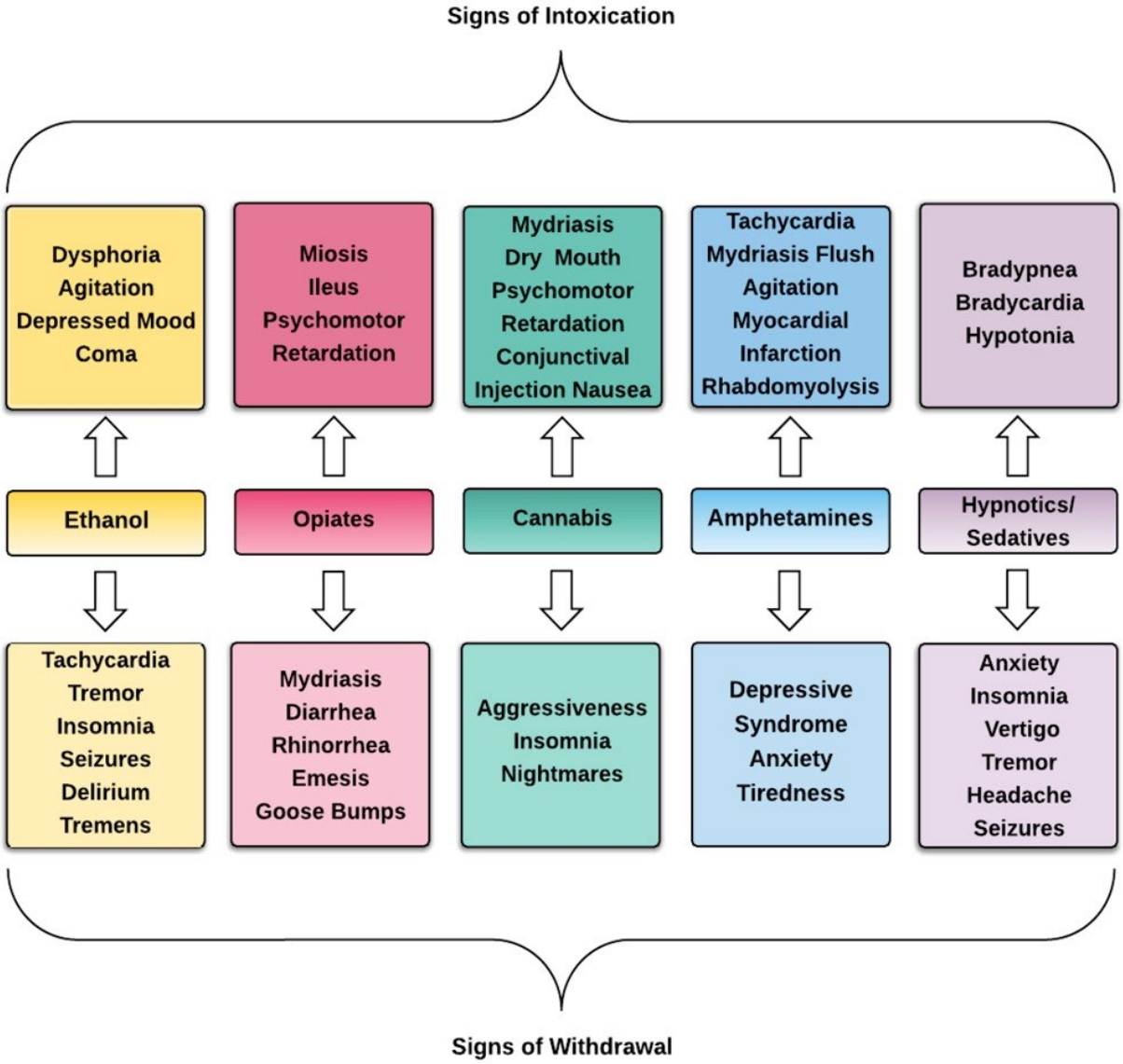
SUBSECTION 6

Intoxication and substance withdrawal

One of the most common causes of agitation is intoxication. It is defined as a transient state affecting alertness, cognition, mood, perception, behavior, and the autonomic nervous system, due to the consumption of one or more substances.

Note: Substance (drug) withdrawal and intoxication can present with similar behavioral symptoms, but abnormal signs related to the autonomic nervous system may differ. Withdrawal symptoms occur with dose reduction of a substance the person is used to, whereas intoxication may occur after first-time use and in dependent persons alike.

FIGURE 12.6.1 Signs of Intoxication and Withdrawal with Regard to Specific Drugs*



**If intoxication is in the differential diagnosis, emergency laboratory screening tests are mandatory. If drug withdrawal is in the differential diagnoses, testing for metabolites might help identify the parent substance.*

SUBSECTION 7

Psychotic Patients

When examining a psychotic patient, **avoid open-ended questions.** The Examiner asks only short questions, avoids abstract questioning style and long periods of silence. As the patient is presumably mistrustful and suspicious, one should make an effort to **explain in detail** the planned procedures and treatment decisions.

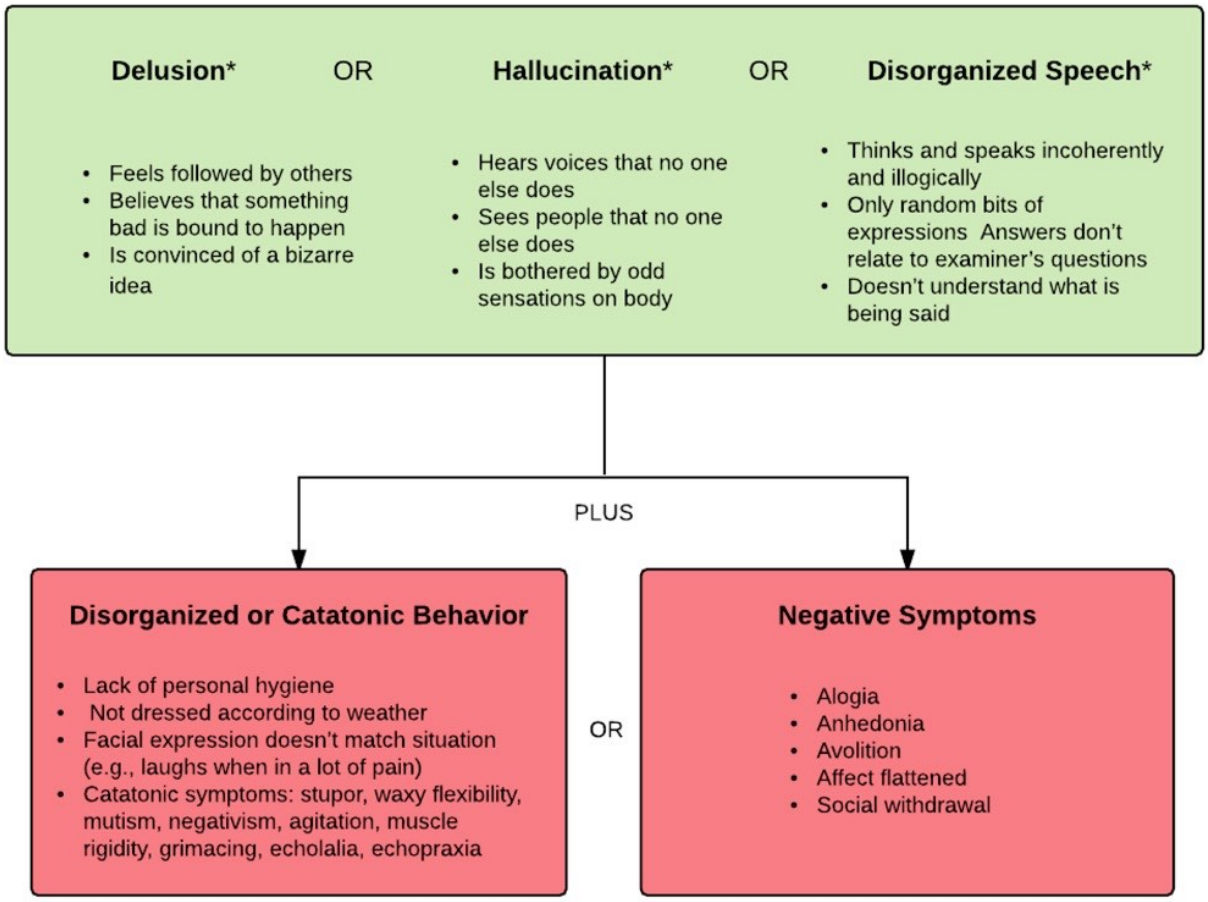
Note: in the emergency room, a definitive diagnosis often cannot be made, nor is it necessary before treating the presenting syndrome.

In the emergency room setting, one of the most common underlying psychiatric causes of psychosis is schizophrenia. Use the following **screening questions** if you suspect the patient may be psychotic:

- In the past month, were you convinced that others were following you or plotting against you?
- In the past month, have you heard a voice saying a few words or sentences, when there was no one around who might account for it?
- In the past month, have you felt that someone was controlling your thoughts or was inserting thoughts into your head that weren't you own?

If any one question is answered with "yes" a further examination regarding schizophrenia by the consultant psychiatrist is warranted.

FIGURE 12.7.1 Symptoms and Signs of Schizophrenia



*Note: At least two symptoms are required for a duration of one month or longer and one of the two needs to be a core symptom as marked by an asterisk**

Video of a patient with psychosis:

https://www.youtube.com/watch?v=ZB28gfSmz1Y&index=4&list=PL42hXq8UeG_PXnD21Eda1qH-p6-1Gh1vt1

SUBSECTION 8

Depressed Patients

Generally, when examining depressed patients one may need to **repeat some questions**, since attention and concentration may be decreased. Furthermore, when ruminations are prominent during the interview, you need **to gently interrupt and redirect the patient**.

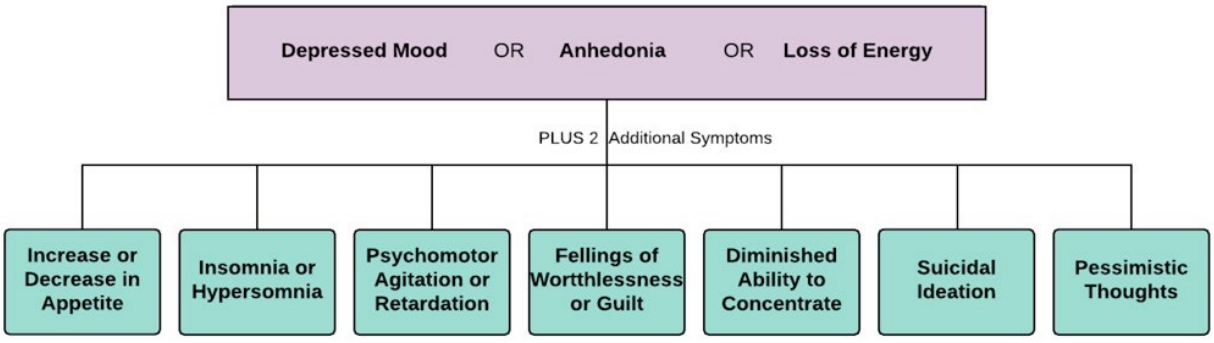
Note: Recent adverse life events are important risk factors for the onset of a major depressive episode. Asking about suicidal ideation is mandatory.

The following **screening questions** are useful:

- In the past two weeks, have you suffered from feelings of depression, sadness, or hopelessness? On how many days?
- In the past two weeks, have you lost interest or joy in your activities/hobbies? On how many days?

If one question is being answered with "almost every day" or both questions are answered "on several or most days", a further examination regarding a depressive disorder by a consultant psychiatrist is warranted.

FIGURE 12.8.1 Diagnosis of a Major Depressive Episode



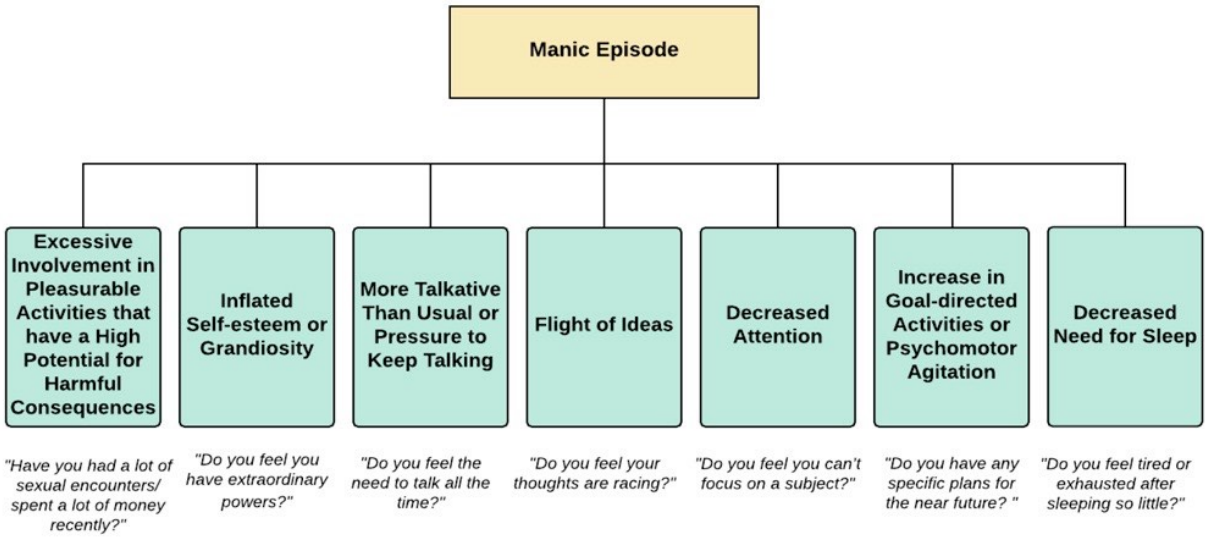
Note: At least two of the three core symptoms and two of the additional symptoms need to be present for at least two weeks.

SUBSECTION 9

Manic Patients

A manic episode is part of a bipolar disorder or it can occur as recurrent manic episodes, or as a schizoaffective disorder. It may also occur secondary to a neurological or general medical condition like during high-dose corticosteroid treatments (e.g. in acute attacks of multiple sclerosis or in other autoimmune diseases, or in malignant hyperthyroidism) or due to cocaine use. Occasionally it is seen in benzodiazepine withdrawal.

FIGURE 12.9.1 Diagnosis of a Manic Episode



Note: At least 3 features are reqired for a period of 1 week or more.

SUBSECTION 10

Borderline Patients

When a patient presents with suicidal ideations in the emergency room setting, the most common cause is a depressive disorder. Importantly, a **borderline personality disorder** must also be considered, especially if further features such as **self-mutilation**, **dissociation**, and **intoxication** with a mixture of substances are present: alcohol combined with prescribed and/or **OTC** drugs). When examining patients, one should ask whether they still have "the weapon" they cut themselves with (e.g., a blade, a knife) and where this weapon is right now to **assess for the current risk**. One should continue to ask if they have many drugs at home (regardless whether prescribed or not). Even if there is no sign of acute intoxication, there is an increased risk of intake when under stress due to high impulsivity being a typical feature of the disorder. Borderline patients can be very mistrusting when under stress. So in order to be able to conduct the interview, the examiner should use **validation strategies**. These include interview techniques displaying that one **understands** the patient regardless of lacking approval of her/his behavior. The physician should **respect** the patient and **should not be judgmental**. Well-intentioned but **unhelpful advice** like "a young woman with so much potential…" is not recommended. (Hong, 2016).

Note: Borderline patients are not always immediately recognizable as such. They often present to the neurologist with symptoms that may mimic epilepsy (i.e., dissociations). They also commonly show somatization tendencies and, thus, have often been pretreated by internists and neurologists prior to a psychiatric consultation (cf. Section 9).

Mnemonic for Borderline Personality Disorder "**IMPULSIVE**"

Impulsive

Moodiness

Paranoia or dissociation under stress

Unstable self-image

Labile intense relationships

Suicidal gestures

Inappropriate anger

Vulnerability to abandonment

Emptiness (feelings of)

SUBSECTION 11

Alcohol Use Disorder Patients

When an alcohol use disorder is suspected, one may use the **CAGE** screening tool, which is applicable in less than a minute:

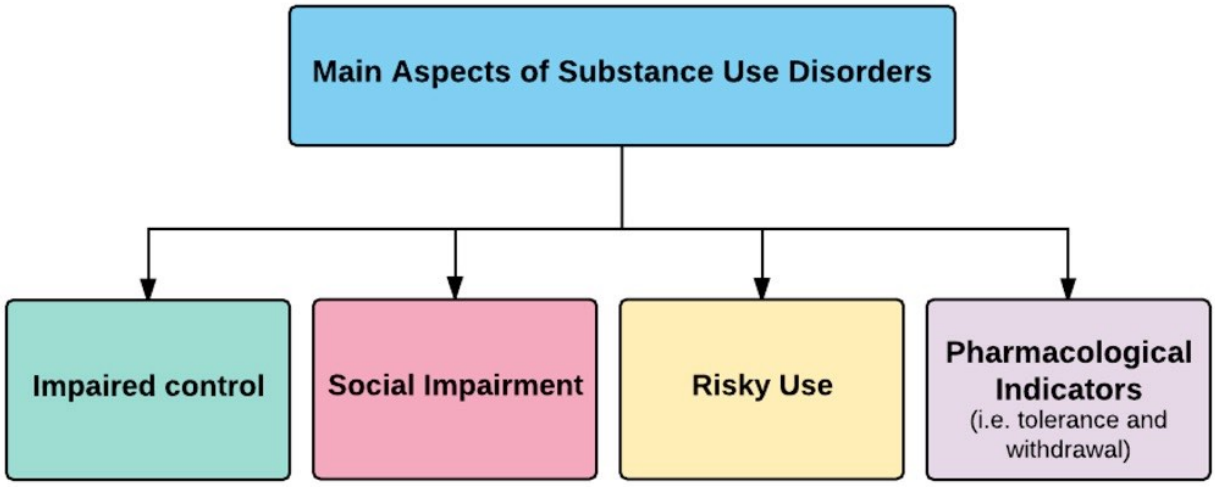
- Have you ever felt you needed to **C**ut down on your drinking?
- Have people **A**nnoyed you by criticizing your drinking?
- Have you ever felt **G**uilty about drinking?
- Have you ever felt you needed a drink first thing in the morning (**E**ye-opener) to steady your nerves or to get rid of a hangover?

Two or more "yes"-answers in the CAGE screening warrant a further examination.

In order to establish the diagnosis of any substance use disorder, the symptoms need to have been present in the past 12 months (American Psychiatric Association. & American Psychiatric Association. DSM-5 Task Force., 2013). The degree of severity is as follows:

- **mild** substance use disorder: the presence of **2-3** out of 11 criteria
- **moderate** substance use disorder: the presence of **4-5** out of 11 criteria
- **severe** substance use disorder: the presence of **6** and more out of 11 criteria

FIGURE 12.11.1 Substance Use Disorders



Mnemonic for Alcohol Use Disorder: "**WITHDRAW IT NOW**"

- **W**ithdrawal
- **I**nterest or Important activities given up or reduced
- **T**olerance
- **H**arm to social life known but still continues to drink
- **D**ange**R** to own health known but still continues to drink
- **A**mount of drinks more than intended
- **W**ork/school/family problems because of drinking
- **I**ncreased chances of getting hurt while or after drinking
- **T**oo much time spent with drinking or aftereffects
- **NO**thing else on mind except for drinking
- **W**anted to cut down or stop but could not

Appendix

Individual Case Presentation - common psychiatric disorders in the Emergency Room illustrating the spectrum of symptoms and signs.

Case 1: A Schizophrenic Patient

A 21-year-old male patient was admitted to the emergency room by paramedics who found him lying on the street. He presented with **mutism** and **stupor**. When physically examining the patient, **muscle rigidity** and **negativism** were noted. When finally the limbs were able to be moved, they remained in the imposed position, a symptom known as **waxy flexibility**. (This is different from "Gegenhalten" as described in Section 3.)

In order to differentiate between possible catatonic features of schizophrenia, and other causes of the clinical symptoms and signs (e.g., delirium, major depressive episode, manic episode, or factitious disorder), other sources of information were of utmost importance. The history of the patient's illness was taken from next of kin after having searched the patient's bag to find the telephone number of his close relatives. The patient had gradually stopped eating and drinking, had become slower in movement and speaking, had **withdrawn from social activities**, had become very **anxious**, had expressed **delusional ideas**, had talked to **imagined voices**, had increased his cigarette smoking and had neglected personal hygiene. A similar episode had occurred the year before. Together, these symptoms were highly suspicious of catatonic schizophrenia. Thus, 1 mg lorazepam was given because of its stupor-relieving potency, which then allowed for the diagnostic evaluation to be completed.

Case 2: A Depressed Patient

A 56-year-old male patient presented with **insomnia** and **anxiety** as his chief complaints. During history taking, we learned that he had

lost his job four months before admission and he was going through a **divorce** that his wife had filed. Prominent features of the mental status examination included **ruminations**, reduced concentration, **depressed mood**, **anhedonia**, and loss of appetite. Over the course of the interview, he admitted **suicidal ideations**. He was offered a place in a specialized inpatient treatment program for depression, which he gladly accepted.

Case 3: A Patient with Mania

A 32-year-old female teacher presented without a chief complaint. She said she came to the emergency room because her general practitioner advised her to do so. In the examination, she appeared to be **euphoric**, was flirtatious, **talked a lot** and was difficult to interrupt, answered questions only tangentially, and showed **flight of ideas**. She further reported that she **did not feel the need to sleep** lately. This suited her just fine as she was planning to **move to Australia** for good and was busy with the preparations. She said "the time has now come" and that she had already booked a ticket. She also had terminated her job as it was "holding her back". She had **spent all of her savings** in the past few days but did not mind to continue buying things she believed were nice to have by **overdrawing her account**. Her libido was increased and she **lacked any insight**. She finally reluctantly agreed to be admitted to the psychiatric ward.

Case 4: A Patient with Borderline Personality Disorder

A 21-year-old female patient presented with **self-mutilation** and **suicidal ideation**. She wore a lot of make-up and has several tattoos and piercings. On her **left forearm**, **several scars** and a fresh wound were noted. In the course of the examination, she started to cry and seemed depressed. When talking about her ex-partner **her mood changed** and she became **very angry**. She had **broken up** with him only a few hours ago because she **suspected him to have an affair**. Afterward she felt so stressed that she cut herself in order to **release her inner tension**. She also reported that she felt an urge to kill herself. She had had these suicidal thoughts for years but had

never attempted suicide, yet. Upon assessing her suicidal risk (see Subsection 12.3), there was no acute risk for suicide evident. After she felt calmer and more relieved, treatment options were discussed and an elective admission to a specialized DBT (dialectical behavior therapy) ward at a psychiatric department was recommended.

SECTION 13

FURTHER READING

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SECTION 14

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SECTION 15

LIST OF FIGURES

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Figure 2.2.4: Images 0 to 4: Courtesy of Professor Dr. Peter Meyer, University Hospital of Basle, Switzerland. Image 5: This image is used under the Creative Commons Attribution 3.0 Unported license, courtesy of Dr. Jonathan Trobe, M.D. University of Michigan Kellogg Eye Center. Image 5 was modified from its original form through color correction and cropping for incorporation into the figure. Original source: http://kellogg.umich.edu/theeyeshaveit/opticfundus/papilledema.html.

Movie 2.3.2: with permission by the patient

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Figure 6.1.1 b: Henry Vandike Carter, revised by Warren H. Lewis

Table 11.11.1: modified from Wijdicks EF et al. 2005, with permission of the author

SECTION 16

ACKNOWLEDGEMENTS

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Epicritical sensation

Sensation of touch, pressure, two-point discrimination, proprioceptive sensation, joint position, vibration sense (pallesthesia)

Position (proprioceptive)

This is tested on index finger and big toe, occasionally in larger joints. Test results are dependent on the joints tested and become less precise in the elderly. Test results are indicated in the approximate angle that can still be appreciated.

Two-point discrimination

Ability to discriminate between a single or a double prick stimulus. “Static” testing means leaving the two stimuli at one site at a time for a few seconds, (and for control purposes also as singlet prick at any level), followed by “dynamic” testing with moving the two pricks along the skin over ~10 mm.

Pallesthesia

Vibration sense

Tuning fork

Metal Instrument for testing vibration sense. The graded Rydell tuning fork used in continental Europe has a frequency of 64 Hz and can be read out by a scale, which is detachable. Without the detachable scales the frequency is 128 Hz. Common U. S. tuning forks have a frequency of 128 Hz (with weights) and 256 Hz without, and are precision instruments if struck to maximum intensity allowing for standardized duration of vibratory sensation. Vibration alarms of smart phones vibrate at frequencies much lower than those provided by the tuning forks and should not be used.

Hyp(o)esthesia

Subjective feeling of a stimulus is simply reduced

Allesthesia

Subjective feeling is felt in a different or strange way like pins and needles

Dysesthesia

Electric shocks or other uncomfortable sensations upon touch

Allodynia

Inappropriately painful sensation upon non-painful stimuli

Protopathic (pain/temp)

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Inappropriately strong pain sensation with mild painful stimuli

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Inappropriately weak pain sensation with strong painful stimuli

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Tactile character perception, i.e. perceiving numbers “written” on the skin

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Directional movement: tested by letting the patient describe the direction of a prick or finger movement on the skin

Extinction

The patient cannot detect two simultaneous stimuli on both sides of the body. By definition, perception of a single stimulus is still possible.

Paresis

Muscle weakness of single muscles, muscle groups or limbs, trunk, head

Paraparesis

Bilateral weakness of limbs (most frequently, both legs; rarely, both arms)

Tetraparesis

Weakness of all four limbs, optional plus trunk and head

Plegia (para-/tetraplegia)

Severe weakness, practically loss of function

Anosmia

Inability or impairment of smell

Miosis

Pupillary constriction

Mydriasis

Pupillary dilation

Anisocoria

Inequality of the pupils

OU/OS/OD

Both eyes/left eye/right eye

Papilledema

Optic nerve swelling due to increased intracranial pressure

Anop(s)ia

Loss of vision: defined by sector, quadrant, hemifield

Argyll-Robertson pupil

Constriction with near focus, but not with light; common in syphilis

Adie pupil

Tonically dilated pupil; postganglionic parasympathetic failure

Horner syndrome

Mild ptosis, miosis, anhydrosis; sympathetic failure

Exo/eso/hyper/hypo-tropia

Outward/inward/upward/downward deviation of the eye also named strabismus or squint

Exo/eso-phoria

Still compensated misalignment of the optic axis, i.e. latent strabismus, comes out when binocular fixation is interrupted or with drugs and fatigue

Ophthalmoplegia

Inability to move the eyeball

Gaze paralysis

Supranuclear pathology causes limitations in gaze, may be asymmetrical

Version

Conjugate movements of both eyes

Duction

Movements of a single eye

Vergence

Movements of the eyes in the same (convergent) or opposite (divergent) directions

Nystagmus

Physiologic rapid eye movements upon body rotation or hot/cold water in the ear (vestibular n,) and when fixating on moving objects (optokinetic n. - train, car). Pathologic if spontaneous or induced by certain head positions. Many different types. The fast phase names the direction of nystagmus except for pendular nystagmus where both phases are equal.

Vertigo

Specifically the perception of spinning/moving either of the patient or the environment often associated with nystagmus

Dysequilibirum

A general sense of unsteadiness, usually of the body

Myotonia Duplikat

Slowness of relaxation after skeletal muscle contraction, sustained contraction also upon percussion with a reflex hammer

Anarthria

Inability or impairment in articulation of syllables and words

Dysphagia

Subjective impression of a patient with swallowing difficulties

Astasia

Inability to stand upright

Abasia

Inability to walk normally; normal base implies that the medial edge of each foot touches an imaginary line in the direction of walking

Ataxia

Being out of balance with increased sway (gait, stance) or reaching out with a large sway (best exemplified in alcohol intoxication). Ataxic gait: Broad-based and unsteady swaying of the body; “drunkard’s gait”, may be mixed with spastic component (spastic-ataxic)

Dysmetria

The movement misses the target (includes hypermetria, exaggerated movement amplitude) and is best detectable on sideways movement toward the examiner's finger

Steppage gait

Lifting of the knees high with feet limply slapping floor due to dorsiflexion weakness

Spasticity

Velocity-dependent increase in tone (velocity dependence may be difficult to detect in very severe cases), predominating in flexors in upper limbs, in extensors in lower limbs

Spastic gait: Stiff and hyperextended legs resulting in forced plantarflexion with circumduction and foot dragging

Festinating gait

Short rapid (often shuffling steps), with difficulty stopping

Magnetic gait

Short delay before foot lift-off, as if stuck to the floor

MRC grades

Grading system by the British Research Council (1942; O'Brian 2010). Intermediate grades have also been used (see Muscle strength testing below)

0= no movement

1= flicker movement

2= muscle or limb movement but not against gravity;

3= muscle/limb movement against gravity in full range

4= good but weaker than normal muscle strength

5= normal strength

A modified system has recently been proposed based on a mathematical model (Rasch) which may have better discrimination in strength testing (Vanhoutte et al., 2012)

UMN

Upper motoneuron: pyramidal neurons from the cortex projecting down to the anterior spinal nerve nuclei

LMN

Lower motoneuron: peripheral α-motor neurons with their cell bodies in the anterior horn cells of the spinal cord

Rigidity

Velocity-independent increase in muscle tone ("lead-pipe like", stiff even at rest) involving all muscles, including the axial musculature

Paratonia

Continual opposition of movements in all directions (Gegenhalten) or continual assistance of movement in all directions (Mitgehen)

Myotonia

Hyperactivation of skeletal muscle contraction resulting in cramps with slowness of relaxation after contraction, and sustained contraction also upon percussion with a reflex hammer

Neuromyotonia

A wave-like, spontaneous movement of muscle regions, sometimes mimicking massive fasciculations, frequently associated with muscle hypertrophy and stiffness

Fasciculations

Short and circumscribed, multifocal, irregular movements of muscle “fascicles”, sometimes with visible change of joint position if in the hands and feet

Dystonia

Involuntary muscle contractions associated with repetitive and often twisting movements as well as awkward, irregular postures

Chorea

Rapid movements of local areas or multifocal activities that may appear almost dancelike

Ballismus

Throwing movements

Myoclonus

Jerky involuntary muscle contractions leading to abrupt movements of the limb, trunk, or facial

Tremor

Resting tremor (hands, feet, head), usually around 3-6 per second; action tremor (only on active innervation, such as stretching out the arms), sometimes accentuated by intentional movements

Postural tremor

A special form of action tremor: postural tremor occurs when the person maintains a position against gravity. Postural tremor may be position dependent. Holding arms out straight in flexion or with fingers held in front of the nose may accentuate tremor with variable frequency.

Kinetic tremor

A special form of action tremor: appears when the patient performs translational movements such as repetitive movements between his/her nose and the finger of the examiner or with pouring water from cup-to-cup.

Intention tremor

A special form of action termor: a.k.a. tremor of goal-directed movement: the closer the patient gets to the target the more irregular and shaky becomes the movement, the amplitude of the tremor increases as the effector (e.g., finger or toe) approaches the goal of the movement.

Reflex hammer

There are various types of hammers around, some carry the names of respected institutions or of famous neurologists of the past (Lanska,1989) . A heavy weight hammer has the advantage of allowing a more reproducible tap.

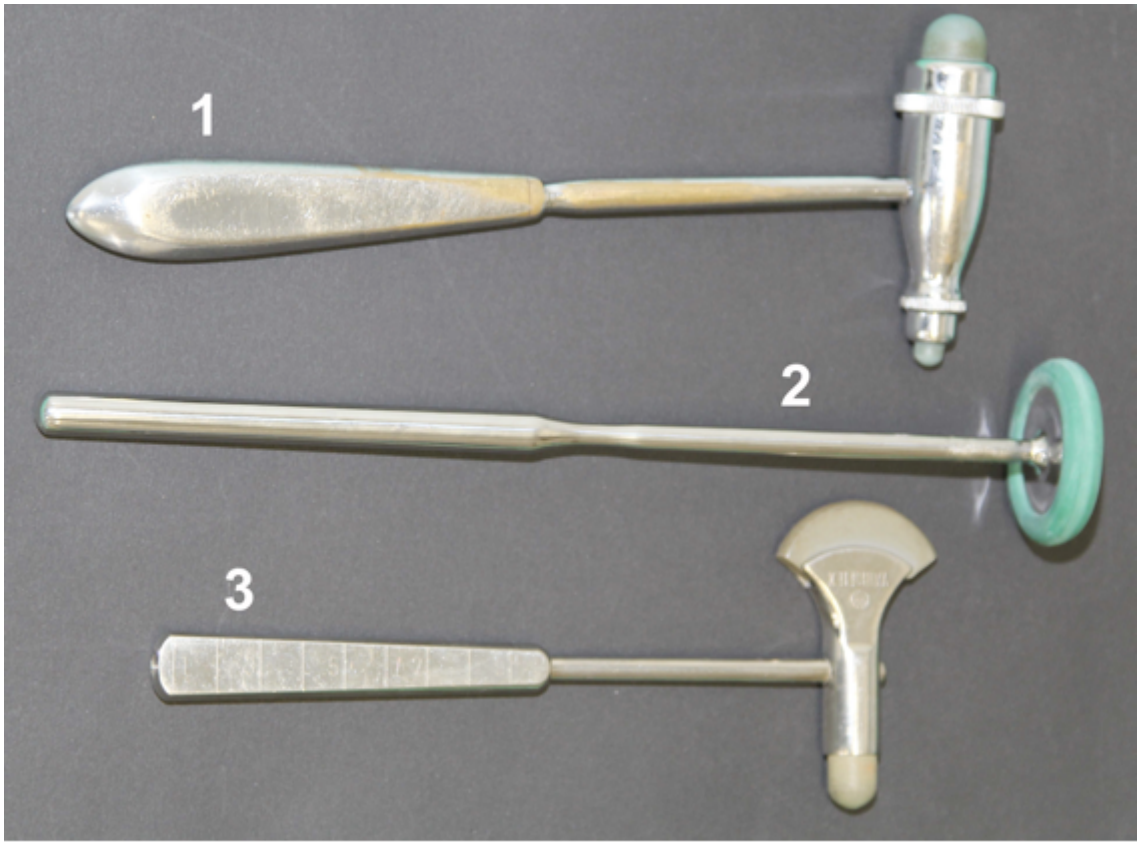

Fig. 4.4.1 Three types of reflex hammers

1 - Trömner hammer (Mayo hammer)
2 - Queen Square hammer
3 - Pediatric hammer

The Taylor hammer is not depicted

Pyramidal signs

This denotes a group of reflexes that are positive when a suprasegmental or supraspinal (central) lesion exists. Unfortunately, the term is a misnomer as well-defined lesions of the pyramidal tract in animal experiments typically lead to flaccid paresis without "pyramidal" signs. The most important and sensitive is the Babinski sign often combined with the Chaddock reflex. It is best elicited by a wooden spatula, or a thick wooden rod.

Dyssynergia

Disorder of finely tuned complex movements (evident in finger-nose-finger testing), in which the movement is broken down into components: reach, turn hand, hit target

Prosody

The “melody” of speech – it varies largely with ethnicity and this is even maintained in the newly learned foreign language. Loss of prosody: automated (computer) language

Paraphasias

Semantic: confusion of words or replacement of one word by another real word, which are more or less related to the target word, e.g. lion for tiger

Phonematic: wrong or displaced syllables/vowels/consonants, e.g. “vast fell I travered to Chicago” for “last fall I travelled to Chicago”

Wernicke's aphasia

Fluent aphasia with promiment paragrammatism, complete or partial inability to comprehend spoken language. The "fluent" aphasia allows them to babble on as if speaking a foreign language fluently.

Dyslexia

Inability to read and comprehend a text or even to read single letters

Dyscalculia

Inability or reduced ability to comprehend and read numbers and perform simple math

Acalculia

Inability or reduced ability to comprehend and read numbers and perform simple math

Alexia

Inability to read and comprehend a text or even to read single letters

Neologisms

There may be newly formed nonsense words as if from a foreign language, in contrast to paraphasias, the target word cannot be recognized.

Paragrammatism

Wrong (although complex) grammar of a sentence, e.g. "I not never next here come back" for "I will never come back here"

Agrammatism

Reduced, telegraphic grammar, e.g. "Monday ... Dad ... hospital"

Speech automatisms

Utterances without any content or meaning which do not fit into the context, severest form: recurring utterances (e.g. "dododo")

Broca's aphasia

Non-fluent aphasia with prominent agrammatism, while comprehension is only mildly affected

Anomic aphasia

a type of aphasia mainly characterised by many word-finding difficulties

Paraesthesia

Subjective feeling is felt in a different or strange way like pins and needles.

Dysarthria

Inability or impairment in articulation of syllables and words

Hyposmia

Inability or impairment of smell

Torsion

Movements of a single eye

Anhedonia

Marked decrease in ability to experience pleasure

Avolition

Lack of interest or engagement in goal-directed behavior

Cluster B personality disorder

Borderline, histrionic, narcissistic, and antisocial personality disorder

Delusion

Uncorrectable conviction that is untrue and implausible or bizarre

Echolalia

Mimicking someone's speech

Echopraxia

Mimicking someone's movements

Flight of ideas

Accelerated stream of thought, where the person jumps from one idea to the next without elaborating on any one thought; a logical connection between thoughts may remain, even if not immediately obvious

Hallucination

Sensory perception in an awake person without external stimulus

Legal highs

Legal drugs and substances that are used for their intoxicating effects

Mutism

Marked decrease in speech production

Rumination

Unpleasant urge to think about own mistakes and adverse events of the past over and over again

OTC

"over the counter"; an attribute used for medication that can be legally bought without a prescription

Stupor

Marked decrease in reactivity to the environment, in spontaneous movements, and in activity in an awake patient (different from stupor in section 11.2)

Waxy flexibility

Limbs of the patient remain in the position they were imposed by the examiner (different from paratonia -cf. section 3.2)

Red glass

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Epicritical sensation

Sensation of touch, pressure, two-point discrimination, proprioceptive sensation, joint position, vibration sense (pallesthesia)

Verwandte Glossarbegriffe

Two-point discrimination

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Position (proprioceptive)

This is tested on index finger and big toe, occasionally in larger joints. Test results are dependent on the joints tested and become less precise in the elderly. Test results are indicated in the approximate angle that can still be appreciated.

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[Kapitel 6 - Section 6 – Sensory system]

Two-point discrimination

Ability to discriminate between a single or a double prick stimulus. "Static" testing means leaving the two stimuli at one site at a time for a few seconds, (and for control purposes also as singlet prick at any level), followed by "dynamic" testing with moving the two pricks along the skin over ~10 mm.

Verwandte Glossarbegriffe

Epicritical sensation

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Kapitel 6 - Sensory Examination
Kapitel 6 - Sensory Examination
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Kapitel 9 - Functional Disorders of Vision, Hearing, and Cutaneous Sensation

Pallesthesia

Vibration sense

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[Kapitel 6 - Sensory Examination](#)

Tuning fork

Metal Instrument for testing vibration sense. The graded Rydell tuning fork used in continental Europe has a frequency of 64 Hz and can be read out by a scale, which is detachable. Without the detachable scales the frequency is 128 Hz. Common U. S. tuning forks have a frequency of 128 Hz (with weights) and 256 Hz without, and are precision instruments if struck to maximum intensity allowing for standardized duration of vibratory sensation. Vibration alarms of smart phones vibrate at frequencies much lower than those provided by the tuning forks and should not be used.

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Hyp(o)esthesia

Subjective feeling of a stimulus is simply reduced

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Allesthesia

Subjective feeling is felt in a different or strange way like pins and needles

Verwandte Glossarbegriffe

Paraesthesia

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Kapitel 9 - Functional Disorders of Vision, Hearing, and Cutaneous Sensation

Dysesthesia

Electric shocks or other uncomfortable sensations upon touch

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Kapitel 6 - Section 6 – Sensory system
Kapitel 9 - Functional Disorders of Vision, Hearing, and Cutaneous Sensation

Allodynia

Inappropriately painful sensation upon non-painful stimuli

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Kapitel 6 - Section 6 – Sensory system

Protopathic (pain/temp)

Pain sensation: pin prick, painful pressure, painful heat, painful cold. Temperature sensation: cool and warm (not painful)

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Hyperalgesia

Inappropriately strong pain sensation with mild painful stimuli

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[Kapitel 6 - Section 6 – Sensory system]

Hyp(o)algesia

Inappropriately weak pain sensation with strong painful stimuli

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[Kapitel 6 - Section 6 – Sensory system]

Stereognosis

Perceiving objects in a 3-dimensional way (its gestalt); this is done by presenting objects of daily livings (such as paper clips) to either hand with the patient asked to identify them with eyes closed.

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[Kapitel 6 - Section 6 – Sensory system]

Graphesthesia

Tactile character perception, i.e. perceiving numbers “written” on the skin

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Kapitel 6 - Section 6 – Sensory system
Kapitel 6 - Sensory Examination

Kinesthesia

Directional movement: tested by letting the patient describe the direction of a prick or finger movement on the skin

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[Kapitel 6 - Section 6 – Sensory system]

Extinction

The patient cannot detect two simultaneous stimuli on both sides of the body. By definition, perception of a single stimulus is still possible.

Index

Paresis

Muscle weakness of single muscles, muscle groups or limbs, trunk, head

Verwandte Glossarbegriffe

Paraparesis, Tetraparesis

Index

Paraparesis

Bilateral weakness of limbs (most frequently, both legs; rarely, both arms)

Verwandte Glossarbegriffe

Paresis

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Kapitel 4 - Monosynaptic (tendon) reflexes
Kapitel 10 - Section 10 – Summarizing, Categorizing, and Structuring Pathologic Findings

Tetraparesis

Weakness of all four limbs, optional plus trunk and head

Verwandte Glossarbegriffe

Paresis

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Abschnitt 2 - Preface

Plegia (para-/tetraplegia)

Severe weakness, practically loss of function

Verwandte Glossarbegriffe

Ophthalmoplegia

Index

Abschnitt 2 - Preface

Anosmia

Inability or impairment of smell

Verwandte Glossarbegriffe

Hyposmia

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Kapitel 2 - Section 2 – Cranial Nerves (CN)

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Pupillary constriction

Verwandte Glossarbegriffe

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Kapitel 2 - Section 2 – Cranial Nerves (CN)
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Kapitel 2 - Oculomotor nerves (CN III - oculomotor n.; CN IV - trochlear n.; CN VI - abducens n.)

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Pupillary dilation

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[Kapitel 2 - Section 2 – Cranial Nerves (CN)]

Anisocoria

Inequality of the pupils

Index

[Kapitel 2 - Section 2 – Cranial Nerves (CN)]

OU/OS/OD

Both eyes/left eye/right eye

Index

[Kapitel 2 - Section 2 – Cranial Nerves (CN)]

Papilledema

Optic nerve swelling due to increased intracranial pressure

Index

Anop(s)ia

Loss of vision: defined by sector, quadrant, hemifield

Index

[Kapitel 2 - Section 2 – Cranial Nerves (CN)]

Argyll-Robertson pupil

Constriction with near focus, but not with light; common in syphilis

Index

[Kapitel 2 - Section 2 – Cranial Nerves (CN)]

Adie pupil

Tonically dilated pupil; postganglionic parasympathetic failure

Index

[Kapitel 2 - Section 2 – Cranial Nerves (CN)]

Horner syndrome

Mild ptosis, miosis, anhydrosis; sympathetic failure

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Miosis

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Kapitel 2 - Section 2 – Cranial Nerves (CN)

Kapitel 2 - Oculomotor nerves (CN III - oculomotor n.; CN IV - trochlear n.; CN VI - abducens n.)

Exo/eso/hyper/hypo-tropia

Outward/inward/upward/downward deviation of the eye also named strabismus or squint

Index

[Kapitel 2 - Section 2 – Cranial Nerves (CN)]

Exo/eso-phoria

Still compensated misalignment of the optic axis, i.e. latent strabismus, comes out when binocular fixation is interrupted or with drugs and fatigue

Index

[Kapitel 2 - Section 2 – Cranial Nerves (CN)]

Ophthalmoplegia

Inability to move the eyeball

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[Plegia (para-/tetraplegia)]

Index

[Kapitel 2 - Section 2 – Cranial Nerves (CN)]

Gaze paralysis

Supranuclear pathology causes limitations in gaze, may be asymmetrical

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[Kapitel 2 - Section 2 – Cranial Nerves (CN)]

Version

Conjugate movements of both eyes

Index

[Kapitel 2 - Section 2 – Cranial Nerves (CN)]

Duction

Movements of a single eye

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[Torsion]

Index

[Kapitel 2 - Section 2 – Cranial Nerves (CN)]

Vergence

Movements of the eyes in the same (convergent) or opposite (divergent) directions

Index

[Kapitel 2 - Section 2 – Cranial Nerves (CN)]

Nystagmus

Physiologic rapid eye movements upon body rotation or hot/cold water in the ear (vestibular n,) and when fixating on moving objects (optokinetic n. - train, car). Pathologic if spontaneous or induced by certain head positions. Many different types. The fast phase names the direction of nystagmus except for pendular nystagmus where both phases are equal.

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[Vertigo](#)

Index

Vertigo

Specifically the perception of spinning/moving either of the patient or the environment often associated with nystagmus

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Nystagmus

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Kapitel 2 - Oculomotor nerves (CN III - oculomotor n.; CN IV - trochlear n.; CN VI - abducens n.)
Kapitel 2 - Vestibulocochlear nerve (syn.: n. statoacusticus; CN VIII)

Dysequilibirum

A general sense of unsteadiness, usually of the body

Index

[Kapitel 2 - Section 2 – Cranial Nerves (CN)]

Myotonia Duplikat

Slowness of relaxation after skeletal muscle contraction, sustained contraction also upon percussion with a reflex hammer

Anarthria

Inability or impairment in articulation of syllables and words

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Dysarthria

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Kapitel 2 - Section 2 – Cranial Nerves (CN)

Kapitel 8 - Short-term and long-term memory

Dysphagia

Subjective impression of a patient with swallowing difficulties

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[Kapitel 7 - Section 7 – Autonomic Nervous System]

Astasia

Inability to stand upright

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[Abasia]

Index

[Kapitel 3 - Section 3 – Motor System (A)]

Abasia

Inability to walk normally; normal base implies that the medial edge of each foot touches an imaginary line in the direction of walking

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Astasia

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Kapitel 3 - Section 3 – Motor System (A)

Ataxia

Being out of balance with increased sway (gait, stance) or reaching out with a large sway (best exemplified in alcohol intoxication). **Ataxic gait**: Broad-based and unsteady swaying of the body; "drunkard's gait", may be mixed with spastic component (spastic-ataxic)

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Spasticity

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Kapitel 9 - Functional Motor Symptoms

Dysmetria

The movement misses the target (includes hypermetria, exaggerated movement amplitude) and is best detectable on sideways movement toward the examiner's finger

Index

Steppage gait

Lifting of the knees high with feet limply slapping floor due to dorsiflexion weakness

Index

[Kapitel 3 - Section 3 – Motor System (A)]

Spasticity

Velocity-dependent increase in tone (velocity dependence may be difficult to detect in very severe cases), predominating in flexors in upper limbs, in extensors in lower limbs

Spastic gait: Stiff and hyperextended legs resulting in forced plantarflexion with circumduction and foot dragging

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Ataxia

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Festinating gait

Short rapid (often **shuffling** steps), with difficulty stopping

Index

[Kapitel 3 - Section 3 – Motor System (A)]

Magnetic gait

Short delay before foot lift-off, as if stuck to the floor

Index

[Kapitel 3 - Section 3 – Motor System (A)]

MRC grades

Grading system by the British Research Council (1942; O'Brian 2010). Intermediate grades have also been used (see Muscle strength testing below)

0= no movement

1= flicker movement

2= muscle or limb movement but not against gravity;

3= muscle/limb movement against gravity in full range

4= good but weaker than normal muscle strength

5= normal strength

A modified system has recently been proposed based on a mathematical model (Rasch) which may have better discrimination in strength testing (Vanhoutte et al., 2012)

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Kapitel 3 - Section 3 – Motor System (A)
Kapitel 3 - Further tests of the motor system
Kapitel 3 - Further tests of the motor system
Kapitel 4 - Monosynaptic (tendon) reflexes

UMN

Upper motoneuron: pyramidal neurons from the cortex projecting down to the anterior spinal nerve nuclei

Index

LMN

Lower motoneuron: peripheral α-motor neurons with their cell bodies in the anterior horn cells of the spinal cord

Index

[Kapitel 3 - Section 3 – Motor System (A)]
[Kapitel 4 - Monosynaptic (tendon) reflexes]

Rigidity

Velocity-independent increase in muscle tone ("lead-pipe like", stiff even at rest) involving all muscles, including the axial musculature

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Paratonia

Continual opposition of movements in all directions (Gegenhalten) or continual assistance of movement in all directions (Mitgehen)

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Kapitel 3 - Further tests of the motor system

Myotonia

Hyperactivation of skeletal muscle contraction resulting in cramps with slowness of relaxation after contraction, and sustained contraction also upon percussion with a reflex hammer

Index

Neuromyotonia

A wave-like, spontaneous movement of muscle regions, sometimes mimicking massive fasciculations, frequently associated with muscle hypertrophy and stiffness

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Kapitel 3 - Section 3 – Motor System (A)
Kapitel 3 - Further tests of the motor system
Kapitel 3 - Further tests of the motor system

Fasciculations

Short and circumscribed, multifocal, irregular movements of muscle “fascicles”, sometimes with visible change of joint position if in the hands and feet

Index

Dystonia

Involuntary muscle contractions associated with repetitive and often twisting movements as well as awkward, irregular postures

Index

Chorea

Rapid movements of local areas or multifocal activities that may appear almost dancelike

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Kapitel 3 - Section 3 – Motor System (A)
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Kapitel 3 - Gait and stance

Ballismus

Throwing movements

Index

[Kapitel 3 - Section 3 – Motor System (A)]

Myoclonus

Jerky involuntary muscle contractions leading to abrupt movements of the limb, trunk, or facial

Index

Tremor

Resting tremor (hands, feet, head), usually around 3-6 per second; action tremor (only on active innervation, such as stretching out the arms), sometimes accentuated by intentional movements

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Intention tremor, Kinetic tremor, Postural tremor

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Postural tremor

A special form of action tremor: postural tremor occurs when the person maintains a position against gravity. Postural tremor may be position dependent. Holding arms out straight in flexion or with fingers held in front of the nose may accentuate tremor with variable frequency.

Verwandte Glossarbegriffe

Tremor

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Kinetic tremor

A special form of action tremor: appears when the patient performs translational movements such as repetitive movements between his/her nose and the finger of the examiner or with pouring water from cup-to-cup.

Verwandte Glossarbegriffe

Tremor

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Kapitel 3 - Section 3 – Motor System (A)
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Kapitel 5 - Finger-nose and finger-nose-finger tests

Intention tremor

A special form of action termor: a.k.a. tremor of goal-directed movement: the closer the patient gets to the target the more irregular and shaky becomes the movement, the amplitude of the tremor increases as the effector (e.g., finger or toe) approaches the goal of the movement.

Verwandte Glossarbegriffe

Tremor

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Kapitel 3 - Section 3 – Motor System (A)
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Kapitel 5 - Section 5 – Coordination (Motor System B)

Reflex hammer

There are various types of hammers around, some carry the names of respected institutions or of famous neurologists of the past (Lanska,1989) . A heavy weight hammer has the advantage of allowing a more reproducible tap.

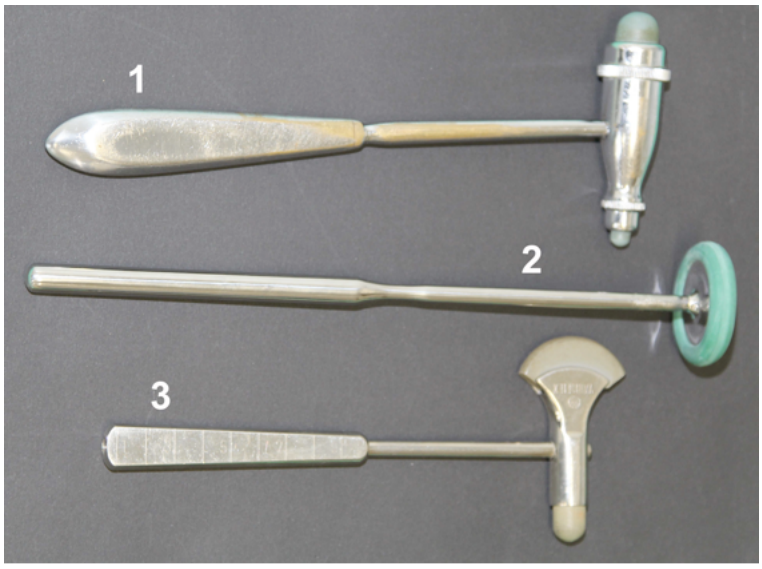

FIG. 4.4.1 Three types of reflex hammers

1 - Trömner hammer (Mayo hammer)
2 - Queen Square hammer
3 - Pediatric hammer

The Taylor hammer is not depicted

Index

Pyramidal signs

This denotes a **group of reflexes** that are positive when a suprasegmental or supraspinal (central) lesion exists. Unfortunately, the term is a **misnomer** as well-defined lesions of the pyramidal tract in animal experiments typically lead to flaccid paresis without "pyramidal" signs. The most important and sensitive is the Babinski sign **often combined** with the Chaddock reflex. It is best elicited by a wooden spatula, or a thick wooden rod.

Index

Dyssynergia

Disorder of finely tuned complex movements (evident in finger-nose-finger testing), in which the movement is broken down into components: reach, turn hand, hit target

Index

Kapitel 5 - Section 5 – Coordination (Motor System B)

Prosody

The “melody” of speech – it varies largely with ethnicity and this is even maintained in the newly learned foreign language.
Loss of prosody: automated (computer) language

Index

Kapitel 8 - Testing for Language and Calculus and Acalculia

Paraphasias

Semantic: confusion of words or replacement of one word by another real word, which are more or less related to the target word, e.g. lion for tiger

Phonematic: wrong or displaced syllables/vowels/consonants, e.g. “vast fell I travered to Chicago” for “last fall I travelled to Chicago”

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Neologisms

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Kapitel 8 - Testing for Language and Calculus and Acalculia
Kapitel 8 - Testing for Language and Calculus and Acalculia

Wernicke's aphasia

Fluent aphasia with promiment paragrammatism, complete or partial inability to comprehend spoken language. The "fluent" aphasia allows them to babble on as if speaking a foreign language fluently.

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Paragrammatism

Index

Dyslexia

Inability to read and comprehend a text or even to read single letters

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Alexia

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Dyscalculia

Inability or reduced ability to comprehend and read numbers and perform simple math

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Acalculia

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Kapitel 8 - Testing for Language and Calculus and Acalculia

Acalculia

Inability or reduced ability to comprehend and read numbers and perform simple math

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Dyscalculia

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Kapitel 8 - Testing for Language and Calculus and Acalculia

Alexia

Inability to read and comprehend a text or even to read single letters

Verwandte Glossarbegriffe

Dyslexia

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Kapitel 8 - Testing for Language and Calculus and Acalculia

Neologisms

There may be newly formed nonsense words as if from a foreign language, in contrast to paraphasias, the target word cannot be recognized.

Verwandte Glossarbegriffe

Paraphasias

Index

Kapitel 8 - Testing for Language and Calculus and Acalculia

Paragrammatism

Wrong (although complex) grammar of a sentence, e.g. “I not never next here come back” for “I will never come back here”

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Wernicke’s aphasia

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Kapitel 8 - Testing for Language and Calculus and Acalculia

Agrammatism

Reduced, telegraphic grammar, e.g. “Monday … Dad … hospital”

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[Kapitel 8 - Testing for Language and Calculus and Acalculia]

Speech automatisms

Utterances without any content or meaning which do not fit into the context, severest form: recurring utterances (e.g. “dododo”)

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[Kapitel 8 - Testing for Language and Calculus and Acalculia]

Broca's aphasia

Non-fluent aphasia with prominent agrammatism, while comprehension is only mildly affected

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[Kapitel 8 - Testing for Language and Calculus and Acalculia]

Anomic aphasia

a type of aphasia mainly characterised by many word-finding difficulties

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[Kapitel 8 - Testing for Language and Calculus and Acalculia]

Paraesthesia

Subjective feeling is felt in a different or strange way like pins and needles.

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Allesthesia

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Kapitel 6 - Section 6 – Sensory system

Dysarthria

Inability or impairment in articulation of syllables and words

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Anarthria

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Kapitel 2 - Hypoglossus nerve (CN XII)
Kapitel 8 - Testing for Language and Calculus and Acalculia

Hyposmia

Inability or impairment of smell

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[Anosmia]

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[Kapitel 2 - Section 2 – Cranial Nerves (CN)]
[Kapitel 2 - Olfactory nerve (CN I)]

Torsion

Movements of a single eye

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Duction

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Kapitel 2 - Section 2 – Cranial Nerves (CN)

Anhedonia

Marked decrease in ability to experience pleasure

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Kapitel 12 - Section 12 – The Psychiatric Examination in the Emergency RoomBooks

Kapitel 12 - Appendix

Avolition

Lack of interest or engagement in goal-directed behavior

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[Kapitel 12 - Section 12 – The Psychiatric Examination in the Emergency RoomBooks](#)

Cluster B personality disorder

Borderline, histrionic, narcissistic, and antisocial personality disorder

Index

Delusion

Uncorrectable conviction that is untrue and implausible or bizarre

Index

Kapitel 12 - Section 12 – The Psychiatric Examination in the Emergency RoomBooks

Kapitel 12 - Mental Status Examination

Echolalia

Mimicking someone's speech

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Kapitel 12 - Section 12 – The Psychiatric Examination in the Emergency RoomBooks

Echopraxia

Mimicking someone's movements

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[Kapitel 12 - Section 12 – The Psychiatric Examination in the Emergency RoomBooks](#)

Flight of ideas

Accelerated stream of thought, where the person jumps from one idea to the next without elaborating on any one thought; a logical connection between thoughts may remain, even if not immediately obvious

Index

Hallucination

Sensory perception in an awake person without external stimulus

Index

Legal highs

Legal drugs and substances that are used for their intoxicating effects

Index

Mutism

Marked decrease in speech production

Index

Rumination

Unpleasant urge to think about own mistakes and adverse events of the past over and over again

Index

OTC

"over the counter"; an attribute used for medication that can be legally bought without a prescription

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Kapitel 12 - Section 12 – The Psychiatric Examination in the Emergency RoomBooks
Kapitel 12 - Borderline Patients

Stupor

Marked decrease in reactivity to the environment, in spontaneous movements, and in activity in an awake patient (different from stupor in section 11.2)

Index

Waxy flexibility

Limbs of the patient remain in the position they were imposed by the examiner (different from paratonia -cf. section 3.2)

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Kapitel 12 - Appendix

Red glass

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