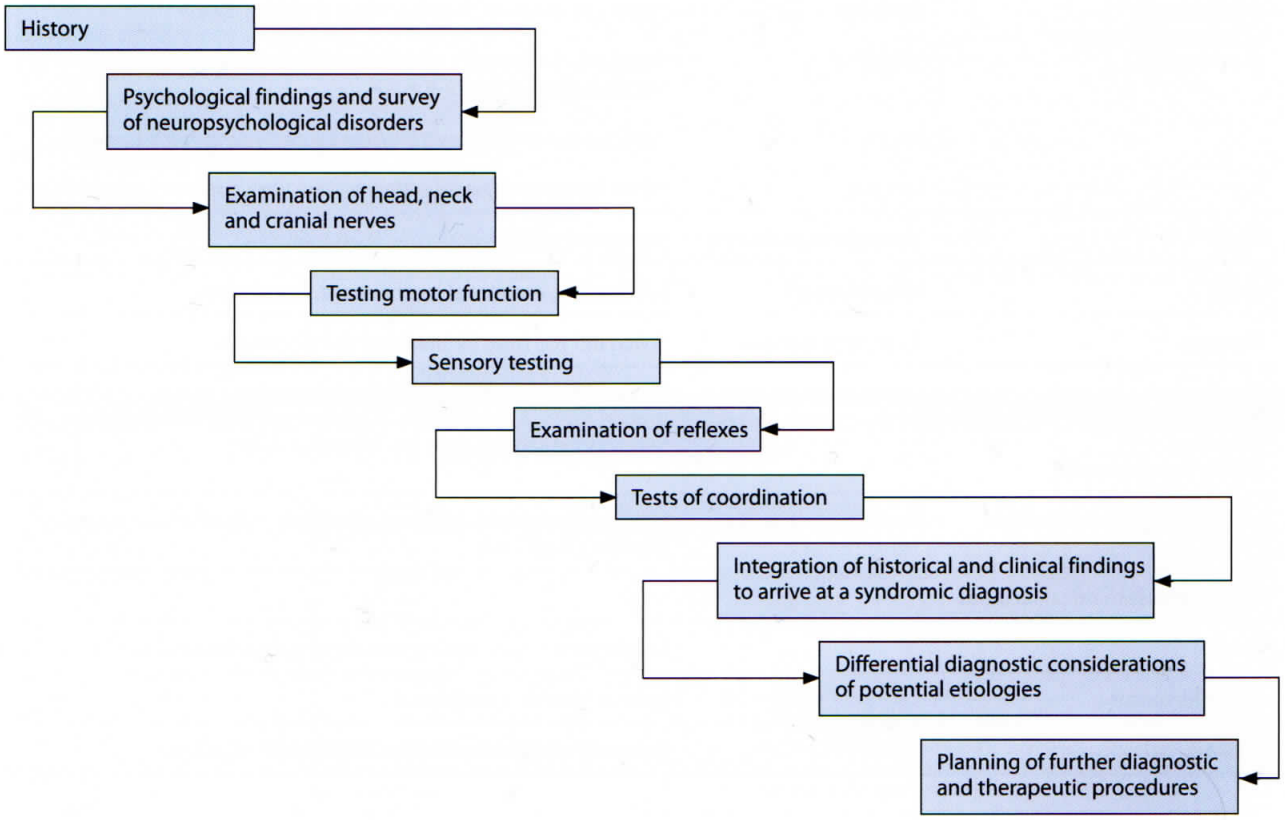
Chapter 21

Neurology

K. Gardill and H. Wiethölter

Normally, patients with visual disturbances initially seek the care of an ophthalmologist, even though the underlying cause of the problem, and more importantly its management, falls more into the field of the neurologist. The ophthalmologist is therefore heir to an important responsibility and must be on the watch for other symptoms that might accompany the visual problems. Only in this way can patients be given prompt and appropriate referral to neurological or neuroradiological consultants. Frequently, the clinician can accomplish this with only a few specific neurological tests, which can yield important information needed for the planning of further investigation and management of the problem.

As a rule, a complete neurological evaluation should be governed by a specific and well-thought sequence of tests (analogous to the sequence discussed in this chapter), to ensure that the function of all relevant systems is covered. Particularly in neurology is it generally possible to identify accurately the disease locus at fault by means of a careful clinical examination, combined with a detailed history. Given the limitations of time, an ophthalmological examination will be able to include only a few specific elements of the neurological examination, and yet with an adequate knowledge of the neuroanatomic and physiologic principles involved, should allow a close topographic localization and etiologic classification, based on a few specific symptoms and their associated findings.



History, Psychological Findings, and Neuropsychological Disorders

At the start of every neurological examination, there should be at least a minimal attempt to take a history of the patient's symptoms. This practice provides the physician with an opportunity to learn in general about the patient's psychological state, including speaking and language disorders or other neuropsychological deficits. Other individual or isolated problems may be difficult to draw out and require a more targeted form of questioning.

Psychological State

Pearl

The state of the patient's awareness of his or her identity and orientation to time and place is essential information and fundamentally important for an understanding of the problems he/she faces (■ Table 21.1).

Clues about the presence of a neurodegenerative disorder (for example, vascular encephalopathy with dementia and ocular motility disturbances, or Richardson-Steele-Olszewski syndrome with dementia and vertical gaze palsy) also help with assessing the reliability of the patient's historical accounts.

Neuropsychological Disorders

Acquired neuropsychological disorders are, as a rule, the product of pathological events affecting the cerebral hemispheres. Most commonly, these are encountered in the setting of a recent stroke. Other potential causes include hemispheric tumors, head trauma, encephalitides, and neurodegenerative diseases.

Table 21.1. Psychological data

Vigilance *(quantitative assessment of awareness)*	State of awareness	Awake, sleepy, stuporous, comatose
Orientation *(qualitative assessment of awareness)*	Temporal	"What day of the week is this? What is today's date?"
	Locational	"What city is this? What is the name of this hospital? Do you know what floor this is?"
	Situational	"Why are you in the hospital? What caused you to come to me?"
	Personal	"What is your anniversary? What are the birth dates of your children?"
Mood	*(General emotional state)*	Depressed, sullen, irritated, euphoric, anxious
Affect	*(Affective state)*	Labile affect, emotional incontinence, emotionless
Memory	*(Short-term memory)*	"What did you have for breakfast this morning?"
Recall	*(Long-term memory)*	"What is the date of your anniversary?"
Attention Ability to concentrate		Stays on the subject, does not allow digression
Intelligence/train of thought	*(Intellectual capacities)*	Slowing, diminished ability to understand, oligophrenia (congenital), dementia (acquired)
Self-awareness/critical faculties		
Drive		Lacking drive, apathetic, hyperactivity, compulsively active
Thought content		Narrow, obsessed, delusional
Illusions/hallucination		Especially visual hallucinations with organic disorders

Aphasia

Definition

Aphasias are central nervous system (CNS) disorders that cause a change in or loss of the communicative use of language. Dysarthrias are different in that they are disorders of the motor functions of speech, i.e., problems of poor verbal articulation because of loss of motor control and/or of sensory feedback within the speaking apparatus.

Pearl

Careful attention to the quality of a patient's spontaneous speech, and tests in which he/she reads aloud or repeats a spoken work or phrase will bring out otherwise unapparent problems and will mostly allow rapid classification of the type or form of the aphasia. This includes having the patient repeat words or sentences, give names to various objects, and fulfill requests to carry out simple tasks (■ Tables 21.2 and 21.3).

Apraxia

Definition

Apraxia is a central disturbance of sequences of movement and motor behaviors, despite retained motor function and coordination, mostly associated with lesions in the parietal lobes (■ Table 21.4; see also Chap. 13).

Important Signs and Symptoms (Tabulated)

Meningismus

Definition

When testing the mobility of the cervical spine, the examiner has the patient lie passively supine while raising his or her head (flexing ventrally). Painful resistance to the flexion (stiff neck) is **meningismus**.

Table 21.2. Primary symptoms of aphasia

Symptom	Definition/description	Example
Neologisms	Making up or substituting new words	"Cutter" instead of "knife"
Agrammatism	Telegram style of speaking	"Yesterday . . . eat . . . home"
Paragrammatism	Defective sentence formation with dropping off, shortening, or doubling of sentence parts	"Today I was . . . it was bread that . . . and my wife . . . no, I mean . . ."
Phonic paraphrasia	Word or syllable transposition	"Roadrail" for "railroad"
Semantic paraphrasia	Confused word use	"Fork" rather than "spoon"
Cliché speech/stereotypes	Meaningless/repetitive expressions	"But certainly nevertheless" "My goodness, my goodness"
Amnesic aphasia	Missing words are replaced with circumlocutions	"That gizmo for cutting" rather than "knife"

Table 21.3. Types of aphasia

Form	Principal signs	Locus of damage
Motor (Broca)	Elevated speaking effort Agrammatism Phonic paraphrasia Understanding of language largely retained	Left frontal lobe
Sensory (Wernicke)	Fluent, spontaneous speech with no meaning Paragrammatism Phonetic and semantic paraphrasia Neologisms Marked difficulty with understanding language	Left temporal lobe
Global	Severe disruption of all expressive and receptive functions of speech	Extensive zones of damage in the areas supplied by the left middle cerebral artery
Amnestic	Principally anomia	Posterior temporal and parietal lobes

Table 21.4. Types of apraxia

Type	Frequency, everyday relevance, affected hemisphere	Principal signs
Ideomotor	Relatively frequent, but scarcely relevant in everyday life Language-dominant hemisphere	Substitution of parts of a movement by parapraxias (= faulty components of movement, e.g., in a military salute, using a fist to the brow, rather than an open hand)
Ideational	Relatively infrequent, relevant in everyday life Language-dominant hemisphere	Cannot carry out a common sequence of actions (e.g., opening a can or preparing coffee)
Constructional	Non–language-dominant hemisphere	Loss of creative acts that depend on visual control (e.g., forming a spatial image from its individual elements – e.g., drawing an image of a house)

Meningismus is associated with meningitides and subarachnoid hemorrhages. Lasègue's sign is a similar finding. When raising the supine patient's straightened leg (flexing the hip joint), increasing pain radiating into the leg can even restrict the range of movement. This finding is also associated with compressive lesions of the lumbosacral nerve roots caused by prolapsed intervertebral discs.

Clinical Testing of Cranial Nerve Function

Isolated deficits in the function of individual cranial nerve can by caused by inflammatory (e.g., borreliosis, or idiopathic hemifacial paralysis [■ Fig. 21.1]) or by space-occupying lesions like tumors or aneurysms. In addition, abrupt mononeuropathies of the cranial nerves that innervate the extraocular muscles is frequently encountered in patients with chronic metabolic disorders, such as diabetes (■ Table 21.5).

Pearl

Clinically it is important to differentiate peripheral mononeuropathies of cranial nerves from disorders of cranial nerves associated with failures of other brainstem systems.

In addition to the central nuclei and the proximal portions of cranial nerves III through XII, which are arranged in descending order through the brainstem (mesencephalon, pons, medulla oblongata), are other important neural structures, all crowded closely together. Numerous afferents, including the dorsal columns, the spinothalamic, and the spinocerebellar pathways that connect the brain and the cerebellum with input from the spinal cord and efferent pathways (especially the pyramidal tract), all pass through this region, where many of them decussate to the contralateral side. Control of eye movements is organized in the

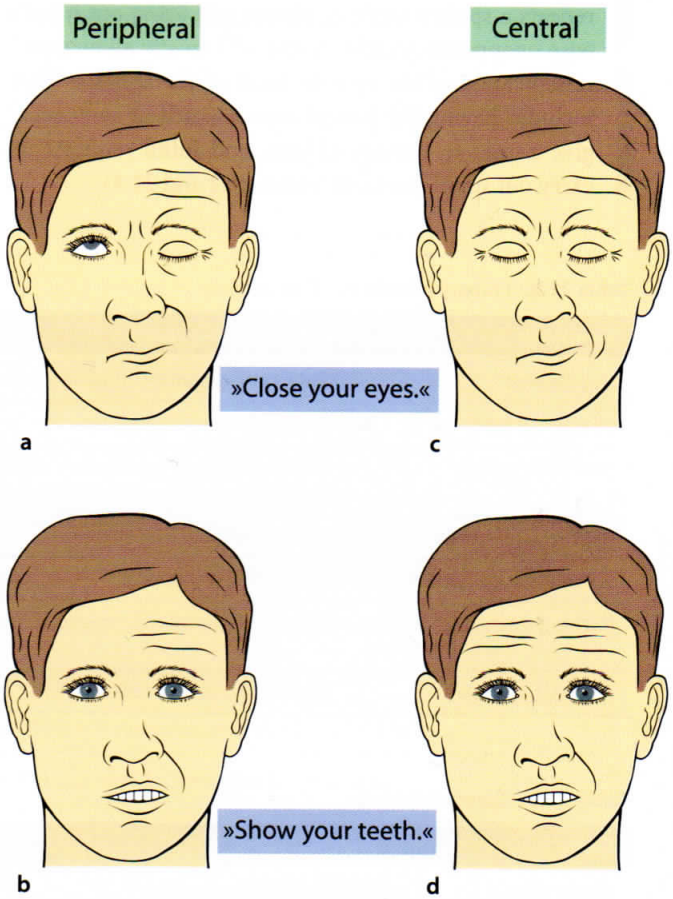


Fig. 21.1. Differential diagnosis of a facial palsy on the right side. **a**, **b** *Peripheral* facial palsy: **a** faulty lid closure with lagophthalmos and Bell's phenomenon; the patient is additionally asked to look forced upwards **b** paralysis of the perioral musculature in company with paralysis of the temporal branch of the facial nerve; **c**, **d** *central* facial palsy – **c** normal lid closure, **d** paresis of the perioral muscles and superior facial symmetry

Table 21.5. Clinical testing of the cranial nerves

Cranial nerve	Test	Pathological signs
I. Olfactory	Testing each nostril separately with eyes closed, using aromatic substances, e.g., vanilla extract or powdered coffee	Hyposmia, anosmia
II. Optic	See Chap. 8 Acuity, visual field (confrontation testing), fundus appearance (optic discs)	Reduced acuity, visual field defects, papilledema, optic disc pallor
III. Oculomotor IV. Trochlear VI. Abducens	See Chap. 10 Interpalpebral fissure, ocular motility (diplopia, oscillopsia), pupillary reactions (light/accommodation), look for nystagmus (positional and with head movement, using Frenzel's magnifying glasses)	Ocular muscle or gaze paresis, chooses eccentric gaze position, nystagmus, anisocoria
V. Trigeminal	Facial sensation, jaw muscles (open mouth against resistance, palpation of the masseters during clenching of teeth)	Peripheral ("isolated trigeminal branch") or central (onion peel dermatome) loss of sensation
VII. Facial	Facial muscles (furrow the brow, force eyes closed, turn up nose, pucker lips, blow cheeks open)	central (sparing the brow) or peripheral paresis (with Bell's phenomenon) See ■ Fig. 21.1
VIII. Acoustic	Monaural hearing tests with light rubbing of fingers together or whispering Tests of balance (nystagmus, standing, and gait tests)	Hypo-, hyper-, or anacusis
IX. Glossopharyngeal X. Vagus	Gag reflex, sensation of gums, and pharynx, swallowing Tests of palatal levators Hoarseness	Faulty swallowing, (unilateral) paralysis of palatal elevation, (pulling of the uvula towards the normal side), hoarse voice caused by paresis of the recurrent laryngeal nerve (a branch of the vagus nerve)
XI. Accessory	Head turning against resistance (sternocleidomastoids contralaterally innervated) and shoulder shrugging (superior trapezius) against resistance	
XII. Hypoglossal	Tongue protrusion	Unilateral paresis of the tongue (deviates to the healthy side) in peripheral disease and caudal brainstem disease, less so with supranuclear disease (cerebral lesions)

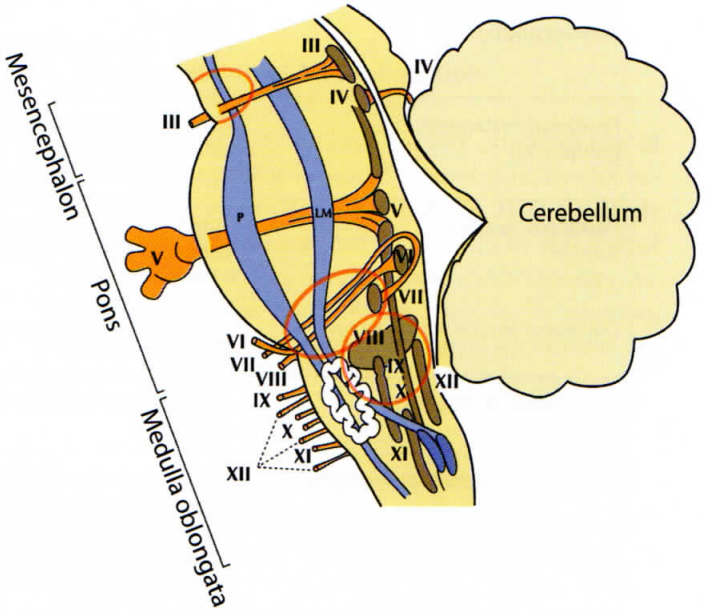


Fig. 21.2. Topographic summary of structures in the brainstem that are important for the localization of pathology: cranial nerve nuclei and cranial nerves (*Roman numerals*), the pyramidal tracts (*P*) (volitional motor function), the posterior columns (lemniscus medialis [*LM*]) and their nuclei (sensory afferents); cerebellar tracts have been left out to improve graphic depiction of other elements; the *red circled areas* are the disease locations for the most important brainstem syndromes (see Chap. 10)

midbrain and pons. In addition, important nuclei include the red nuclei in the midbrain and the olives in the medulla oblongata. Other important centers regulate consciousness, respiration, and cardiovascular circulation (■ Fig. 21.2). By analyzing the patterns of dysfunction discovered during the examination, it is possible to localize the site of a brainstem lesion with a high degree of accuracy. The most frequent causes of brainstem damage are demyelinating diseases (multiple sclerosis) in the young and ischemic vascular disease in the elderly (see Chap 10).

Nystagmus

Definition

Nystagmus is an involuntary, rhythmic oscillatory movement, usually of both eyes. In a jerk nystagmus, a slower drift in eye position is countered by sequential, intermittent saccadic movements in the opposite direction. By convention, the direction of a nystagmus is named according to the direction of the fast phase.

Physiological forms of nystagmus include positional (end gaze) nystagmus, vestibular nystagmus, and optokinetic nystagmus (OKN). Pathological types of nystagmus are most commonly the result of disorders of the brainstem or cerebellum. A simplifying rule states that nystagmus without vertigo has a central cause, whereas the type that is accompanied by vertigo is usually (though not always) caused by disease that lies outside of the CNS (■ Table 21.6; see Chap. 11).

Gaze Palsies

Definition

Gaze palsies are supranuclear disturbances of conjugate movements of both eyes.

This implies that the extraocular muscles and their peripheral innervation, including the cranial nerve nuclei, are intact. The most important causes of gaze palsies are cerebral infarcts, multiple sclerosis, tumors, Wernicke's encephalopathy, and neurodegenerative disorders, such as progressive supranuclear palsy (PSP) (■ Fig. 21.3, ■ Table 21.7; see also Chap. 11).

Table 21.6. Types of nystagmus

Spontaneous nystagmus	Congenital: equally fast phases (pendular)	Intensifies with fixation, no vertigo
	Peripheral: (labyrinthine disease, vestibular neuropathy)	Severe vertigo
	Central: (brainstem, cerebellum)	No vertigo
Periodic alternating nystagmus (PAN)	*Congenital or acquired, with disease of the vestibular nuclei or cerebellum. Regularly changes direction of fast phase, waxing and waning with a periodicity of about 120 s*	*No external influence*
Gaze-evoked nystagmus (present only when the direction of gaze is diverted from the primary position)	Physiologic (so-called end-gaze nystagmus)	Fatigue
	Pathologic: brainstem lesion (reticular formation), drug intoxication	Asymmetric Does not fatigue
Positional nystagmus (peripheral)	Benign paroxysmal or positional vertigo, closed head trauma	Following a rapid change in position wit a latency of several seconds, fatigues, produces vertigo
Positional nystagmus (central)	Tumors of the posterior fossa, cerebellar lesions, intoxications	Lateral position when lying, without latency and nonfatiguing, minimal or no vertigo
Downbeat or upbeat nystagmus	*By lesions in the medulla oblongata or cerebellum*	*Mostly with a dependent head position*
Gaze paretic nystagmus	Seen with partial (and even not visible) gaze palsy	
Dissociated (disjunctive) nystagmus	*Nystagmus predominantly in one eye in association with brainstem disease*	*Most common form is that seen in internuclear ophthalmoplegia*
Optokinetic nystagmus (OKN)	Physiologic, e.g., gazing out at passing landscape from a moving vehicle	Can be tested with an optokinetic drum or tape, pathological asymmetry found with lesions of the temporoparietooccipital region (Area 19) see ■ Fig. 13.1

Table 21.7. Gaze palsies

Form	Signs and symptoms	Site of damage	
Horizontal gaze palsy	Volitional gaze movement to the left or right is not possible	• Ocular motility centers of the frontal cortex (Brodmann area 8) • Corticonuclear pathways • Pons (paramedian pontine reticular formation or PPRF)	There is usually a transient period of conjugate deviation to the ipsilateral side (with cortical disease) or the contralateral side (with brainstem lesions)
Vertical gaze palsy	Volitional gaze movement up or down is not possible	• Midbrain	Note that restricted upgaze in the elderly is an aging phenomenon and is not usually pathological
Internuclear ophthalmoplegia (INO)	Paralysis of adduction of one eye with retained convergence (indicating a functional medial rectus muscle) and a simultaneous monocular abducting nystagmus of the contralateral eye (see ■ Fig. 21.3)	• Posterior medial longitudinal fasciculus (MLF) connecting the cranial nerve nuclei of the third and sixth cranial nerves (pontomesencephalic)	Frequently subtotal and evident as a visible slowing of saccadic velocity of the adducting eye Frequently bilateral
One-and-a-half syndrome	A combination of an INO and a gaze paresis to the ipsilateral side (see Chap. 11)	• Pons	

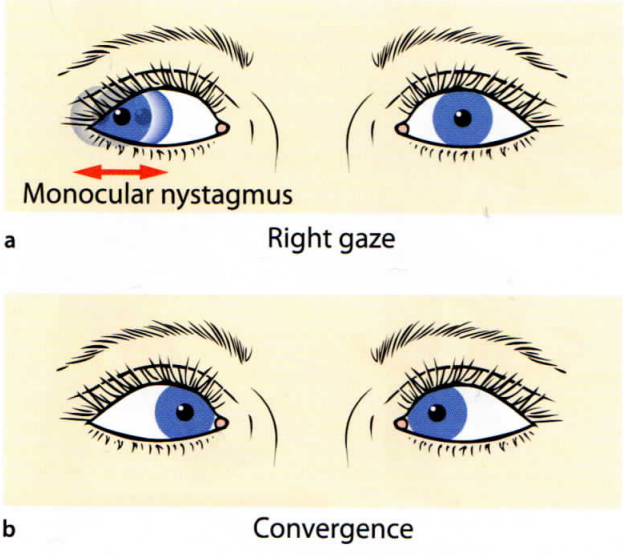


Fig. 21.3. Internuclear ophthalmoplegia (INO), left side. **a** Paresis of adduction of the left and monocular nystagmus of the right eye on gaze to the right side. **b** Normal adduction during accommodative convergence, proving that the medial rectus itself is unaffected

Dysarthria

Definition

Dysarthria is a disturbance of the motor control of speech, resulting in poor articulation.

In contradistinction to aphasia, which is categorized as a neuropsychological deficit (see above), dysarthria is a purely motor disturbance of the mechanics of speech. The understanding of language is unaffected, so communication is less severely affected by dysarthrias than it is by aphasias (■ Table 21.8).

Clinical Testing of Motor Function

Tests of arm and leg positional stability provide a way of quickly detecting the presence of a motor deficit, as for example, following a suspected stroke (■ Fig. 21.4). Relatively subtle motor disturbances can be detected by this sort of testing, although it does not contribute much to a differential diagnosis of the source of the problem. In addition, peripheral pareses of individual nerves or nerve roots can sometimes be missed.

Table 21.8. Dysarthrias

Form	Principal signs and symptoms	Locus of disease
Cortical	Poor articulation of consonants	Cerebral (e.g., following ischemic stroke)
Pseudobulbar	Poor consonants, monotone intonation, and slowing of speech	The cerebral cortex and the pathways to the cranial nerves (supranuclear, frequently associated with small vessel disease)
Bulbar	Muffled speech ("a lump in my throat") or nasal intonation of speech	Brainstem (e.g., ischemic disease), peripheral nerves (e.g., in amyotrophic lateral sclerosis (ALS), motor end plates (e.g., myasthenia), musculature (e.g., muscular dystrophy)
Extrapyramidal	Monotone intonation, soft voice	Basal ganglia or their connections (e.g., Parkinson's disease)
Cerebellar	Fragmented, irregular, blurting speech Inappropriate stressing of words, a raw and deep voice	Cerebellum (particularly with multiple sclerosis)

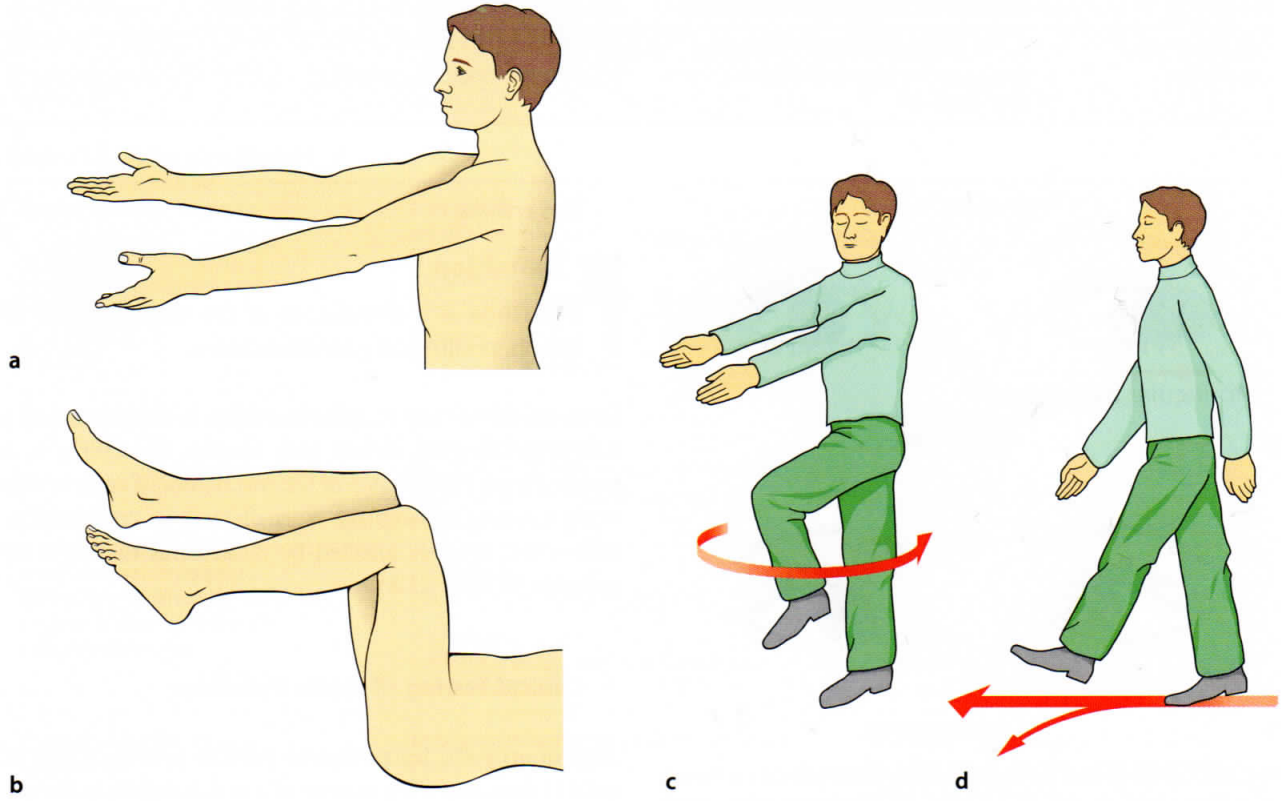


Fig. 21.4. **a** Downward drift and pronation of the left arm during an attempt to hold the limb in a steady and extended position. **b** Sinking of the left leg during an attempt to hold it in a steadily raised position. **c** Unterberger's test: The patient stands and "walks in place" with eyes closed. A fresh vestibular lesion is indicated when a body turn of more than 40° appears within 1–2 minutes (the patient turns in the direction opposite to that of the nystagmus). **d** Walking with closed eyes in a straight line results in a deviation to one side or the other: the side opposite to that of the nystagmus.

Pearl

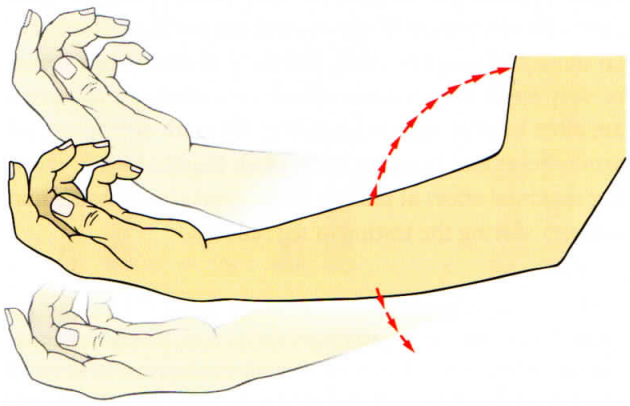
For a complete analysis of motor deficits, the positional stability tests should be augmented with attention to the signs of atrophy, the strengths of specific muscle groups, and the muscular tone of the neck and the extremities (■ Fig. 21.5, ■ Table 21.9).

Diagnostic conclusions about the location of damage can be drawn from the topical distribution of pareses. Therefore, for example, a hemiparesis immediately alerts the examiner to the presence of a central disorder (■ Fig. 21.6). Additional clues for distinguishing between central and peripheral pareses are listed in ■ Table 21.10.

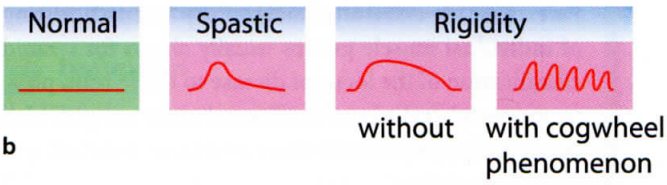
Video 21.1

Table 21.9. Examination of motor function

Inspection		
Signs of atrophy	Particularly in peripheral neuropathies and myopathies	
Movement unrest	Myokymias (quivering movements spread across broad muscle groups)	Frequently normal Often facial in patients with multiple sclerosis
	Fasciculations (brief contractions of individual muscle portions [motor units])	Sometimes normal, pronounced in ALS, spinal muscle atrophy, and poliomyelitis (including postpolio syndrome)
	Fibrillations (contractions of individual muscle fibers, very small, directly visible only in the tongue)	In ALS
Muscle tone	Passive movement of limbs during complete relaxation of muscles, especially those of the neck, elbows, wrists and knees (see ■ Fig. 21.5)	
Diminished	Sleep/hypotony	Acute central paresis Peripheral paresis Cerebellar disease
Elevated	Spasticity Elastic resistance to movement „clasped knife phenomenon"	Most central pareses
	Rigidity Constant resistance to movement, often with the "cogwheeling phenomenon"	Extrapyramidal motor disease
Muscular strength		
Stationary holding of arm position/ leg position	Detects mild or "latent" pareses when weakening is not readily apparent	See ■ Fig. 21.4
Severity of pareses	Degree of weakness: 0/5 = totally paralyzed to 5/5 = completely normal strength	Test the strength of the most important joint movements: The patient is asked to contract a certain muscle or muscle group with maximal force, while the examiner resists the movement
Diadochokinesia (ability to perform rapidly alternating movements)	A test of fine motor control; also detects mild pareses	Rapid alternating pronation and supination of the wrists, typing, replacing light bulbs
Hyperkinesias		
	Tremor, chorea, athetosis, ballismus, dystonia, tics, myoclonus	Largely CNS disease, extrapyramidal disease



a



b

Fig. 21.5 a Testing of the muscle tone around the elbow joint by passive manipulation during maximal relaxation of the muscles. **b** Diagram of important pathological findings the pocketknife phenomenon (fading of resistance when under steady pressure), and cogwheeling (joint rigidity with repeated, brief, and uniform increases in tonus over the entire range of motion of the joint) in case of muscle rigidity in Parkinson's disease

Table 21.10. The differential diagnosis of central and peripheral pareses

	Central pareses	Peripheral pareses
Fine motor control	Diminished	Largely unaffected
Muscular atrophies	None	Manifest
Tonus	Elevated spastically	Weakness
Deep tendon reflexes	Amplified (and possibly associated with clonus)	Weakened or extinguished
Pathological reflexes	Manifest	None
Coarse movements	Manifest	None

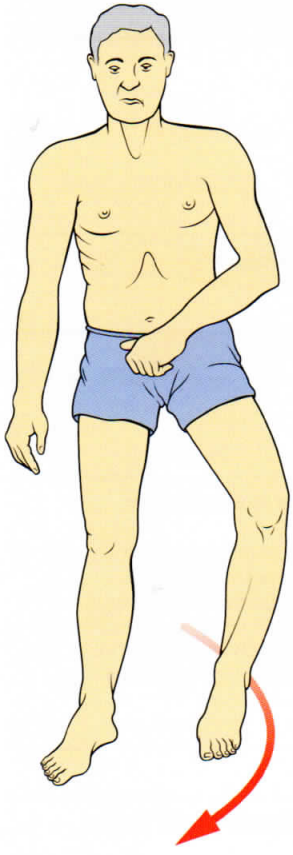


Fig. 21.6. Wernicke-Mann gait with hemiparesis of the left side following an infarct in the region supplied by the right middle cerebral artery: flexed, pronated, and adducted position of the left arm, extended left leg with plantar flexion and a partially supinated foot, forcing the patient's movement of the leg into an arc that swings out to the side

Pearl

Pareses of an extremity analyzed by strength testing of individual muscle groups usually allows for a rapid classification of the locus of disease to nerve root, plexus, or peripheral nerve. Additional clues are provided by accompanying disturbances of sensory function and testing of reflexes.

Clinical Testing of Sensory Function

When testing sensory function, the examiner has to rely on the attention and cooperation of the patient. A thorough testing of the most important sensory modes can be time-consuming (■ Tables 21.11 and 21.12).

Pearl

During a quick survey, or when examining patients that are only poorly cooperative, testing of pain sensation (with a needle or a toothpick) provides the most useful information.

From the topography and the quality of sensory disturbances, one can frequently reach an accurate conclusion as to the localization and etiology of the neural pathology. Typical examples are illustrated in ■ Fig. 21.7.

Clinical Testing of Reflexes

In general a minimal neurologic examination should include the most important arm and leg reflexes (■ Tables 21.13, 21.14, and 21.15), as well as testing of the presence of one of the Babinski signs. Deep tendon reflexes should be tested in positions that allow maximal relaxation of the involved muscle groups. This is most likely to be the case with the patient in the recumbent position. If muscular tension is fully excluded, and the patient still has absent or very weak deep tendon reflexes, facilitation maneuvers are often helpful, such as clenching the teeth during tests of arm reflexes and grasping both hands together while exerting maximal effort at pulling them apart (Jendrassik's maneuver), during the testing of leg reflexes.

Table 21.11. Esthesiometry (estimates of sensory function)

	Sensation	Method of examination	Pathological signs
External sensation	Touch	Stroke with fingertip, or even better use a small sable brush or wad of cotton	Hypesthesia Anesthesia Hyperesthesia
	Pain	Toothpick Disposable safety pin	Hypalgesia Analgesic Hyperalgesia
	Temperature	Test tubes filled with cold and warm (not hot!) water	Thermhypesthesia Thermanesthesia
Deep sensation	Position	With eyes closed, move limbs to the positions indicated by examiner's manipulation of the corresponding contralateral limbs	
	Motion	With eyes closed detect the direction of passive movements of distal interphalangeal joints	
	Vibration	Apply tuning fork to surfaces with palpable subcutaneous bone (0–8/8)	Pallhypesthesia Pallanesthesia
Combined	Stereognosis	With eyes closed identify small objects placed in the hands	Astereognosis

Table 21.12. Additional pathologic signs/symptoms of sensory systems

Dysesthesia	Sensory perception is unpleasantly altered, e.g., touch is felt as pain
Paresthesia	Unpleasant sensations (itching, burning, formication) that are spontaneous, or elicited by gentle surface stimuli
Hyperpathia	Sensations are amplified and are felt as unpleasant, e.g., hyperalgesia
Dissociated sensory loss	Isolated sensory modalities lost within a body part, with other sensory modalities retained

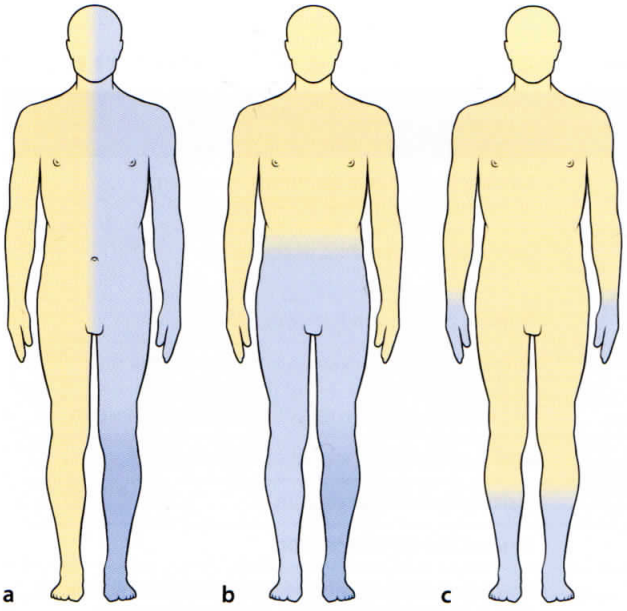


Fig. 21.7. Common patterns of sensory loss. **a** Unilateral loss of sensation with lesions of the contralateral cerebral hemisphere. **b** Loss of sensory perception in both legs with transverse damage to the spinal chord. **c** Glove-and-stocking pattern of sensory loss associated with a polyneuropathy

Deep tendon reflexes are variably elicitable under normal circumstances among patients in general. They can be quite weak in some patients, and lively in others. In order to identify correctly pathological weakening, care should be taken to compare one side to the other, to compare leg reflexes with those of the arm, or the arm and leg reflexes to those of the masseter. Pathological weakening of these reflexes should be diagnosed only when a clear and definite departure from normal can be detected.

Clinical Testing of Coordination – Ataxia

Definition

Disturbances of coordination, i.e., of smoothly cooperative alternation of agonists and antagonists during the course of complex movements like walking, are in general referred to as **ataxia**.

Table 21.13. Types of reflexes

Type	Neurophysiology	Example	Peripheral disease	Central disease
Deep tendon reflexes (DTRs)	Stimulus site and responding body part are in the same muscle, monosynaptic reflex arc, no adaptation	Achilles reflex	Diminished or extinguished	Amplified
Superficial reflexes	Receptors mostly in the skin, responses in neighboring musculature, polysynaptic reflex arc spread over several adjacent spinal dermatomes, adaptation is typical	Physiologic: abdominal reflexes	Weakened or extinguished (measured only by comparisons of symmetrical sites)	Weakened or extinguished (measured only by comparisons of symmetrical sites)
		Pathologic: Babinski reflex	Not elicitable	Elicitable

Table 21.14. Important reflexes of cranial nerve function

	Neurophysiology	Examination method	Significance
Masseter reflex	Monosynaptic muscular reflex Receptor and effector functions served by trigeminal nerve Synapse located in pons	Examiner places index finger transversely across slack-hanging jaw, striking the finger with a reflex hammer The response is a brief elevation of the mandible	Very brisk or amplified with supranuclear lesions affecting the corticopontine pathways (first motor neuron)
Corneal reflex	Oligosynaptic reflex, afferent path in the first (ophthalmic) division of the trigeminal nerve, efferent path via the facial nerve, synapses located in pontomedullary brainstem	Touch cornea with a cotton wisp that is not visible to the patient, response is a blink produced by a contraction of the orbicularis oculi	Attenuation caused specifically by lesions of the trigeminal or facial nerve or their central synaptic connection

Table 21.15. The most important intrinsic and extrinsic muscle reflexes

Reflex	Common abbreviation	Segment/peripheral nerve	Examination technique
Biceps tendon reflex	BTR	C6/musculocutaneous	Striking the index finger of the examiner, as it is held against the biceps tendon
Brachioradialis reflex (radioperiosteal reflex)	RPR	C6/radial	Striking the distal third of the radius
Triceps tendon reflex	TTR	C7/radial	Striking the tendon just above the olecranon
Trömner's reflex		C8/median > ulnar	Fingers 2–5 of the examiner quickly tap the tips of the flexed fingers 2–5 of the patient with his or her hand held in a relaxed, dorsiflexed position
Adductor reflex	ADR	L3–4/obturator	Striking the medial aspect of the knee joint
Patellar tendon reflex	PTR	L4/femoral	Striking the patellar tendon
Achilles tendon reflex	ATR	S1/tibial	Striking the Achilles tendon
Abdominal reflex	AR	Th7–12	Examiner uses a small wooden dowel or similar blunt object to quickly stroke the abdominal surface (from the lateral aspect towards the midline) in the superior, middle, and inferior thirds

Table 21.16. Tests of coordination

Test	Method	Pathological signs
Diadochokinesis	Screwing in a light bulb Rapid alternating pronation/supination at the wrist Playing piano/typing	Dysdiadochokinesis is found in pareses, lesions of the basal ganglia or cerebellum, and disturbances of deep sensation.
Finger-to-nose test Finger-to-finger test Heel-to-knee test	Look for: Smooth accuracy of movement Fluency of the movement Is there an intention tremor?	Dysmetria (see disturbances of cerebellar function)
Rebound movement	The patient (with eyes closed) presses with an extended arm with maximal upward force against the examiner's resistance. Cessation of resistance results in a quick response that prevents uncontrolled elevation of the patient's arm.	Abnormal rebound movement is found in patients with cerebellar dysfunction: The arm rises uncontrollably.
Bárány's pointing test	The patient lowers an extended arm, first with eyes open, then with eyes closed, until his/her index finger is aligned with that of the examiner's.	Poorly controlled movement with cerebellar or vestibular disease. Effect magnified with eyes closed, removing visual feedback control
Standing test, including Romberg	The patient stands with lowered, then with extended arms, and the test is repeated with eyes closed.	Unsteadiness (standing ataxia) with cerebellar disease Increasing with eyes closed, when there is also loss of sensory afferent control (loss of proprioception and touch sensations).
Unterberger step test ■ Fig. 21.4 c	Walk in place with arms extended and eyes closed (must be repeated several times)	Turning around the truncal axis (toward the diseased side) in patients with cerebellar or vestibular disease
Gait test ■ Fig. 21.4 d	Walking movements under various conditions of increasing difficulty (eyes closed, simulated tightrope walking, hopping on one leg)	Gait ataxia (e.g., in cerebellar disease) Small steps with faulty or absent associated movements like arm swing in patients with Parkinson's disease Wernicke-Mann gait after middle cerebral artery infarct

Table 21.17. Summary of signs and symptoms of cerebellar disease

Disorder	Clinical test or finding
Cerebellar dysarthria	See above, see ■ Table 21.8
Resting and gaze-evoked nystagmus	See above, see ■ Table 21.6
Intention tremor	In the finger-to-nose test, the tremor appears at the end of the movement just in front of the nose
Truncal, postural, and gait ataxia	Standing and walking test (■ Table 21.16)
Dysmetria	Poor judgment of targeted movements (pointing test; ■ Table 21.16)
Abnormal rebound movements	Hypermetric movements (■ Table 21.16)
Asynergy – dysdiadochokinesis	Faulty coordination of muscles for specific movements
Muscle flaccidity	See above (motor function)

Normal coordination depends on the simultaneous participation in the function of multiple neuronal systems (■ Table 21.16). Among these are the cerebral hemispheres, the cerebellum, the spinal, extrapyramidal, and vestibular systems, as well as the afferents and efferents of the peripheral nervous system.

Disturbances of Cerebellar Function

Cerebellar disorders are detected most sensitively with tests of coordination (■ Table 21.17). Important clinical signs of cerebellar dysfunction have been described in patients with multiple sclerosis with the so-called Charcot's triad: intention tremor, nystagmus, and scanning speech.

Conclusion

When visual changes are thought to be caused by a disease process affecting the nervous system, the ophthalmologist should, as a "non-neurologist," be able to elicit important clinical signs that will help to clarify the neurological diagnosis. This should help reduce the number of suspected disorders in the differential diagnosis, and by eliminating some possibilities, will facilitate the planning of further diagnostic procedures. This is especially important when assessing the level of urgency associated with the findings, and to participate in the strategic planning of the patient's further care.

Further Reading

Delank HW, Gehlen W (2006) Neurologie. Thieme, Stuttgart
Duus P (2001) Neurologisch-topische Diagnostik. Thieme, Stuttgart
Mumenthaler M (1993) Klinische Untersuchung und Analyse neurologischer Syndrome. Thieme, Stuttgart
Mumenthaler M (1997) Neurologische Differentialdiagnostik. Thieme, Stuttgart
Poeck K, Hacke W (2006) Neurologie. Springer, Berlin Heidelberg New York
Schenck E (1992) Neurologische Untersuchungsmethoden. Thieme, Stuttgart