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Patient Guide to Dizziness and Balance Disorders

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Preface

Dizziness and balance disorders are among the most common complaints of patients in all areas of medicine: one in three people suffers from them once or multiple times in their life. Fortunately, in most cases today, the cause can be identified and successfully treated. However, the reality often looks quite different: time and again, we see patients in specialized outpatient clinics who have undergone an odyssey of doctor visits before a diagnosis is made and effective therapy can be initiated.

The diagnosis is largely based on the symptoms the patient observes and what he/she reports to the physician. Key factors for diagnosis include the time course (episodes, acute onset, longer lasting or persisting) and triggers of the symptoms, as well as any accompanying symptoms. What does the physician do? Initially, the symptom profile is obtained through a systematic conversation. The subsequent physical examination is conducted with particular attention to balance function, eye movements, and hearing ability. Usually, only a few additional instrumental examinations are necessary to determine the cause. This should be carefully explained to the patient. Finally, the treatment and the time it takes until a meaningful improvement of the can be expected should be discussed in detail. What can patients do? In terms of therapy, it is important to carefully follow the physician's treatment recommendations and to keep a dizziness diary documenting the symptoms in order to assess the effectiveness of the therapy and to detect any possible adverse effects of the treatment. During follow-up medical appointments, the treatment is evaluated and adjusted if necessary.

This book provides a clear and comprehensible description of the most common dizziness disorders and the corresponding medical approaches. Additional typical cases, including both favorable and challenging courses, make the presentation particularly vivid. Furthermore, current scientific findings on the various causes of these disorders are also presented.

We wish the readers insight and even enjoyment as they read, all the more so as this will enable physicians to quickly access important information to promptly classify dizziness and balance disorders. Our many years of experience have shown that many patients often already suspect what they might be suffering from: we would like to support you in this. Finally, readers can also be of valuable assistance to relatives and acquaintances who suffer from dizziness and balance disorders.

For improved readability (and to simplify the language), only one gender form is used here, namely the masculine. All gender identities are always intended to be included.

In October 2025

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1

Introduction

Vertigo and dizziness are common complaints and a sensation, but not a disease in and of itself. For some people, vertigo or dizziness are a one-time experience, for others it is a frequent occurrence. However, these leading symptoms can mean something different for each affected individual. For some, it is merely a vague physical discomfort. Others use the term to describe an unusual, false perception of their surroundings, such as a spinning or swaying movement of the things around them. Still others experience a spinning or swaying sensation of their own body, usually in the head area, or a feeling of lightheadedness ("like one beer too many"). A subjectively perceived disturbance of spatial orientation is also possible: What is up, what is down? Where am I right now? This often makes it difficult for patients and doctors to correctly classify the symptoms. Fortunately in clinical practise, the type of these complaints is usually not that important for making the correct diagnosis.

However, the temporal course of the symptoms is important. Some forms of vertigo and dizziness cause persistent symptoms, others occur in episodes, and still others appear suddenly and last longer. In the latter case, caution is advised: it could also be a stroke. Finally, there are patients with a combination of these temporal patterns.

The possible triggers of these complaints, such as changes in position, as well as accompanying symptoms that may originate from the ear or from central brain regions such as the brainstem or cerebellum, are also important.

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This list already makes it clear: vertigo and dizziness can have a variety of underlying causes. There may be disorders within the body's balance system. The balance systems consist of the vestibular organ in the ear and the peripheral vestibular nerve, which runs from this organ to the brain, as well as the central balance centers in the brain. These central balance centers are located in the brainstem, cerebellum, and other regions of the brain. If acute damage occurs in central brain structures, such as a stroke, vertigo and dizziness can indicate a medical emergency. In medicine, the vestibular organ and vestibular nerve are referred to as peripheral vestibular, while the balance structures in the brain are called central vestibular.

In addition to disorders of the balance system—diseases of vertigo and dizziness in the narrower sense—disturbances in other organ systems can also cause sensations of dizziness. These include, for example, disorders of the visual system with double vision or blurred vision, as well as diseases of the peripheral nerves in the legs that are responsible for sensation in the feet and thus for the perception and control of standing and walking. Cardiovascular diseases, such as high or low blood pressure and cardiac arrhythmias, can also trigger sensations of dizziness.

Vertigo and dizziness do not always have to have a pathological background: although such sensations experienced during a car, ship, or carousel ride are referred to as motion sickness, the organic structures important for balance perception are healthy in this case. Rather, during "passively" experienced movement, there can be a "mismatch" of sensory stimuli from the eyes and the vestibular organ, bodily sensations, and the pre-programmed stimulus patterns stored in the brain. This can affect any of us and, depending on the intensity of the stimulus situation, trigger a sensation of dizziness. The same applies to visual height intolerance. With increasing height, those

affected experience a swaying dizziness, often accompanied by additional symptoms such as sweating, inner restlessness, palpitations, and fear of falling. In rare cases, these symptoms escalate into a fear of heights, an anxiety disorder that can be treated psychotherapeutically.

In terms of history, dizziness and vertigo are known as a symptom known since ancient times. In Greek, Roman, and Chinese antiquity, various authors mention these complaints in numerous passages, which—while caution is always advised when drawing such parallels—can, according to current knowledge, be assigned to some modern disorders, including dizziness occurring at heights, during sea voyages, or in connection with alcohol consumption. In ancient literature, diseases such as vestibular migraine (dizziness with headache, sensitivity to light and/or noise) or Menière's disease (dizziness with hearing loss, tinnitus, or a feeling of pressure in the ear) have already been described.

The vestibular organ in the ear was not described until the 19th century. It is located in an especially dense bone, the petrous bone, directly behind the middle ear, and consists of three fine, fluid-filled tubes called semicircular canals. These are arranged in the three spatial planes, that is, at 90-degree angles to each other (one canal is horizontal, the other two are

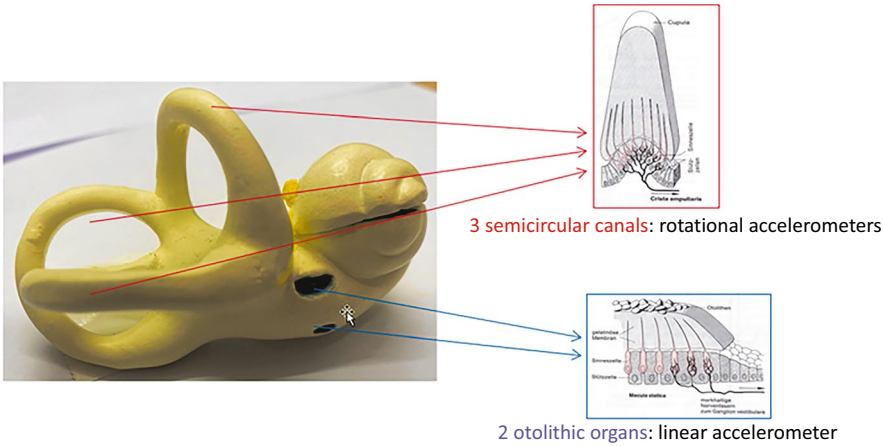

Fig. 1.1 Model of the vestibular organ. (Receptors from: Neurologisch-topische Diagnostik, Anatomie – Funktion – Klinik; Mathias Bähr, Michael Frotscher (eds.), 10th edition, Georg Thieme Verlag Stuttgart)

obliquely vertical). Sensory cells in each canal report the body's rotational movements in space to the brain, thereby ensuring bodily stability during movement (Fig. 1.1). The vestibular organ is filled with a fluid called endolymph. During head rotations, the fluid lags behind due to inertia, causing the sensory hairs in the canals to bend like aquatic plants in a stream. The second measuring system of the vestibular organ is the otoliths, gravity sensors that measure linear accelerations and gravity. In the two otolith organs, the utricle and saccule, calcite crystals rest on the surface of bundles of sensory hairs, as if small lumps were lying on the bristles of a brush.

The semicircular canals and otolith organs fulfill three important functions, disturbances of which can explain the unpleasant symptoms experienced by those with vertigo or dizziness. The first function is the already mentioned perception of position, movement, and orientation in space; the second function is gaze stabilization (i.e., keeping the eyes steady) during head movements. The third function is body control through balance reflexes (so-called vestibulospinal reflexes), in which the vestibular organ activates the appropriate muscles via the spinal cord to prevent increased swaying or a fall. Disease-related disturbances of this balance control lead to gait instability, deviation while walking, and a tendency to fall.

This is why acute dizziness often feels so unpleasant to those affected, sometimes even threatening, because it causes nausea and, in some cases,

vomiting. The reason for this is the close connections between the vestibular organ and the vomiting center in the brainstem. There are also nerve connections to brain structures involved in the anxiety system, which is why dizziness is often accompanied by anxiety and a sense of impending doom.

Adjacent to the vestibular organ is the cochlea (shown in the model in Fig. 1.1 on the right), which explains why some vestibular disorders are also accompanied by symptoms such as tinnitus, a feeling of pressure in the ear, and hearing loss, namely Menière's disease.

If vertigo or dizziness persist, a doctor should be consulted to determine its cause. The various underlying causes of the symptom are associated with different accompanying symptoms, or different triggering situations.

Patient Interview: The Medical History and What to Look For

A careful patient interview—the medical history—is the first and most important step in the medical examination:

- **Type of dizziness:** Is it vertigo, swaying dizziness, gait instability, tendency to fall, or lightheadedness?
- **Duration of dizziness:** How long has the patient been experiencing dizziness: days, weeks, or even years? For episodes of dizziness, the duration of each episode is important. Do they last seconds to a few minutes, many minutes to hours, or many hours to days? How often do episodes occur—several times a day or only very rarely?
- **Triggering factors:** When does the dizziness occur, e.g., with changes in head or body position, coughing, or straining? Do dizziness episodes occur in specific situations, such as in crowds at a shopping mall, on bridges, or at heights? Is the dizziness present at rest, does it occur spontaneously, during movement, after getting up, or while walking?
- **Accompanying symptoms:** Are there simultaneous symptoms originating from the inner ear, such as hearing loss, tinnitus, or a feeling of pressure in one ear? Is the dizziness accompanied by nausea and vomiting, headaches, sensitivity to light and noise?
- **Other relevant conditions:** Are there any heart or blood pressure disorders, or has diabetes been diagnosed? Is there a possibility of a potentially dangerous cause of dizziness originating from the brain that requires interdisciplinary diagnostics?

This detailed patient interview is the starting point of the medical examination and already allows the symptom of dizziness to be narrowed down relatively precisely in terms of its underlying cause. The next steps are the

clinical examination and various instrumental tests to assess the balance system. Taken together, these evaluations provide clues as to the location of the disorder: Is there a disturbance in the vestibular organ or in the brain, or is it due to other causes?

Affected individuals, family members, and professionals such as physiotherapists or psychotherapists who come into contact with people suffering from dizziness will find in this guide an overview of the most important questions about dizziness. The aim is not to replace the conversation with a doctor, but to provide starting points and information for classifying dizziness, for dealing with it, and for understanding its possible consequences. The guide is intended to outline both the course of dizziness disorders and available support options.

The guide is structured as follows:

The **General Section** describes the physical examination, focusing on key aspects for the diagnostic classification of dizziness. In addition, general treatment principles are presented.

The **Special Section** contains individual chapters and sections on the ten most common causes of dizziness, their symptoms, physical and instrumental findings, therapy, and the course with and without treatment. Central and peripheral causes are distinguished.

Overview

Central causes (brain):

- Stroke
- Vestibular migraine
- Cerebellar diseases or diseases with movement disorders, such as Parkinson's disease
- Functional dizziness
- Other causes such as blood pressure regulation disorders, adverse drug effects, diseases of the peripheral nerves

Peripheral causes (ear):

- Peripheral positional vertigo
- Acute unilateral loss of a vestibular organ
- Menière's disease
- Bilateral vestibulopathy
- Vestibular paroxysmia
- Rare causes such as the third mobile window syndrome

Table 1.1 Frequencies of various dizziness syndromes in 47,748 patients at a specialized outpatient clinic

Diagnosis	Frequency	
	n	%
1. Functional dizziness	8121	17.1
2. Vestibular migraine	6407	13.4
3. Peripheral positional vertigo	6299	13.2
4. Central vestibular dizziness	6044	12.7
5. Menière's disease	4799	10.1
6. Unilateral vestibulopathy	4237	8.9
7. Bilateral vestibulopathy	2966	6.2
8. Vestibular paroxysmia	1389	2.9
9. Third mobile window syndrome	207	0.4
10. Unclear dizziness syndromes	2131	4.5
Other	5133	10.8
Total	**47,748**	

Table 1.1 shows a statistical survey of the ten most common dizziness syndromes among more than 47,000 patients at the German Center for Vertigo and Balance Disorders (DSGZ) and the Neurology Clinic of LMU Munich. They account for 80–90% of diagnoses and are described in this guide.

2

The Physical Examination

The symptoms of vertigo and dizziness are highly diverse and can be accompanied by various other disturbances such as gait disorders, tendency to fall, double vision, or oscillopsia. Therefore, in addition to a thorough medical history, a physical neurological examination by a physician is very important for accurate classification. During this examination, deviations in various neurological systems are assessed, e.g., abnormalities in eye movements, motor function, and coordination. Furthermore, some instrumental tests to assess the balance system, including eye movements, can be important for diagnosis. Taken together with the medical history, these examinations provide clues to the location of the disorder—whether it is a peripheral (in the ear) or central (in the brain) vestibular disorder, or whether the cause lies in other sensory systems—as well as to the underlying disease responsible for the symptoms.

2.1 Examination of Eye Movements

Since disturbances in the balance system are often associated with disorders of eye movements and also with involuntary, jerky eye movements (nystagmus), testing eye movements is a central component of the neurological examination. This examination is performed by the physician and if needed by specially trained orthoptists. Involuntary eye movements can also be observed in people who are looking out at the landscape from a moving train and fixate on the next point of interest with jerky eye movements. This

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is called optokinetic nystagmus, which is not pathological; rather, its absence would be considered abnormal. In addition to **visual acuity**, the subjective visual vertical is also examined, i.e., whether the patient perceives an objectively vertical object as vertical or as tilted.

The **subjective visual vertical** can be measured very simply using the so-called bucket test, a sensitive test that indicates an acute unilateral peripheral or central disorder in the balance system (Fig. 2.1). The patient looks into a bucket with a movable rod at the bottom, which they are to align vertically after the examiner rotates the bucket. The examiner can read off by how many degrees the patient's perception of vertical deviates from the objective vertical. The deviation should not exceed 2.5°.

During the examination of eye movements, **abnormal eye movements** are precisely measured. It is tested whether the eyes are aligned parallel or, for example, whether the visual axis of one eye is above that of the other. In the cover test, after covering one eye, vertical adjustment movements of the uncovered eye occur—a sign often found in brainstem disorders. The range of eye movements is also examined by having the patient follow a pen in eight directions (horizontal, vertical, oblique up and down). In this way, dysfunction of eye muscles or their nerves can be detected. In addition to these peripheral palsies, there are also central gaze palsies, for example, when both eyes can no longer move upward, as can occur in certain metabolic diseases.

In addition, attention is paid to **involuntary eye movements**. When the patient looks straight ahead, a **nystagmus** may occur in certain vestibular disorders. This spontaneous nystagmus is examined without or with the aid of Frenzel or M-glasses, which, through special lenses, prevent fixation on an

Fig. 2.1 Measurement of the subjective visual vertical using the bucket test

object. In diseases of the vestibular organ or vestibular nerves, spontaneous nystagmus increases in intensity under the glasses. If this is not the case, it suggests a disorder of the central balance system in the brainstem or cerebellum. Depending on the disease, eye movements may be sawtooth-like (jerk nystagmus) or pendular (pendular nystagmus). Nystagmus can beat horizontally, vertically downward or upward (downbeat, upbeat nystagmus). It is also checked whether nystagmus occurs after repeated head shaking (provocation nystagmus).

The examination of eye movements also includes the assessment of **slow pursuit movements**. It is observed whether these are performed smoothly or whether a saccadic, i.e., jerky, pursuit movement occurs. This can have various causes, such as central vestibular dysfunction; it can also be a side effect of certain medications or age-related. Furthermore, **vergence and convergence reactions** are tested. The patient fixates on an object moving toward the eye, and the physician checks whether both eyes move inward. The examination of **saccades** (rapid eye movements; Fig. 2.2a) is performed by having the patient quickly look back and forth between two targets held by the examiner about 80 cm in front of the patient. The accuracy and speed of the saccades are observed. This can also be assessed using the optokinetic drum. Speed is tested with the optokinetic drum (Fig. 2.2b). Here, a rotating drum with stripes or figures is placed in front of the patient's eyes, which they are to follow. If the previously mentioned nystagmus occurs, as can be observed when looking out of a moving train, the finding is normal. If this nystagmus is absent or shows a direction other than expected, this is considered pathological.

Examination of the function of the organs of balance in the ear. Another important examination is the **head impulse test**. This assesses the vestibulo-ocular reflex, which enables precise fixation on a point during rapid

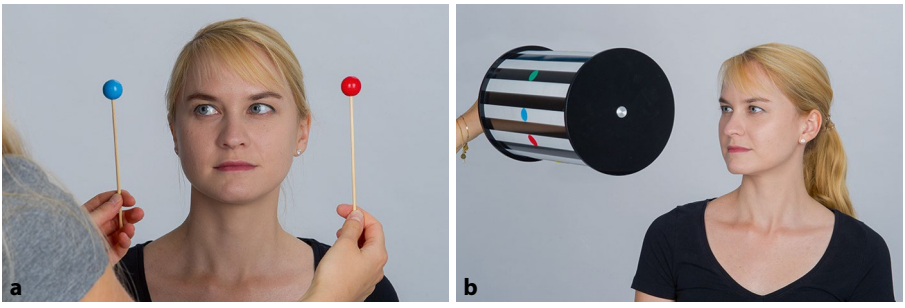

Fig. 2.2 **a** Examination of saccades, **b** Examination using the optokinetic drum

head movements, allowing us to see clearly despite head motion. Stabilization of gaze during head movements is an important function of the vestibular organ. A head movement to the right triggers an equally large eye movement to the left, so that when walking or running we can see faces and read signs instead of seeing blurred images. In this test, the examiner moves the patient's head quickly to the right and left while the patient fixates on a central point. The examiner observes whether the eyes quickly return to the central point after head rotation or whether corrective eye movements are needed.

Examination for a peripheral paroxysmal positional vertigo. Balance testing also includes **positional maneuvers** (Fig. 2.3). These have to be performed in every patients and are particularly important when the patient complains of recurrent short episodes of spinning vertigo triggered by head or body movements. The physician tests this positional vertigo by positioning the patient on the examination table in various directions and observing the eye movements using special glasses (Frenzel/M-glasses) that prevent the patient from fixing their gaze. If the posterior semicircular canal is affected by positional vertigo, the examiner sees rotatory eye movements after positioning to the side following a 45° head turn, which stop after a maximum of one minute. The patient experiences a spinning sensation. For the horizontal semicircular canals, positional tests are performed in the supine position. The

Fig. 2.3 Examination for peripheral paroxysmal positional nystagmus

head is turned 60 to 90° to the right and left. Here, the nystagmus observed by the physician, which beats horizontally, may persist longer.

2.2 Examination of Gait and Posture

Since the vestibular organ is connected to the leg muscles via pathways in the spinal cord (vestibulospinal reflexes), dizziness is often accompanied by unsteadiness of gait and posture as well as a tendency to fall. Testing the ability to stand and walk is therefore another important part of the physical examination.

Postural stability is assessed using the Romberg test under various conditions with different levels of difficulty: feet apart, feet together, tandem stance, standing on one leg. Each condition is tested with eyes open and closed. If the patient sways significantly more with eyes closed, this suggests a sensory disturbance (impairment of sensation) of the vestibular system or the somatosensory system, e.g., in diseases of the leg nerves (polyneuropathy). The direction of the tendency to fall during the examination—unilateral, forward, backward—provides the physician with clues to the underlying disorder. In cases of dizziness not caused by organic disease but by psychosomatic factors, standing improves with distraction, e.g., by writing numbers on the patient's back with the finger.

Gait is also tested under various conditions: walking with eyes open and closed at self-selected speed, walking along an imaginary line, turning, walking with eyes open and closed while performing additional tasks, e.g., counting backwards. The examiner observes gait speed, stride length, gait rhythm, consistency of steps, base of support, posture, and symmetry of gait, as well as arm swing and the number of intermediate steps when turning. Here too, by considering the various possible gait abnormalities, conclusions can be drawn about the underlying disease.

2.3 Examination of Coordination and Motor Function

In addition to these examinations of the balance system, eye movements, posture, and gait, the physical examination includes a complete neurological assessment of coordination—that is, dexterity and fine motor skills of the hands and trunk stability—as well as motor function and sensation. In

particular, the examination of coordination provides information about the functionality of the cerebellum, a brain structure that is very important for balance.

Coordination is assessed by having the patient perform fine finger movements, follow the examiner's rapidly moving finger, touch the nose with the index finger with eyes closed, and slide the heel down the opposite shin. Each of these is tested on both the right and left sides. The examiner observes whether these movements are smooth and targeted or jerky and overshooting, and whether tremor occurs during movement. Trunk stability is assessed while sitting.

The examination of **motor function** includes strength testing of the arm and leg muscles and assessment of muscle tone. It is checked whether involuntary tremor of the arms or legs occurs during certain movements. In addition, the deep tendon reflexes of the arms and legs are tested. Pathological reflexes (e.g., the Babinski reflex) should not be present. In the **sensory examination**, sensation of touch (sharp and dull), pain, and temperature are tested on the arms and legs, comparing both sides of the body. Vibration sense is tested with a tuning fork applied to the ankles and wrists. In diseases of the leg nerves, vibration sense is usually reduced.

2.4 Instrumental Examinations

In addition to systematic history taking and physical examination, instrumental diagnostic testing is the third important pillar of diagnosis, but not needed in every patient. Instrumental laboratory tests serve to precisely determine and document functional deficits in the balance system, as well as to monitor the course of the disease and the effects of therapy. Depending on the clinical findings, different methods of balance testing are used, which are non-invasive and painless.

As already mentioned, the vestibulo-ocular reflex is examined using the head impulse test. Sometimes, small corrective eye movements after head rotation, which can indicate a disorder of the vestibular organ, are missed during clinical examination. Therefore, this test is also performed instrumentally by recording eye and head velocities with a video camera (**video head impulse test**; Fig. 2.4). A specific velocity ratio between eye and head movement is required. If the eye movement is too slow or the eye must make very small corrective movements to fixate the point, this indicates a disorder of one or both vestibular organs, depending on whether the test is abnormal on one or both sides.

Fig. 2.4 Video head impulse test. The function of the vestibular organs can be measured very quickly using rapid head rotations and simultaneous measurement of the resulting eye movements

Fig. 2.5 Caloric testing. Irrigation of the ears with warm and cool water to assess the function of the vestibular organs

Another test also assesses the vestibular organ: the **caloric test** (Fig. 2.5). By irrigating the ear canal with warm and cold water, the fluid in the semicircular canals moves up or down. This triggers a stimulus similar to a head movement, resulting in rapid eye movements (nystagmus). These are

recorded with an eye camera and quantitatively compared between sides. In this way, it is assessed whether the vestibular organs are functioning or not.

The **subjective visual vertical** can also be tested instrumentally in addition to the bucket test (see Sect. 2.1 and Fig. 2.1). In this test, the patient sits in front of a hemispheric dome larger than their visual field, preventing orientation to external structures (Fig. 2.6). At the back of the hemisphere is a rod that the patient is to align vertically in several trials. Again, only a deviation up to 2.5° is considered normal. In addition, **eye position is determined using a fundus camera**. The patient looks into a device that examines and measures the fundus of the eye. This allows determination of whether the fundus, and thus the eyes, are rotated—a sign of a peripheral or central vestibular dysfunction.

In addition to clinical examination, eye movements can be precisely measured using an eye camera with **video-oculography**. Especially for monitoring the course of the disease, including during therapy, this quantitative oculomotor measurement is an important examination. The presence of spontaneous nystagmus is tested. In addition, pursuit, gaze holding, saccades, and optokinetic nystagmus are quantitatively assessed.

Fig. 2.6 Instrumental measurement of the subjective visual vertical using a hemispheric dome and light dots to measure perceived verticality

Fig. 2.7 The function of the otolith organs, which are important for linear movements and the perception of gravity, can be assessed by measuring vestibular evoked potentials

Another test of vestibular organ function, specifically of the otolith organs within it, is the measurement of **vestibular evoked cervical and ocular myogenic potentials** (cVEMP, oVEMP; Fig. 2.7). The otolith organs are involved in various reflex arcs via nerve impulses, which can be measured. When the otolith organs are stimulated, e.g., with a small vibrator on the forehead (utricle reflex arc) or by a click sound in the ear (saccule reflex arc), the nerve signals arriving at certain muscles can be measured: cervical evoked potentials at the neck and ocular evoked potentials at the eyes. In certain vestibular disorders, these may be increased or decreased.

Examination using **posturography** (Fig. 2.8) enables objective testing of postural and balance regulation. Body sway can be measured under various conditions, for example, standing with eyes open or closed, standing on firm ground or foam, and standing with the head tilted back. Using artificial intelligence, i.e., analysis with a neural network that compares sway patterns with many stored patient data, the sway parameters measured in the patient can be assigned to specific diseases, e.g., a disorder of the vestibular organ, cerebellar dysfunction, or functional dizziness.

Quantitative gait analysis (Fig. 2.9) enables the diagnosis of gait disorders by examining gait on a gait analysis mat. The patient walks at various speeds (self-selected, slow, fast), with eyes open or closed, with changes in head position, or while shifting concentration, across the gait carpet. Stride length, speed, base of support, and gait variability are automatically analyzed

Fig. 2.8 Posturography for measuring postural regulation under various conditions, here standing on foam with eyes open and closed

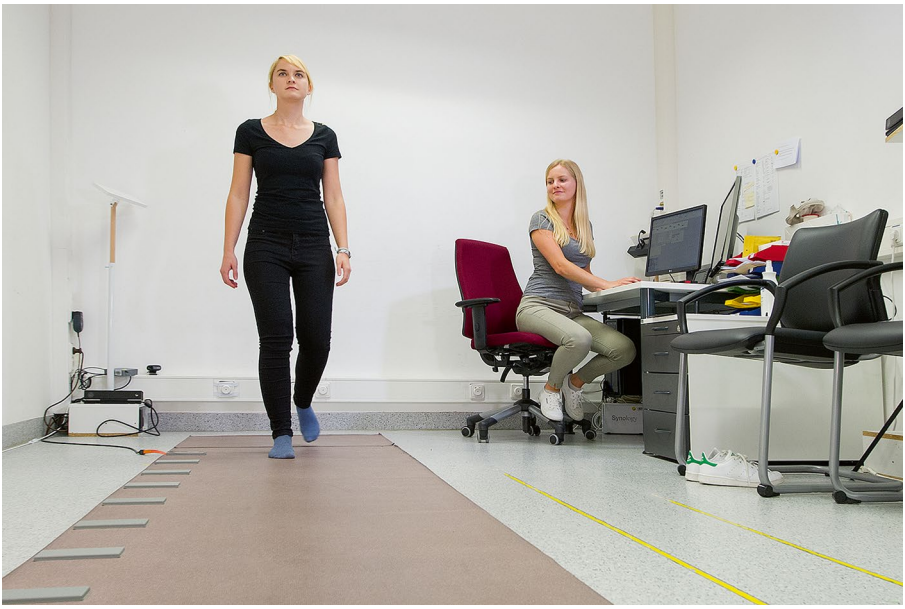

Fig. 2.9 Gait analysis for precise assessment of walking ability, e.g., with eyes open and closed

using a neural network and compared with patterns from patients with diseases associated with gait abnormalities. This allows the gait pattern to be assigned with high accuracy to one of several causes.

In addition to these instrumental balance examinations in the narrower sense, further supplementary tests may be appropriate depending on clinical and instrumental findings. These include assessment of hearing using **pure tone audiometry** as well as imaging techniques: **high-resolution computed tomography** of the temporal bone or **magnetic resonance imaging** of the brain with particular attention to the temporal bone as well as brainstem and cerebellar regions. In some cases, a **lumbar puncture** (cerebrospinal fluid examination) and **genetic testing** may also be necessary.

3

What Matters in Therapy: General Treatment Principles

The various underlying conditions and the differing courses of disease lead, depending on the cause, to physiotherapeutic, pharmacological, psychotherapeutic, or, very rarely, surgical treatment options.

In every case, the foundation of therapy is thorough education about the mechanisms underlying the symptom, the disease itself, its progression, and the time it takes until treatment usually leads to for the patient noticeable improvement. Most vertigo syndromes have a favorable spontaneous course or can be successfully treated, thus offering a good prognosis. It is advisable to keep a vertigo diary in which the affected individual records episodes of vertigo according to type, severity, duration, accompanying symptoms, and triggering situations. This is particularly important for establishing the diagnosis and for monitoring the effectiveness of therapy.

3.1 Physiotherapeutic Treatment

Balance training is an essential component of therapy for many peripheral and central vertigo syndromes. Deficits in the vestibular system can thus be compensated for more effectively, as the brain learns to better cope with the new situation—such as the loss of function in one vestibular organ—and to suppress the sensation of vertigo. Regular balance exercises also improve disorders of the visual system or leg nerves. Special exercises are performed for

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the balance system, eye movements, and sensory system, i.e., for feedback from the leg nerves. These are carried out under the supervision of a physiotherapist and should also be performed independently on a daily basis. Ideally, balance should be practiced several times a day for 10 to 15 minutes under varying conditions. During these exercises, the head is rotated in all three spatial planes while a visual target is fixed. In addition dynamic exercises with head movements while walking, and various walking and standing exercises—such as tandem gait, cross-step walking, single-leg stance, etc.—are performed. It is important to select the difficulty level of an exercise so that the patient is challenged but not overwhelmed. For balance exercises, this means, for example, that the patient should hold on as much as necessary but as little as possible. Safety must always be ensured.

For peripheral paroxysmal positional vertigo (Sect. 4.1.1), repositioning maneuvers are the only effective therapy. These are performed in a very specific manner (Fig. 3.1). After medical instruction, they can be performed independently. Positional therapy under physiotherapeutic supervision is also possible (see Sect. 4.1.1).

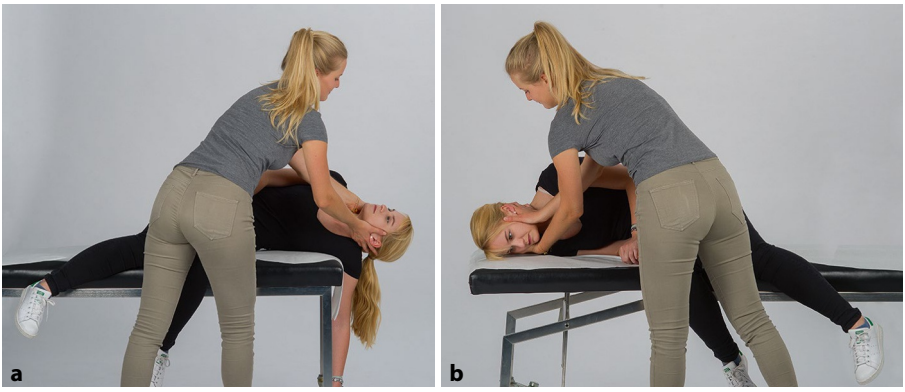

Fig. 3.1 Therapeutic positioning maneuvers, here the Semont maneuver, for peripheral paroxysmal positional vertigo of the left posterior semicircular canal: First, the patient's head is turned 45° toward the unaffected right side; then the patient is laid down 90° toward the affected left side (**a**). After one minute, the patient is then moved about 190° to the unaffected right side (**b**). Important: The head must always remain turned 45° toward the unaffected side. After another minute, the patient sits up again (not shown). These exercises should be performed three times in the morning, three times at midday, and three times in the evening until symptoms resolve, i.e. no positional vertigo can be triggered in three consecutive mornings

3.2 Pharmacological Therapy

Medications are an important component in the treatment of vertigo syndromes. There are medications that are intended to alleviate accompanying nausea or vomiting in acute vertigo disorders: antiemetics. This **symptomatic treatment** should not be continued for more than three days. Sometimes such treatment is necessary to allow physiotherapeutic positioning maneuvers to be performed without nausea. Relevant preparations include dimenhydrinate, ondansetron, and metoclopramide. Benzodiazepines can also be taken for severe vertigo, but they cause drowsiness and unsteady gait. In addition, there is a risk of medication dependence with prolonged use. To prevent motion sickness, scopolamine transdermal patches can be applied behind the ear before travel. However, this preparation also causes drowsiness.

In addition to this symptomatic treatment, there is, depending on the cause of vertigo, a **specific** or **causal pharmacological treatment** (see the respective sections on the disease). Cortisone (glucocorticoids) improves recovery of peripheral vestibular function in unilateral loss of vestibular organ function (see Sect. 4.1.1). Seizure suppressing medications such as lacosamide are used when vertigo is caused by close contact between the vestibular nerve and a small arterial vessel (vestibular paroxysmia; see Sect. 4.1.5). Betahistine is used to prevent vertigo episodes in Menière's disease, a disorder of the inner ear with endolymphatic hydrops (see Sect. 4.1.3). For prevention in vestibular migraine (see Sect. 4.2.2), beta-blockers, calcitonin gene-related peptide (CGRP) antibodies, or antidepressants are used, the latter also for the treatment of functional dizziness (see Sect. 4.3). For oculomotor disorders and cerebellar dysfunction (see Sect. 4.2.3), there are various preparations whose efficacy has only been increasingly studied in recent years, including 4-aminopyridine or acetyl-L-leucine. For downbeat nystagmus and episodic ataxia type 2 (EA 2) 4-aminopyridine is used, and if ineffective in EA 2, acetazolamide is administered (see Sect. 4.1.6).

3.3 Psychotherapeutic Treatment Methods

For the frequently occurring functional dizziness (see Sect. 4.3), if other measures are not effective (explaining the disease to the patient, desensitization by self-exposure, regular sports (but no specific vestibular exercises), or

SSRI), psychotherapeutic treatment is a method of choice. Therapy includes psychoeducational approaches, in which the development and mechanism of the disorder as well as specific behavioral instructions are explained, such as engaging in regular physical activity. In addition, cognitive behavioral therapy can be performed, for example with desensitization, in which triggering situations are deliberately confronted, initially in a low-threshold and then in a more intensive manner, instead of being avoided. Behavioral therapy methods should also be used for vertigo accompanied by additional psychological and psychiatric disorders.

3.4 Surgical Treatment

Rarely, the cause of vertigo is a benign tumor of the vestibular nerve (vestibular schwannoma, formerly known as acoustic neuroma). The tumor is treated surgically or by targeted radiation (Gamma Knife/Cyberknife). In vestibular paroxysmia with close contact between the vestibular nerve and a small arterial vessel (see Sect. 4.1.5), which leads to pulsations and irritation of the nerve, surgery is only required if the affected side can be reliably identified and the patient responds convincingly to medication but cannot tolerate it. In this case, a synthetic material (interposition) is inserted. The third mobile window syndrome with insufficient bony coverage of the semicircular canal (see Sect. 4.1.6) requires surgical treatment in certain cases, usually involving closure of the affected semicircular canal.

4

The Most Common Causes of Vertigo, Dizziness, Balance, and Gait Disorders

The spectrum of possible causes of vertigo, dizziness, balance disorders, and gait disturbances is broad, yet manageable. Many medical specialties, from general medicine to neurology, otorhinolaryngology, and even psychiatry, deal with these common complaints.

After familiarizing yourself in the previous chapters with the history-taking and physical examination of patients with vertigo, dizziness and balance disorders, we now present the most important and common clinical syndromes that may be associated with these leading symptoms.

A fundamental classification is made into disorders that

1. originate from the inner ear and the vestibular nerves: peripheral vertigo and dizziness (see Sect. 4.1).
2. originate from the brainstem or cerebellum: central vertigo and dizziness (see Sect. 4.2).
3. are caused by increased self-perception of balance: functional dizziness (see Sect. 4.3).
4. occur as sequelae of other diseases, e.g., internal medicine conditions (see Sect. 4.4).

Peripheral vertigo has various causes. These include acute unilateral loss of function of a vestibular organ, peripheral positional vertigo caused

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by dislodged crystals that move back and forth in the semicircular canals, Menière's disease, and the usually insidious onset of bilateral loss of vestibular organs (bilateral vestibulopathy). Rare forms include lightning-like, spontaneously occurring vertigo episodes lasting seconds (vestibular paroxysmia), as well as disorders in which symptoms are triggered by pressure changes or loud sounds (syndrome of the third mobile windows).

The most important **central vestibular disorders** include stroke, vestibular migraine, and various forms of cerebellar and so-called basal ganglia diseases (such as Parkinson's disease). The latter are usually characterized by persistent swaying dizziness and gait disturbances.

The most common cause of all is **functional dizziness**. This usually presents as persistent swaying or lightheaded dizziness. Sometimes, an organic disease precedes as a trigger.

Other conditions that can lead to vertigo and dizziness include, for example, adverse drug effects, disturbances in blood pressure regulation, movement disorders, or diseases of the peripheral nerves (polyneuropathies), typically presenting with swaying dizziness and gait instability.

For all these clinical syndromes, typical case examples and video sequences are presented at the outset, followed by a clear and realistic description of the key steps in diagnosis, therapy, and prognosis.

4.1 Vertigo and dizziness originating from the vestibular organs or vestibular nerves: What to look out for

Vertigo and dizziness originating from the vestibular organs or vestibular nerves can be classified based on the symptoms and examination findings of affected patients into three groups: recurrent episodes, sudden-onset with prolonged symptoms, and persistent symptoms (details below). As a patient, you should observe the typical symptoms, their duration, triggers, and accompanying symptoms. In this way, you can help the physician quickly make the correct diagnosis and initiate effective treatment.

The temporal course is particularly important:

1. Recurrent episodes: The duration of episodes can range from seconds to minutes ormany hours. Pay attention to the minimum and maximum duration so you can inform your doctor. If the episodes last

- less than a minute, this may indicate peripheral paroxysmal positional vertigo (PPPV) (see Sect. 4.1.1) or a disorder in which a blood vessel compresses the vestibular nerve (vestibular paroxysmia; see Sect. 4.1.5).
- many minutes, Menière's disease may be the cause (see Sect. 4.1.3).

2. Sudden-onset, prolonged, severe spinning vertigo: If this originates from the vestibular nerve, the most likely cause is inflammation of the vestibular nerve (acute unilateral vestibulopathy/vestibular neuritis; see Sect. 4.1.2).
3. Persistent swaying dizziness with gait disturbances, usually developing slowly over weeks to months: If this originates from the vestibular organs in the ear, it is usually due to bilateral reduction or even loss of function of these organs (bilateral vestibulopathy; see Sect. 4.1.4).

Furthermore, you should note whether the symptoms occur out of the blue or whether there are factors that trigger, worsen, or alleviate them.

1. Occurrence without a trigger is typical for Menière's disease, unilateral loss of a vestibular organ, or vestibular paroxysmia.
2. Triggers can include:

- Changes in head or body position; these are typical for peripheral paroxysmal positional vertigo (PPPV)
- Movements while walking, with vertigo worsening in the dark or on uneven ground; this is typical for bilateral loss of vestibular organs (bilateral vestibulopathy)

Finally, symptoms that occur in addition to vertigo are important for classification. Hearing loss, ear noises, or tinnitus are typical for disorders originating from the inner ear. These are indicative of Menière's disease.

4.1.1 Brief vertigo episodes when turning over and sitting up: Peripheral paroxysmal positional vertigo

In brief

In peripheral paroxysmal positional vertigo (PPPV), typically, spinning vertigo episodes lasting seconds to a minute occur, triggered by certain movements, such as turning over in bed, lying down, sitting up from a lying position, bending forward, or tilting the head back. The episodes are often accompanied by

malaise and nausea, occasionally by vomiting and visual disturbances with oscillopsia, as well as mild swaying dizziness when standing or walking. The vertigo episodes occur over a period of several days to months and may recur in half to the patients throughout life. A very typical feature is the onset of symptoms in the morning when first sitting up in bed. The cause of PPPV is a mechanical disturbance in the vestibular organ located in the inner ear. Small crystals, also called otoliths or ear stones, enter one of the three semicircular canals (usually the posterior canal, less often the horizontal) and, when the head moves relative to gravity, stimulate the sensory receptors, which is perceived as vertigo. Using the diagnostic positional maneuver, vertigo is provoked and, based on the patient's eye movements, it is determined which side (right or left) and which of the three semicircular canals is affected. With specific repositioning maneuvers, the ear stones are moved out of the canal and back to their original location, where they no longer cause vertigo episodes. The subsequent swaying dizziness indicates that the crystals are back in the correct place, which is desirable. This dizziness may last for a few days and is a good sign, although unpleasent, because it indicates that the crystals are "back home".

A typical case

You wake up at 6:30 a.m., turn over in bed, and suddenly the whole world is spinning. You keep your head and body very still, and after half a minute the vertigo subsides. You turn your head again, and again everything spins. You notice slight nausea and a rising urge to vomit. You try to sit up in bed, and again everything spins—it feels like the end of the world. You remain calm and still, and the spinning vertigo slowly fades. You try to get up to go to the bathroom; now everything is swaying as well. You hold onto the wall and just manage to reach the bathroom. Standing at the sink and looking in the mirror, you feel okay for the moment. Then you bend your head forward, and the carousel starts again. You quickly return to bed and lie down, and again the spinning vertigo occurs as you lie down. At rest, everything subsides. You try to sleep. After an hour, you wake up and the symptoms are less severe. However, you can hardly eat breakfast because you still feel nauseous.

This is the typical onset of peripheral paroxysmal positional vertigo: the first symptoms typically occur in the morning when turning over or sitting up, spinning sensations that slowly subside at rest, accompanied by nausea, retching, and even vomiting. Additionally, there is swaying dizziness and gait instability when walking.

What you need to watch out for are additional symptoms that may indicate a serious cause, such as double vision, swallowing difficulties, speech disturbances, paralysis, sensory disturbances, or coordination problems in the arms or legs. In such cases, you should see a doctor immediately, or better yet, go to a hospital, as this could also be a stroke.

What happens next? The same day, you visit your general practitioner. You tell him your story. He asks about other symptoms. He also asks whether the symptoms were preceded by a fall or prolonged bed rest, which is not the case. You also have no other illnesses, especially no ear diseases.

The general practitioner now moves you from a sitting position to the right and left with the head turned in the opposite direction. When turning you to the right, he triggers severe spinning vertigo. He observes, using particular goggles, that your eyes are jumping. With this, he quickly makes the diagnosis: This is peripheral paroxysmal positional vertigo on the right side, in your case originating from the posterior semicircular canal.

The doctor explains the condition to you: Small crystals, which normally reside elsewhere, have fallen into your posterior semicircular canal. This causes brief vertigo episodes whenever you change the position of your head relative to gravity.

He now shows you how you can be treated: In an upright sitting position, you should turn your head 45° to the left and then quickly throw yourself onto your right shoulder, landing on the back of your head so that you are looking diagonally at the ceiling. This again triggers severe, but intentional, spinning vertigo. You remain in this position for 60 seconds. Then comes the "big move": You swing yourself forcefully onto your left side, so that you now land with your forehead facing down. After a few seconds, spinning vertigo and nausea occur again. You hold this position for another 60 seconds, then sit up. Now you feel pulled backward; the doctor holds you so you don't fall backward. You repeat these exercises two more times. Afterwards, the doctor ensures that you can perform the exercises independently.

You now stand up and notice increased swaying, which is a good sign. It shows that the small crystals have at least partially returned to where they came from and belong. The general practitioner gives you a follow-up appointment in a week.

At home, you do the exercises three times in the morning, three times at noon, and three times in the evening. On the third morning, you notice that you can no longer trigger spinning vertigo, but you still have severe swaying vertigo with gait instability. As advised by your doctor, you now move around a lot, and after a few days these symptoms also subside.

You go for a follow-up visit to your doctor and report that you were already feeling well by the third day. The doctor moves you back and forth again and is pleased that neither spinning vertigo nor these jumping eye movements can be triggered.

Finally, your doctor tells you that the risk of such symptoms recurring for the rest of your life is 50%. He recommends performing the same treatment maneuvers if these symptoms recur, so that you can heal yourself in this way.

A typical not-so-typical case

You have suffered for years from recurrent phases lasting days to weeks with swaying dizziness, gait instability, and mild nausea. You avoid quick movements, lie down in bed very slowly, and sit up in bed very slowly. You also turn over in bed very slowly. You do not suffer from episodes of spinning vertigo, the leading symptom is dizziness and imbalance.

Because of these symptoms, you have already seen many doctors: general practitioner, orthopedist, neurologist, and ENT specialist. None of them found anything, except that you sway more when standing.

Since you keep having these symptoms and feel significantly impaired during these times, you go to a dizziness center because your symptoms are particularly severe at the moment. Here, you are first carefully questioned and examined. Positional maneuvers are also performed. When you are thrown to the left, you experience severe brief spinning vertigo, like after a carousel ride. Additionally, the doctor observes your eyes jumping upward and to the left. This allows the diagnosis to be made very quickly and easily: You are suffering from peripheral positional vertigo originating from the left posterior semicircular canal.

The doctor asks you why the diagnosis had not been made before. You reply that you have never been moved back and forth in this way and that you often saw doctors when you were symptom-free.

What can we learn from your case?

1. Patients with peripheral paroyxsmal positional vertigo may primarily suffer from swaying dizziness, imbalance and gait instability, especially if they perform the movements that trigger spinning vertigo very slowly or avoid them altogether.
2. If patients visit a doctor during a symptom-free period, the physical examination is often unremarkable.
3. Unfortunately, even today, not all patients are examined using positional maneuvers, which in our opinion should be performed on every patient with vertigo and dizziness, regardless of medical history, at every doctor's visit, to identify the easily treatable peripheral paroxysmal positional vertigo.
4. If you suffer from vertigo, ask every doctor to perform the positional maneuvers on you, so that peripheral paroxysmal positional vertigo is not overlooked.

A common cause of brief vertigo episodes lasting seconds to a minute is peripheral paroxysmal positional vertigo, formerly known as benign paroxysmal positional vertigo. In the typical form, brief spinning vertigo episodes often first occur in the morning when turning over, subside at rest, and may be accompanied by nausea, vomiting, and oscillopsia (see video in Fig. 4.1). However, there are also patients who primarily suffer from swaying dizziness and gait instability (see video in Fig. 4.2), in whom the diagnosis of peripheral paroxysmal positional vertigo is often missed. Therefore, it is important that patients with vertigo and dizziness are examined with positional maneuvers at every doctor's visit.

Peripheral paroxysmal positional vertigo (PPPV) is the most common cause of vertigo originating from the inner ear. It can be diagnosed very precisely, as the diagnostic positional maneuvers can determine whether the right or left ear is affected and in which of the three semicircular canals in the inner ear the crystals are located. The doctor can also identify the underlying mechanism during the maneuvers.

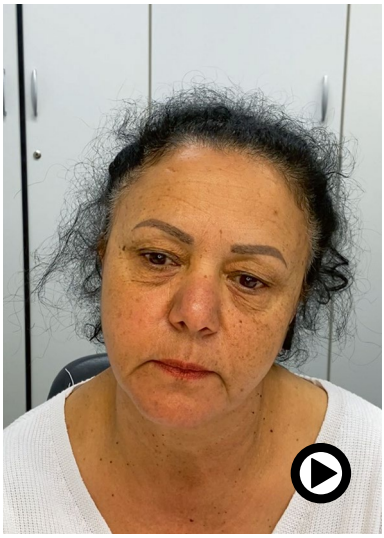

Fig. 4.1 Typical case of peripheral paroxysmal positional vertigo (PPPV). The diagnosis was confirmed by diagnostic positional maneuvers and the patient was quickly and successfully treated (▸ https://doi.org/10.1007/000-ky3)

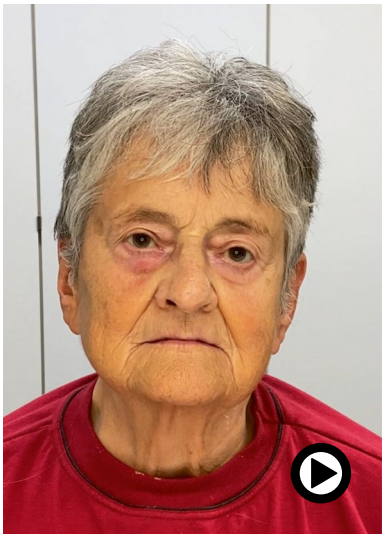

Fig. 4.2 An atypical typical case. The consequence: All patients with vertigo should be examined for positional vertigo at every doctor's visit, regardless of medical history. This patient suffered from typical peripheral paroxysmal positional vertigo of the right posterior semicircular canal and was quickly and successfully treated (▸ https://doi.org/10.1007/000-kxg)

Peripheral paroxysmal positional vertigo should not be overlooked, as it

- leads to an impairment of quality of life and functioning in affected individuals. For example, the risk of falls is significantly increased, with the danger of bone fractures.
- is one of the conditions that can be most accurately classified. This is done simply by performing the positional maneuvers. These should be performed on all patients with vertigo and dizziness at every doctor's visit.
- can be treated very quickly and easily with repositioning maneuvers, without expensive diagnostic or other treatment procedures.

Important

After successful treatment, patients often suffer from movement-dependent swaying dizziness and gait instability. These are good signs, even if this initially seems counterintuitive. They indicate that the ear stones in the inner ear have returned to where they came from and belong. The doctor should inform patients of this before performing the treatment maneuvers, so that they interpret this positively and do not feel unsettled. This also reduces the risk of patients developing functional dizziness. Peripheral paroxysmal positional vertigo can recur at any time. The risk is about 15–20% in the first year and, after one episode, 50% over the course of a lifetime.

Patient symptoms

Typically, patients report recurrent episodes of vertigo lasting seconds to a minute. Most often, this is spinning vertigo, like being on a carousel. The episodes are typically triggered by changes in head position relative to gravity, such as turning over, sitting up, or lying down in bed. The patient then intuitively lies still, after which the spinning vertigo subsides. With renewed head or body movements, for example when bending forward or tilting the head back, it recurs. Symptoms often also occur in the morning when first sitting up or turning over, because the small crystals that cause the condition clump together overnight.

Risk factors

In most patients, peripheral paroxysmal positional vertigo occurs in the context of otherwise complete health. Risk factors for its occurrence are older age, prolonged bed rest, e.g., as a result of another illness such as bone fracture, traumatic brain injury, as well as any other disease affecting the inner ear, e.g., after vestibular neuritis, Menière's disease (with vertigo episodes

and hearing loss), or vestibular migraine (vertigo episodes with headache, photophobia, or phonophobia). Peripheral paroxysmal positional vertigo occurs twice as often in women as in men, and the right side is affected twice as often as the left, since most people sleep on their right side.

What does the doctor observe during the physical examination?
Horizontal semicircular canal: The doctor should perform the diagnostic positional maneuvers for both the right and left sides for all three semicircular canals. Nowadays, the examination begins with the horizontal canal: The patient lies on their back with the head elevated by 30°. The head is then turned to the right and left, and the doctor observes through special goggles whether nystagmus can be triggered. This is accompanied by brief spinning vertigo.

Posterior semicircular canal: Then the posterior canal is examined. For this, the patient sits upright, and the doctor turns the head 45° in one direction and then throws the patient 90° in the other direction. If the patient's description of symptoms suggests which side is likely affected, that side is examined first. Otherwise, the right side is examined first, as it is affected twice as often as the left.

Anterior semicircular canal: A very rare form is peripheral paroxysmal positional vertigo originating from the anterior semicircular canal. For this, the patient is laid flat on their back and the head is extended 30° backward.

All these maneuvers have the advantage that treatment of peripheral paroxysmal positional vertigo begins immediately with diagnosis, i.e., the first steps of diagnosis correspond to the first steps of treatment.

Causes of brief vertigo episodes: Which disorders may be behind them?
The causes of brief vertigo episodes can be classified as

- peripheral vestibular disorders associated with recurrent brief vertigo episodes. Primarily, these include
  - the aforementioned peripheral paroxysmal positional vertigo originating from the posterior or horizontal semicircular canal, or, rarely, from the anterior canal. The doctor can diagnose this based on the direction of the eye movements (positional nystagmus) triggered during the maneuvers.
  - a peripheral paroxysmal positional vertigo in which two or three semicircular canals are affected; in this case, it is advisable to wait a few days and then re-examine the patient, as spontaneous recovery rates are high.

  - bilateral peripheral paroxysmal positional vertigo, which is relatively more common after a head injury with impact to the head.

- another disorder, e.g., due to compression of a vestibular nerve by a blood vessel or a bony defect in the inner ear. These are described in more detail in other chapters of this book.
- central causes, especially central positional vertigo/nystagmus due to cerebellar disorders. The doctor can distinguish these by physical examination. In these cases, magnetic resonance imaging of the head is necessary to search for the cause.
- other disorders, e.g., orthostatic dizziness (see Sect. 4.4.1). Here, symptoms typically occur when standing up and rising from the left or right lateral position, but not when lying down, which is important for differentiation.

Therapy

The basic prerequisite for treatment is that the affected side, i.e., right or left, as well as the affected semicircular canal, i.e., posterior, horizontal, or anterior, have been identified. It is important that the doctor does not overlook bilateral peripheral positional vertigo, especially after a fall. Very rarely, multiple canals on one side may be affected. The goal of the repositioning maneuvers is to move the crystals out of the affected canal.

Treatment of the most common form: peripheral paroxysmal positional vertigo of the posterior semicircular canal

Semont maneuver:

- While sitting, the patient first turns the head 45° toward the unaffected side (to bring the posterior canal into the plane of movement).
- The patient is then moved at moderate speed 90° toward the side of the affected labyrinth; this position must be held for 60 seconds.
- Then comes the so-called big move: The patient is moved 180° to the affected side, where they must remain for another 60 seconds.
- Finally, the patient sits upright for 60 seconds.

These maneuvers should be performed three times in the morning, three times at noon, and three times in the evening until symptoms resolve.

A more effective method for treating positional vertigo of the posterior semicircular canal is the Semont-plus maneuver, in which the patient's head is moved 60° below the horizontal when turning to the affected side (see Fig. 4.3).

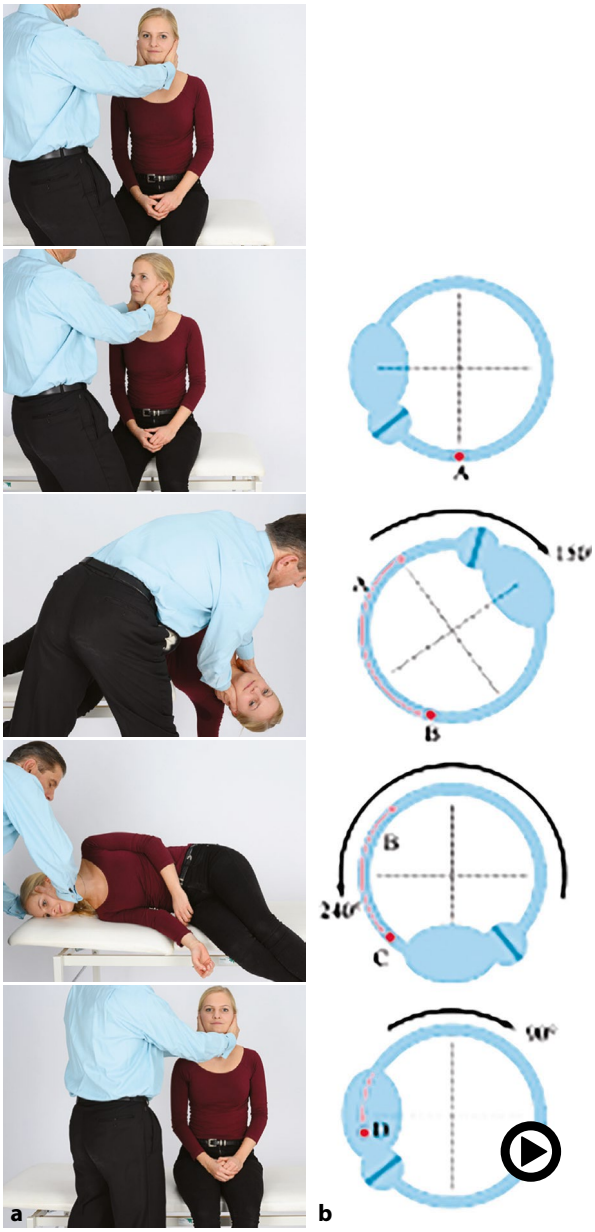

Fig. 4.3 **a** Treatment of a patient with peripheral paroxysmal positional vertigo of the right posterior semicircular canal using the Semont-plus maneuver. Each position must be maintained for one minute. The exercises must be performed three times in the morning, three times at noon, and three times in the evening until no positional vertigo can be triggered in the morning. **b** Schematic representation of the movement of the otoliths during the Semont-plus maneuver (▸ https://doi.org/10.1007/000-kxh)

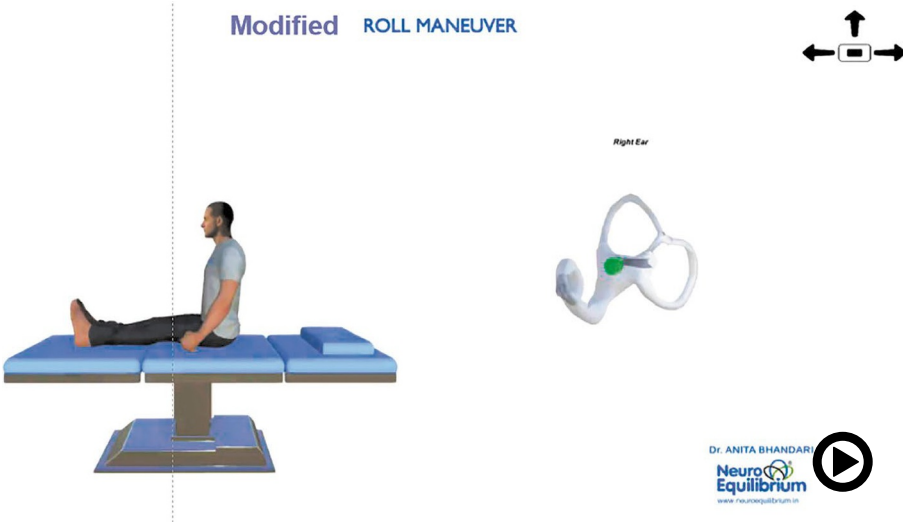

Fig. 4.4 Schematic representation of the treatment of a patient with peripheral paroxysmal positional vertigo of the right horizontal semicircular canal using the modified roll maneuver. The patient is first turned to the affected side (right) and then rotated in 90° increments toward the unaffected side. Each position should be maintained for 30 seconds. The more rotations, the better. Afterwards, the patient should stand up. On the right side, the changing position of the semicircular canals in the inner ear during the roll maneuver is shown (video without sound) (▸ https://doi.org/10.1007/000-kxj)

Treatment of the second most common form: peripheral paroxysmal positional vertigo of a horizontal semicircular canal with freely moving crystals

We recommend the roll maneuver in the supine position (see video in Fig. 4.4):

- The patient first turns toward the side identified by the doctor as affected,
- then in 90-degree steps along the body's longitudinal axis toward the unaffected side, remaining in each position for 30 seconds.
- Afterwards, the patient rests for 12 hours on the unaffected side.

Treatment of the third most common form: peripheral paroxysmal positional vertigo of a horizontal semicircular canal with adherent crystals

By shaking the head, which is bent forward by 120° while sitting, the adherent crystals can be dislodged (video in Fig. 4.5).

Fig. 4.5 Treatment of a patient with positional vertigo of a horizontal semicircular canal with adherent crystals: Here, simple head shaking helps, and as often as possible (video without sound) (▸ https://doi.org/10.1007/000-kxk)

Self-treatment by the patient at home

It is important that the patient receives precise instructions from the physician on how to perform the maneuvers independently at home. This applies to all forms. The physician should ensure in the office that the patient is performing the repositioning maneuvers correctly; if so, the cure rate is over 95%. It is helpful if family members perform the maneuvers together with the patient and check for correct execution.

Frequency of repositioning maneuvers

The repositioning maneuvers for each type of peripheral paroxysmal positional vertigo should be performed three times in the morning, three times at midday, and three times in the evening. The exercises are usually most effective in the morning, as the crystals clump together overnight, making it easier to expel the clump than to expel individual crystals.

Assessing the success of repositioning maneuvers

If the patient is unable to trigger any episodes of rotational vertigo for three consecutive mornings, it can be assumed that the treatment was successful. With successful treatment, a movement-dependent swaying vertigo occurs, which is desirable. This swaying dizziness is due to the crystals falling back to where they came from and belong. The treatment of choice for this swaying dizziness is regular physical activity. The more frequently the patient moves, the faster the swaying dizziness will subside within the following days to a few weeks.

Undesirable effects of repositioning maneuvers and what can be done about them

A temporary side effect may be nausea, especially with repeated repositioning during a session. This nausea can be prevented with medication, e.g., by administering 50 mg dimenhydrinate half an hour before the maneuvers. Some studies show that central compensation of these symptoms can be improved by betahistine.

In most successfully treated patients, swaying or lightheaded dizziness with gait unsteadiness occurs for one to four weeks when walking and moving, due to partial repositioning of the crystals to the utricle (post-repositioning otolith dizziness). Patients should be informed of this before treatment begins so that they can interpret this dizziness as a positive sign.

Risk of recurrence

According to follow-up observations over an average of ten years, the risk of recurrence in treated patients is about 50% overall. This usually happens in the first year, then becomes less frequent, and is more common in women than in men.

What to do if symptoms persist or recur?

If episodes of positional vertigo persist for more than two weeks, the patient should ideally see the physician again to check the diagnosis and treatment success.

Possible causes for persistent symptoms include:

- **The diagnosis was incorrect**: The diagnosis should be reviewed by the physician. Referral to a specialist may be indicated. There may be bilateral peripheral positional vertigo (for example, after a mild traumatic brain injury), or the crystals may have migrated into another semicircular canal. However, it may also be a disorder originating from the cerebellum (central positional vertigo; see Sect. 4.2). Episodes of positional vertigo can also occur in vestibular migraine (see Sect. 4.2.2).
- **The repositioning maneuver was not performed correctly**: This should be checked by the physician.
- **Prolonged swaying dizziness:** In this case, increased exercises with balance training are recommended.
- **Development of secondary functional dizziness** (see Sect. 4.3).

4.1.2 Acute prolonged vertigo

In brief

Sudden onset of severe spinning vertigo lasting for days up to one to two weeks is the leading symptom in acute failure of a vestibular nerve (vestibular neuritis, also referred to as acute unilateral vestibulopathy). In addition, patients often experience nausea up to vomiting, unsteadiness when standing and walking with a tendency to fall, and visual disturbances with an apparent movement of the visual surrounding (oscillopsia). Movements of the head and body exacerbate the symptoms. Diagnosis is made using the head impulse test (rapid head turns to the right and left while observing eye movements) and the physician detects specific eye movement disorders (nystagmus). Important: Attention should be paid by the patient to additional symptoms such as double vision, swallowing or speech disturbances, paralysis, or sensory disturbances, which may indicate a stroke. These must be reported to the physician immediately, who, if in doubt, will order a CT scan with vascular imaging and later an MRI. In the first one to three days, anti-vertigo medications can be taken to treat nausea and vomiting. Taking corticosteroids (starting treatment within three days of symptom onset) for a short period can lead to improved recovery of the vestibular organ. Based on animal studies, betahistine should also be given to improve symptoms and so-called central compensation. Early balance training and regular physical and sports activity train the balance system and promote rapid improvement. Most patients recover very well in the long term. Recurrence is rare.

A typical case

You are sitting comfortably in a café when suddenly the world starts spinning. You have never experienced this before. The spinning intensifies. The images before your eyes jump back and forth. You begin to feel nauseous. You try to stand up, but fall to the left and immediately sit back down. Your companion notices that you are very pale. The emergency doctor is called because you feel so unwell and have now vomited. You are taken to the nearest hospital. During the ride, vertigo and nausea worsen, also due to the movement of the vehicle. The doctor gives you medication for the nausea. After 15 minutes, you arrive at the hospital and are treated in the emergency department.

The doctor asks about your symptoms. You describe those mentioned above. Then the doctor wants to know if there are any additional symptoms: double vision, swallowing difficulties, speech disturbances, sensory disturbances in the face or limbs, paralysis, or coordination problems as well as headache, hearing problems and prior episodes of vertigo. You answer no to all of these. He also asks if you have high blood pressure, diabetes, or lipid metabolism disorders. You answer no here as well. You have also never had a stroke or heart attack.

The doctor then examines you. He looks into your eyes and sees that they are twitching slightly to the right (nystagmus). He puts special goggles (so-called nystagmus goggles) on you, under which everything appears blurry. Now he can see that the eyes twitch to the right (fast phase of the eye

movements to the right, this means a nystagmus to the right). He asks you to look right and left. Then he quickly turns your head to the left and right and tells you that the left vestibular organ in the inner ear, or more precisely, the left vestibular nerve, has failed. Then your ability to stand and walk is tested: you tend to fall to the left, especially after closing your eyes. The doctor finds no other abnormal findings.

The doctor now discusses the findings and the cause of the illness with you: It is very likely an inflammation of the left vestibular nerve, usually triggered by herpes viruses. The medical term is vestibular neuritis or acute peripheral vestibulopathy. There were no indications of a stroke or other ear diseases, namely Menière's disease or vestibular migraine.

Now it is time for treatment. For the nausea, you receive dimenhydrinate for one day, three times daily. For the inflammation and swelling of the vestibular nerve, you receive corticosteroid tablets each morning, in a tapering dose. Additionally, you are given a medication to improve symptoms and central compensation: betahistine, three times daily. And as a fourth measure, you undergo intensive balance training, three times daily. Initially in bed with rapid head shaking to the right and left, at least 100 times per day. The next day you can already get up, and the physiotherapists conduct balance training with you, which you should then continue independently. On the second day, you are already feeling well enough to leave the hospital and go home accompanied.

At a follow-up visit three months later, the doctor finds that your vestibular nerve has largely recovered and you have almost perfect balance: you can stand on one leg with your eyes closed for four seconds. Finally, the doctor informs you that the risk of having this illness again is less than 10%.

Case example 2

The videos in Fig. 4.6 and 4.7 show the history and examination of a patient with vestibular neuritis.

Patient symptoms

The acute failure of a vestibular organ initially presents with sudden onset of severe spinning vertigo, which can last at least 24 hours and up to one to two weeks. The vertigo is usually accompanied by nausea, vomiting, unsteady gait with a tendency to fall to the affected side, and visual disturbances with the image of the visual surrouding is jumping infront ot the patient's eyes (oscillopsia). Head movements exacerbate the symptoms, so patients intuitively remain still. Brief episodes of vertigo may precede the acute onset.

What patients should pay special attention to and which symptoms do not fit acute vestibular nerve failure

Important: Acute, prolonged rotational or swaying vertigo can also be caused by a stroke in the brainstem or cerebellum (see Sect. 4.2). Central

Fig. 4.6 Acute prolonged vertigo. History of a patient with failure of a vestibular nerve (vestibular neuritis, also called acute unilateral vestibulopathy) (▸ https://doi.org/10.1007/000-kxm)

neurological symptoms such as double vision, swallowing and speech disturbances, paralysis, or sensory disturbances indicate a stroke and should be watched for in particular. Acute hearing loss, tinnitus, or headache are also not part of the clinical picture of vestibular organ failure. Acute vertigo with hearing loss, tinnitus, or ear pressure suggests Menière's disease, but can also occur with a circulatory disorder of a vestibular organ. Vertigo with headache can also be caused by a stroke. If vertigo with headache occurs frequently, vestibular migraine should be considered. If these symptoms are present, they should be reported to the physician immediately. The risk of stroke is increased in older people, those with high blood pressure, diabetes mellitus, high cholesterol, or a history of stroke or heart attack. These risk factors should also be reported to the physician so that the correct diagnosis can be made quickly.

What can patients do if acute severe prolonged vertigo occurs for the first time?

Patients should pay attention to the symptoms mentioned above and especially to any additional symptoms that may indicate a stroke.

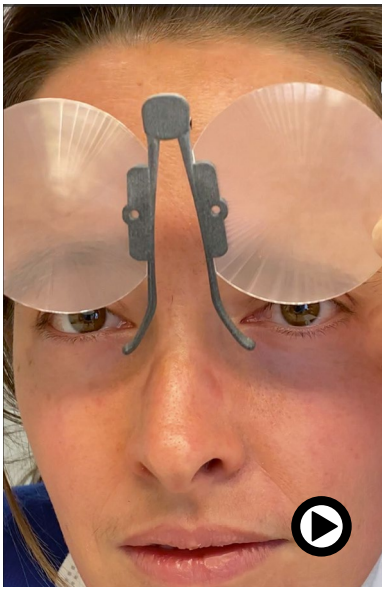

Fig. 4.7 Acute prolonged vertigo. Examination of a patient with failure of the left vestibular nerve (vestibular neuritis or acute unilateral vestibulopathy). The jumping eye movements (spontaneous nystagmus) are largely suppressed by fixation. Under the M-glasses: Jumping eye movements to the right (horizontal-torsional spontaneous nystagmus). Intensified when looking to the right. No change in direction when looking to the left, i.e., there is no evidence of gaze-evoked nystagmus. Head impulse test to the left: The eyes move with the head to the left and the patient must make a rapid corrective eye movement to the center. This indicates that the left vestibular nerve has failed (▸ https://doi.org/10.1007/000-kxn)

Patients should seek medical attention as soon as possible, ideally in an emergency department, so that a diagnosis can be made quickly and a serious cause ruled out.

Patients should rest as much as possible in the first hours, as movement exacerbates the symptoms.

What does the doctor see during physical examinations?

1. Examination for spontaneous nystagmus without and with a specific examination instrument, the Frenzel or M-glasses: Here, a spontaneous nystagmus beating toward the unaffected side is seen. Its intensity increases under the glasses and decreases when looking at a target.
2. Rapid head impulse test to assess vestibular function: Here, the gaze deviates with head movement toward the affected side and a corrective movement back to the initial fixation point is necessary.

3. Examination of standing ability using the Romberg test: Here, increased swaying is observed, typically toward the side of the failed vestibular nerve, with a marked increase after closing the eyes.

Which findings argue against the diagnosis of vestibular neuritis?

1. On examination of eye movements
   - one eye is higher than the other,
   - the direction of the jumping eye movements changes when looking to the right and left (gaze-evoked nystagmus).
2. On examination of hearing
   - new unilateral hearing loss and
   - new tinnitus are detected.
3. On neurological examination
   - swallowing and speech disturbances,
   - paralysis of the face or arms or legs,
   - sensory disturbances in the face or arms or legs, and
   - coordination disorders of the arms or legs.

Instrumental examinations

Instrumental examinations to assess vestibular function include the head impulse test with video oculography. If this is not available, ear irrigation with warm and cold water can be performed.

To rule out other diseases, especially stroke, a CT scan and later an MRI of the head are performed if in doubt.

Cause

The cause of acute vestibular nerve failure (vestibular neuritis or acute unilateral vestibulopathy) is thought to be inflammation of the vestibular nerve. In most cases, this is probably caused by reactivation of "dormant" herpes viruses. Almost everyone carries these viruses. Normally, they cause no symptoms other than cold sores. The inflammation causes swelling of the nerve, which runs through a narrow bony canal, thereby impairing nerve function.

Other causes of acute prolonged vertigo (differential diagnoses)

The diagnosis of vestibular neuritis is a diagnosis of exclusion, i.e., there must be no evidence of other causes. The most important other diseases characterized by acute, prolonged vertigo are

- Stroke in the brainstem or cerebellum (see Sect. 4.2.1)
- Stroke in the inner ear: sudden rotational vertigo and hearing loss
- Episode of vestibular migraine (see Sect. 4.2.2)
- Menière's disease: episodes of vertigo with hearing loss (see Sect. 4.1.3)

How do rotational vertigo and jerky eye movements arise?
A special feature of the vestibular nerves is that their nerve fibers are active even without head movement, i.e., they fire regularly. If one side fails, the other side remains active—even at rest—leading to an imbalance. As a result, rotational vertigo and nystagmus occur. Both subside after days to a few weeks. This process is called central compensation and can be accelerated and improved by medication and physiotherapy.

Therapy
Based on the above-mentioned cause of vestibular neuritis, the following three treatment principles can be derived:

Therapy of acute symptoms: rotational vertigo, nausea, and vomiting In the first one to three days, anti-vertigo medications can be taken to treat nausea and vomiting. We usually give dimenhydrinate (50–100 mg). These medications should not be given for more than three days, as otherwise central compensation is inhibited and patients remain ill longer than necessary.

Treatment of the underlying disease Taking corticosteroids (starting treatment within three days) for a short period can lead to better recovery of the vestibular nerve. We usually give methylprednisolone, initially 100 mg orally per day, and reduce the dose by 20 mg every fourth day.

Improvement of balance through medication and physiotherapy Early balance training and regular physical and sports activity challenge and train the balance system to achieve rapid improvement. We recommend stepwise physical training for about 15 minutes three times per day over a period of about four weeks. This should initially be supervised by a physiotherapist and mainly involves rapid head turns to the right and left. Subsequently, training sessions to improve standing, posture, and gait regulation during eye-head-body movements should be performed, which the patient can later do independently. It is important that the difficulty of balance and coordination exercises is continuously increased to a level above normal requirements.

Course of the disease and complications

- **Acute phase**

The first phase of the disease is characterized by severe spinning vertigo, directional tendency to fall, nausea up to vomiting, and usually a severe feeling of illness. Since symptoms worsen with movement, patients often intuitively seek rest. The symptoms then gradually subside over days to one to two weeks. Afterwards, there is freedom from symptoms at rest, i.e., when lying down and sitting.

- **Further course and follow-up**

After the initial severe spinning vertigo with tendency to fall, mild swaying vertigo and gait unsteadiness may persist for some time. In most cases, however, the function of the affected vestibular nerve improves again. But even if the function of the vestibular nerve does not fully recover, significant persistent symptoms over months to years are not a typical consequence of the disease. Only with rapid head movements or in situations that place high demands on the balance system, such as balancing or walking in the dark, may unsteadiness be noticeable for a long time.

For follow-up of the therapeutic effect, outpatient follow-up visits after four weeks, six months, and twelve months are advisable, also to assess the extent of recovery of vestibular nerve function and to provide appropriate advice to the patient.

The risk of developing vestibular neuritis again is less than 10%. However, if prolonged episodes of vertigo occur repeatedly, other causes such as vestibular migraine (see Sect. 4.2.2) or Menière's disease (see Sect. 4.1.3) should be considered.

About 15% of patients develop post-infectious peripheral paroxysmal positional vertigo (see Sect. 4.1.1) or secondary functional dizziness (see Sect. 4.3).

4.1.3 Episodes of Vertigo with Hearing Loss: Menière's Disease

In Brief

Recurrent episodes of vertigo lasting many minutes to hours, accompanied by hearing loss, tinnitus, or a feeling of pressure in the affected ear: these are the typical symptoms of patients with Menière's disease. The cause is a disorder of

the inner ear: too much fluid, endolymph, is produced. Initially, this leads to swelling of the inner ear space, then to the tearing of a thin membrane and the mixing of two fluids, endolymph and perilymph, within the inner ear. This results in episodes of vertigo, hearing loss, tinnitus, and pressure on the affected side. When the pressure eventually decreases and the leak in the membrane closes, the episode subsides. In addition to a physical examination, a hearing test and ear irrigation with warm and cold water to assess the vestibular system are performed. The hearing test typically shows reduced hearing for low frequencies. During an acute episode, vestibular suppressants such as dimenhydrinate can be taken. More important, however, is preventive (prophylactic) treatment. The goal is to reduce the frequency of vertigo episodes. For this, high-dose betahistine, or in combination with a medication (both "off-label") that reduces the breakdown of betahistine in the body (so-called monoamine oxidase B inhibitor), is recommended for at least six to twelve months (this personal recommendation is based on the observation in single cases and a so-called phase 1 study and so far not proven in clinical trials). To assess the treatment effect and adjust therapy, a vertigo diary (days with an episode of vertigo) should be kept.

A Typical Case

You suffer from recurrent episodes of vertigo that last for many minutes to hours. In addition, you have noticed that during such episodes you hear worse in your right ear, and recently also a very little in your left ear. You have also had tinnitus on the right side for many years, which fluctuates in intensity. Because of these symptoms, you saw your general practitioner, who suspected Menière's disease. He is now referring you to an ear, nose, and throat (ENT) specialist.

The ENT specialist carefully asks you about your symptoms. You recount the above history. He asks if you experience a feeling of pressure in your ear, which you confirm for the right side. He also asks how long the vertigo episodes last. You state that they usually last many minutes to a few hours. He wants to know since when and how frequently you have had these symptoms. The symptoms have been present for three years and have recently occurred two to three times per month.

The doctor also asks whether you have or have had migraines, which you deny. He asks whether the vertigo episodes are accompanied by sensitivity to light or noise; you also answer no to this.

The ENT specialist now tests your hearing during the physical examination using a tuning fork and rubbing sounds. He finds a marked hearing loss in the right ear. The rest of the examination is normal.

Instrumental examinations follow: Testing your hearing with an audiogram confirms hearing loss on the right in the low-frequency range. On the left, there is mild low-frequency hearing loss. Next, your ears are irrigated with cold and warm water to test the vestibular organs. This shows that the right vestibular organ is not functioning properly.

The doctor discusses the likely diagnosis with you. Since you have not yet had a head MRI, he refers you to a radiologist. This examination, with and without contrast, shows normal findings, in particular no evidence of a vestibular nerve tumor.

You return to your ENT specialist with the images. He now explains that you do indeed have Menière's disease, much more pronounced on the right than the left. This is a disorder of the inner ear in which too much fluid is produced, putting the entire system under pressure. If the pressure becomes very high, a membrane tears and a potassium-rich fluid leaks out. This leads to episodes of vertigo and hearing loss.

Because of the high frequency of vertigo episodes, he recommends prophylactic treatment. He prescribes you betahistine, 48 mg three times a day (off-label dosage (approved dosage is 48 mg per day, higher dosages are based on observations in single cases, no clinical trials yet). To assess the effect of therapy, you are to keep a vertigo diary (days with an episode of vertigo).

For the treatment of the episodes themselves, you are to take a medication for vertigo and nausea; he prescribes dimenhydrinate.

You return to your ENT specialist after three months, but unfortunately there has been no improvement. For this reason, the doctor increases the dose of betahistine to 96 mg three times a day (see above). You are to continue keeping your vertigo diary.

After another three months, you return for another outpatient visit. Now, you have been free of vertigo for the past two months, so the treatment is successful. Further check-ups at six and twelve months also show a good course, so that the daily dose of betahistine can be reduced by 24 mg every third month.

Case Example 2

In the video in Fig. 4.8, a patient with Menière's disease describes her medical history.

Fig. 4.8 Patient with Menière's disease experiencing episodes of vertigo and hearing loss (▸ https://doi.org/10.1007/000-kxp)

Patient Symptoms

Typically, there are recurrent episodes of vertigo, usually accompanied by hearing loss, tinnitus, and/or a feeling of pressure in the affected ear. The duration of episodes is usually between 20 minutes and 12 hours. In about one third of cases, the often abrupt onset vertigo is preceded by an increase in tinnitus and ear pressure as well as hearing loss. A temporal association between acute hearing loss and vertigo episodes within ±24 hours is often only reported by the patient when specifically asked, which is important for diagnosis. Also typical for the disease are pronounced fluctuations over time, with vertigo episodes, sudden hearing loss with improvement in hearing, varying degrees of tinnitus, and sometimes more or less pronounced ear pressure. After many episodes, hearing loss may progress and become permanent; vertigo episodes then often occur without other ear symptoms. Over the years in about 20 to 40% both ears may be affected as the disease progresses. Very rarely, sudden falls out of nowhere occur (the medical term is Tumarkin's otolithic catastrophy). Important: Patients with Menière's disease may also simultaneously suffer from vestibular migraine (see Sect. 4.2.2) or peripheral paroxysmal positional vertigo (see Sect. 4.1.1).

What Patients Should Pay Special Attention To

It is important to note the duration of symptoms, i.e., vertigo and hearing loss (minimum and maximum duration of episodes). Have there previously been episodes of sudden hearing loss or tinnitus? Has hearing ever been tested? Do the symptoms occur spontaneously, are there triggers, aggravating or alleviating factors? All of this should be communicated to the doctor. Has a head MRI with contrast ever been performed? If so, the images and findings should be presented to the doctor.

Since patients can suffer from both Menière's disease and vestibular migraine (see Sect. 4.2.2), the following additional questions are important: Does or did the patient have migraine? Are the vertigo episodes accompanied by headache or hypersensitivity to light or noise?

This also applies to the simultaneous occurrence of peripheral paroxysmal positional vertigo, which can frequently occur with any other inner ear disorder (see Sect. 4.1.1).

What Can Patients Do If They Suffer from Vertigo with Hearing Loss?

1. The patient should pay attention to whether the hearing loss occurs in episodes, i.e., whether sudden hearing loss resolves or is permanent. How long has this been present?

2. Is the hearing loss episodic or slowly progressive? The latter is also seen with a benign tumor of the vestibular nerve (vestibular schwannoma, formerly called acoustic neuroma).
3. Does the patient suffer from tinnitus or a feeling of pressure in the ear?
4. How does the patient describe the tinnitus?
5. Is only one ear or are both affected?

Cause

Menière's disease is a disorder of the inner ear. For various reasons, not all of which are known, too much fluid (endolymph) is produced. Patients perceive this as a feeling of pressure in the affected ear. Initially, the inner ear space swells, eventually a thin membrane tears, and two fluids, endolymph and perilymph, mix within the inner ear. This leads to excitation and then inhibition of function, with spinning—rarely swaying—vertigo, hearing loss, and/or (increased) tinnitus. When the pressure drops again and the leak closes, the episode subsides. If the membrane tears frequently, permanent deficits in balance and hearing function develop. Marked fluctuations due to variable excess pressure in the ear are typical for the disease.

What Does the Doctor See During Physical Examinations?
Physical examination between vertigo episodes:

- Assessment of hearing with rubbing sounds and tuning fork (Rinne and Weber tests)
- Examination of the vestibular system and eye movements: In Menière's disease, ear irrigation with cold and warm water often shows reduced function in the affected ear, whereas the rapid head impulse test is typically normal (see below).
- Examination of eye movements for central disturbances, which may indicate vestibular migraine, for example

During an episode (rarely observed in practice):

- Assessment of hearing: Typically, hearing is markedly reduced during an episode. Nowadays, this can be examined by the patient using an "hearing-App" between the episodes and during an episode which is highly recommended to get first-hand information.
- Examination of eye movements: Initially, there is unilateral, transient, minutes-long excitation of the affected ear: the eyes then beat rapidly toward the affected side. Later, a functional deficit develops and the eyes beat for many minutes toward the other side.

It has proven diagnostically helpful for the patient to record their eye movements during an episode and as mentioned above and below to self-test hearing using an app.

Instrumental Examinations
In addition to the physical examination, a hearing test and assessment of the vestibular organ are performed. All examinations should be repeated during the course of the disease, as the typical changes are often not detectable at the onset of the disease.

Hearing Test (Audiometry)
According to current diagnostic criteria for Menière's disease, audiometry is the most important instrumental examination. It requires evidence of significant hearing loss in the low-frequency range, also to identify the affected ear. The exact values are: increased bone conduction threshold by more than 30 dB at at least two consecutive frequencies below 2000 Hz.

Assessment of Vestibular Organ Function
This involves irrigating the ears with warm and cold water. This often reveals reduced function of the affected ear. In contrast, testing with the rapid head impulse test (video head impulse test) is usually normal. This combination is typical for Menière's disease and also helps distinguish it from other vestibular disorders.

Imaging
To rule out a (benign) tumor of the vestibular nerve (vestibular schwannoma) or other space-occupying lesions in this area, a head MRI with and without contrast should be performed.

Other Conditions That Can Cause Similar Symptoms
Here, three groups of symptoms should be distinguished:

- Recurrent episodes of vertigo are the main feature
- Combination of vertigo with hearing loss, i.e., accompanied by hearing loss or tinnitus
- First occurrence of prolonged vertigo

Other Conditions with Recurrent Episodes of Vertigo
Longer duration: In cases of recurrent episodes of vertigo, vestibular migraine (see Sect. 4.2.2) is another possibility. This is also characterized by

episodes lasting minutes to many hours. The following features help distinguish it and point toward vestibular migraine: absence of relevant hearing loss despite many episodes, migraine-typical symptoms during the episode, and migraine or a history of migraine.

It is important to note that patients can simultaneously have both Menière's disease and vestibular migraine, both of which must then be diagnosed and treated.

Shorter episode duration: Here, consider vestibular paroxysmia (episodes usually lasting less than a minute, occurring up to 100 times per day; see Sect. 4.1.5) and peripheral paroxysmal positional vertigo (brief episodes of vertigo triggered by changes in head position relative to gravity; see Sect. 4.1.1). Importantly, peripheral paroxysmal positional vertigo can frequently occur with any other inner ear disorder. This also applies to Menière's disease, so patients may suffer from both conditions. Therefore, patients should request to be examined for peripheral paroxysmal positional vertigo at every doctor's visit.

Other Conditions with Vertigo, Dizziness, and Hearing Loss or Tinnitus
Many diseases are associated with a combination of vertigo, dizzines, and hearing problems.

- Do the symptoms occur suddenly? In this case, inflammatory causes are possible, e.g., herpes zoster oticus, accompanied by pain and skin changes in and around the ear as seen in shingles, since it is the same virus. Treatment is with an antiviral medication. A rare but serious disease is Cogan's syndrome. This is an autoimmune disease characterized by vertigo and dizziness, bilateral hearing loss and tinnitus, and eye inflammation. Cogan's syndrome is a medical emergency, must be diagnosed quickly, and initially treated with high-dose intravenous corticosteroids. Another cause of sudden onset of vertigo and hearing loss is an inner ear infarction.
- Are the symptoms worsening slowly? This may be a (benign) tumor of the vestibular nerve (vestibular schwannoma). Therefore, an MRI of the brain with and without contrast should be performed.

First Occurrence of Prolonged Vertigo
This is usually due to unilateral loss of a vestibular nerve with sudden onset of spinning vertigo, nausea, oscillopsia, and tendency to fall (see Sect. 4.1.2). The absence of ear symptoms helps to distinguish this. Important: Rarely, in the first episodes of Menière's disease, vertigo is the predominant symptom,

making initial differentiation difficult, as hearing loss develops only later. Another possible cause is a stroke in the brainstem or cerebellum (see Sect. 4.2.1).

Course

The disease most commonly begins between the ages of 40 and 60, rarely in childhood. Initially, patients are symptom-free between episodes, then develop increasing tinnitus and hearing loss (usually low-frequency loss), which fluctuate greatly. The disease starts unilaterally with very irregular, initially increasing, then decreasing frequency of episodes, which may later affect the other ear. The longer patients are followed, the more often bilateral disease occurs: untreated, after 20 years, up to 40% of those affected. This also explains why Menière's disease is the so-far second most common identifiable cause of bilateral vestibular loss.

Treatment

- **Treatment of the episodes**

During episodes, vestibular suppressants such as dimenhydrinate can be taken to treat vertigo and nausea or vomiting.

- **Prophylactic treatment**

If patients experience two or more episodes per month, preventive treatment is advisable, aiming to reduce the frequency of vertigo episodes and prevent permanent damage to vestibular and hearing function. Many treatment methods have been proposed in the past. Unfortunately, scientific evidence for their efficacy is low to very low (this assessment is based on six Cochrane reviews from 2023). This applies, for example, to low-salt diets, injection of corticosteroids or gentamicin into the inner ear, inner ear surgery, and certain oral medications: diuretics or betahistine in the previously studied daily doses.

We currently recommend taking betahistine in a high, escalating dose up to 96 mg three times daily (the approved daily dose is 48 mg, so this is an off-label treatment, not proven so far by clinical trials) for at least six to twelve months. We have also achieved good treatment results with the combination of betahistine (48 mg or 96 mg three times daily) and a medication that inhibits the breakdown of betahistine (monoamine oxidase B inhibitor, e.g., rasagiline 1 mg daily or selegiline 5 mg daily; also off-label treatment, based on single cases and a phase 1 study, so far not proven in clinical trials). Betahistine often takes up to three months to reach its full effect. The efficacy of the combination treatment has to be investigated in studies.

To assess the effect of treatment and adjust therapy, the patient should keep a vertigo diary (days with an episode of vertigo).

4.1.4 Persisting postural imbalance and gait instability: bilateral vestibulopathy

In brief

The condition known as bilateral vestibulopathy, a partial or complete loss of function of both balance organs, usually develops gradually. The most common symptom is unsteadiness while standing or walking, worsening in the dark and on uneven ground. In addition, affected individuals often report blurred vision and a bouncing or shaking of the visual field during fast head movements or while walking. Treatment with certain antibiotics (aminoglycosides, such as gentamicin) that damage the inner ear, bilateral Menière's disease, a previous episode of meningitis, or genetic disorders (as recently discovered) are possible causes of this bilateral loss of function. However, in about one-third of all cases, the cause of bilateral vestibulopathy remains unclear. The most important therapeutic measure is intensive, daily, lifelong balance physiotherapy and gait training.

A typical case

For the past two years, you have noticed increasing unsteadiness and progressive gait instability. You feel fine when lying down or sitting. As soon as you stand up, you feel unsteady. You have also noticed that it is worse in the dark—such as when you have to go to the bathroom at night. You also feel increasingly unsteady when walking on uneven ground and have nearly fallen several times. Finally, you have noticed that during walking the image in front of your eyes is moving up- and downward and you have to stop walking in order to read a sign or recognize a face. You have already seen your general practitioner, who could not find a cause for your symptoms. She is now referring you to a neurologist.

You describe the above symptoms to the neurologist. She asks whether your unsteadiness and gait instability worsen in crowds, department stores, or other specific situations. You deny this. She also asks whether your symptoms improve after small amounts of alcohol or during physical activity. You deny this as well. Finally, she wants to know whether you were treated with intravenous antibiotics before your symptoms began—this was not the case.

The neurologist now examines you. First, she observes your eye movements. These are normal. Then she asks you to fixate on her nose and quickly turns your head to the right and left, observing your eye movements. With this test, she assesses the function of your balance organs. She observes that during these rapid head turns to the right and left, your eyes move with your head, and you then make a quick eye movement to see the examiner's nose again. This indicates that both balance organs are not functioning properly. She tests

your visual acuity and hearing; both are normal. Your sense of touch, tested with a tuning fork on your feet and hands, is also unremarkable. Reflex testing with a reflex hammer is likewise normal. This demonstrates that your peripheral nerves are functioning well. The doctor then asks you to stand upright. This allows her to assess your ability to stand. It becomes apparent that you sway much more when you close your eyes. You are unable to stand on one leg with your eyes closed—there is a risk of falling.

- **What further examinations are performed?**
  With a device that simultaneously measures the velocity of head and eye movements when the head is quickly turned to the right and left, the function of the balance organs can be analyzed in more detail. This reveals that both balance organs are largely, but not completely, nonfunctional.
  The neurologist also recommends a magnetic resonance imaging (MRI) scan of the head, both with and without contrast agent. This imaging of the head is unremarkable; in particular, there is no evidence of a tumor of the vestibular nerves.
  The doctor now asks you to return for an outpatient follow-up, bringing the MRI images. She discusses all findings, the diagnosis, and the treatment with you.
- **What is the cause of the symptoms?**
  The neurologist explains that both of your balance organs are not functioning properly. The medical term for this condition is bilateral vestibulopathy. This leads to the movement-dependent unsteadiness and gait disorder you are experiencing, both of which typically worsen in the dark and on uneven ground. Typically, you have no symptoms when sitting or lying quietly.
  The doctor also explains that the underlying cause of the condition often cannot be identified. From your medical history, it can at least be concluded that the dysfunction of the balance organs was not caused by certain antibiotics. It is thought to be due to premature aging in combination with genetic factors found recently of the hair cells and the vestibular nerve in the inner ear. It is important that the other sensory organs relevant for balance, especially the eyes and peripheral nerves, and central structures in the brainstem and cerebellum are most often intact. This means that, contrary to the fears of many patients, a wheelchair—even in the long-term—will not be needed for this condition.
  After the conversation, you are initially relieved that, although it is an unpleasant condition, it is not actually dangerous. You are now, of course, interested in what can be done about it.
- **What does the treatment look like?**
  The most important treatment principle is lifelong balance training, i.e., you need to do certain exercises every day for about 20 minutes to gradually improve your balance over weeks and months. It should be noted that a treatment effect is often only noticeable after two to three months. The same applies to deterioration if the patient stops balance training.
- **What is the course of the condition?**
  After three months, you return for a follow-up. The neurologist again tests your balance organs with the head impulse test: the dysfunction remains unchanged. In the assessment of your ability to stand, it is pleasing to see that you are now able to briefly stand on one leg with your eyes closed.

A further follow-up takes place after twelve months. Here, it is found that your vestibular function remains unchanged, but your ability to stand and walk has noticeably improved. You also report that you are coping better in everyday life. Fortunately, there have still been no falls. The doctor encourages you to continue the exercises daily and lifelong, just as you have been doing.

Case example 2

The videos in Figs. 4.9, 4.10, and 4.11 show the medical history, head impulse test, and assessment of standing ability in patients with bilateral vestibulopathy.

Bilateral vestibulopathy refers to the partial or rarely complete loss of function of both balance organs. Symptoms usually develop gradually and are barely noticeable to the patient at first. The earliest complaints most commonly reported are mild balance or gait disturbances or blurred vision during walking and rapid head movements. Imbalance and gait disturbances are especially noticeable when walking in darkness and on uneven terrain (e.g., forest floor, sand). Less commonly, bilateral dysfunction develops over a short period. This can occur, for example, during treatment with certain antibiotics (e.g., aminoglycosides) or in the context of meningitis.

Various reflex pathways between the vestibular organ, the eyes, and the legs play a crucial role in this often-overlooked condition. The

Fig. 4.9 Medical history of a patient with bilateral vestibulopathy, a bilateral loss of function of the balance organs (▸ https://doi.org/10.1007/000-kxq)

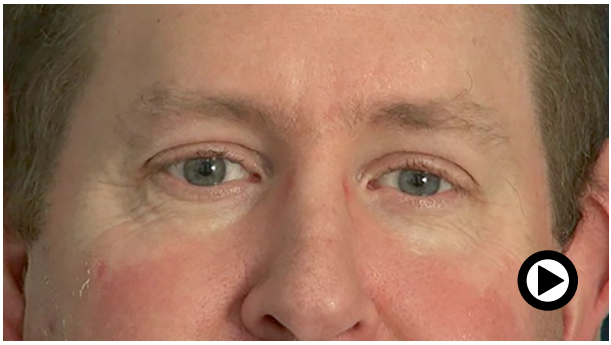

Fig. 4.10 Rapid head impulse test in a patient with bilateral vestibulopathy, a bilateral loss of function of the balance organs. During rapid head turns, the eyes initially move with the head, and the patient must make a quick eye movement back to center to fixate on the target again (video without sound) (▸ https://doi.org/10.1007/000-kxr)

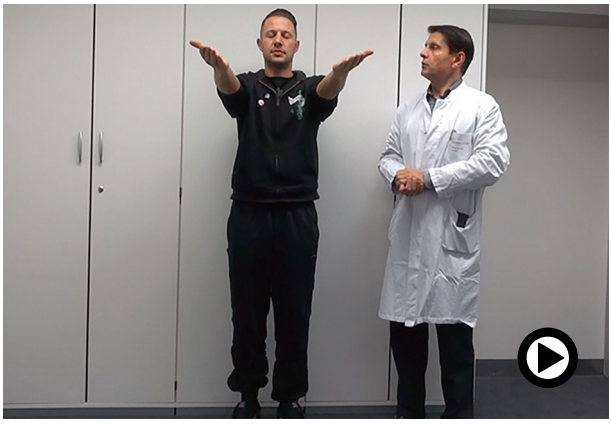

Fig. 4.11 Assessment of standing ability with eyes open and closed. With eyes closed, the patient sways significantly more. This is a typical finding in bilateral vestibulopathy, a bilateral loss of function of the balance organs (video without sound) (▸ https://doi.org/10.1007/000-kxs)

vestibulo-ocular reflex serves to stabilize the eyes during head movements. It relies on precise information from the rotational acceleration sensors (semicircular canals) of the vestibular organ. If these signals are absent, as in bilateral dysfunction of the balance organs, the eyes can no longer be stabilized during rapid head movements. Patients perceive this loss of function as blurred vision or as a bouncing/jittering of the visual field.

Other regulatory circuits originating from the vestibular organ are vestibulospinal reflexes, which project from the balance organs to the spinal cord and legs and are important for maintaining an upright posture while walking and standing. The brain relies on signals from three channels: the eyes, the rotational sensors (semicircular canals) and the sensors for gravity and linear movements (otolith organs) located in the balance organs as well as the sensors in the legs (muscles, joints and skin).

The diagnosis of bilateral vestibulopathy is often made only after a significant delay. The actual cause remains these days still unknown in about one-third of all patients, even after comprehensive diagnostics. Bilateral vestibulopathy can significantly impair quality of life, mainly due to the markedly increased risk of falls, movement-dependent dizziness and imbalance, and blurred vision during walking and rapid head movements. The most common known causes of bilateral vestibulopathy include vestibulotoxic medication side effects (damage to the balance organ), Menière's disease, genetic causes, and meningitis. Vestibulotoxic medications include certain antibiotics, such as aminoglycosides (gentamicin, streptomycin), which are used for severe infections, various cancer drugs (chemotherapeutics), and medications for treating cardiac arrhythmias (amiodarone). Traumatic brain injuries and skull fractures can also lead to a bilateral, but then sudden, loss of function of the balance organ. Bilateral vestibulopathy can also occur in combination with other functional deficits. These include various genetic cerebellar disorders (see Sect. 4.2.3), genetic inner ear hearing loss, and damage to the peripheral nerves.

Patient symptoms

Nearly all patients with bilateral vestibulopathy report unsteadiness while standing and walking, which usually begins gradually (video in Fig. 4.9). They also experience dizziness and sometimes have to take lateral compensatory steps while walking. These symptoms become noticeably worse in the dark or on uneven ground and can lead to (near-)falls. If the loss of function of the balance organs occurs abruptly on both sides or sequentially—first on one side, then the other—short episodes of spinning vertigo with nausea/vomiting may occur temporarily. Nearly half of all patients report visual instability (oscillopsia) as soon as they walk or move their head quickly. As a result, patients cannot read a street sign or recognize a face while walking; they must stop to see clearly. These symptoms are due to a reduced or absent vestibulo-ocular reflex (the connection between the inner ear sensors and eye movements). Head movements are then not adequately compensated by opposite eye movements. You can literally visualize this instability by filming with your

smartphone while moving it rapidly back and forth. Without image stabilization, the picture becomes shaky. Patients with bilateral vestibulopathy are typically symptom-free when sitting or lying down under static conditions. Several studies have also found evidence that bilateral vestibulopathy may be associated with impaired spatial orientation. Patients with bilateral vestibulopathy may secondary develop depression or anxiety disorders due to reduced quality of life. This is often accompanied by avoidance behavior and social withdrawal, to the point where affected individuals no longer dare to leave the house alone and limit social contacts to a minimum. This can be avoided by doctors who explain the cause and the course of the disease.

What does the doctor observe during physical and instrumental examination?

The clinical examination is guided by the reported symptoms. If imbalance, gait disturbance, and movement-induced blurred vision are reported, the focus is on the balance organ, but also on the nerves in the feet and the function of the cerebellum. Loss of function of the vestibular system can occur at the level of the semicircular canals, the vestibular nerves, or the brainstem nuclei.

Physical examination

If bilateral vestibulopathy is suspected, assessment of the function of the balance organs is paramount. The head impulse test allows evaluation of the vestibulo-ocular reflex at the bedside. The patient is asked to fixate on an object in the room (usually the examiner's nose) while the examiner performs rapid head turns to the left and right. If the vestibulo-ocular reflex is intact, the eyes immediately compensate for the head turn. If the vestibulo-ocular reflex is reduced, this immediate, rotation-sensor-mediated eye movement in the opposite direction is absent (see Sect. 2.1). The corrective eye movement—also called a catch-up saccade—occurs, which the doctor can observe (video in Fig. 4.10). This usually indicates damage to the balance organ itself. Namely in long-standing bilateral vestibulopathy, the catch-up saccades may be triggered so early during the head turn that they are no longer visible to the examiner; the reflex then appears falsely normal. Assessment of standing function (Romberg test) is performed under various conditions. Both the nature of the surface (firm vs. soft) and vision (eyes open vs. closed) can be varied (video in Fig. 4.11). When standing on a soft surface with eyes closed, mainly the balance organs are available to stabilize the body. Patients with bilateral vestibulopathy can fall during this test. All patients with dizziness and/or balance disorders should also undergo detailed

gait assessment. This includes testing normal gait as well as walking under challenging conditions, e.g., walking on an imaginary tightrope (walking on a straight line) with eyes open or closed, or walking on tiptoes or heels. Patients with bilateral vestibulopathy typically walk with their feet farther apart (broad-based gait). They walk unsteadily and look down at the ground to spot unevenness or obstacles in time. In these patients, the cerebellum, which is important for movement coordination, usually functions normally. Sensation in the feet is also preserved. However, in some conditions, both balance organs and the cerebellum may be affected simultaneously. These include genetic cerebellar disorders, which involve a combination of cerebellar dysfunction, cerebellar nystagmus, reduced sensation in the feet, and bilateral vestibulopathy, or, rarely but well treatable and should therefore not be overlooked, acute vitamin B1 deficiency.

Instrumental findings

The prerequisite for diagnosing bilateral vestibulopathy is evidence of bilateral partial or rarely complete loss of function of the sensors or the vestibular nerve. The video head impulse test and caloric irrigation allow detailed assessment of the rotational sensors and vestibular nerves. The function of all the semicircular canals in the balance organs is measured and the extent of dysfunction is calculated. The video head impulse test takes about 5 minutes to perform. Before the introduction of the video head impulse test, vestibular function was assessed only by caloric stimulation of the horizontal semicircular canals with warm and cold water, which induces spinning vertigo and characteristic nystagmus if the balance organs are intact. Measurement of body sway while standing using force plates allows quantification of postural instability. Hearing is tested to detect possible hearing loss. In addition, magnetic resonance imaging of the brain is performed to visualize the balance organ, vestibular nerve, brainstem, and cerebellum in detail. The main focus is on detecting tumors of the vestibular nerve, or rarely blood deposits on the vestibular nerve (siderosis), and inflammatory changes. A cerebrospinal fluid examination (lumbar puncture) is useful in rapidly progressive, unclear cases.

Therapy

Treatment of bilateral vestibulopathy depends on the underlying cause, but is only possible in rare cases. If the condition occurs suddenly or over a short period and, for example, is due to autoimmune processes, steroids can be tried, or in other cases of vitamin B1 deficiency, the loss of function of those brainstem nuclei that process signals from the balance organs (vestibular nuclei) can be reversed by rapid administration of vitamin B1. Such

a vitamin B1 deficiency can be caused by repeated vomiting, malnutrition or poor diet, or excessive alcohol consumption. If initial symptoms appear during ongoing treatment with aminoglycoside-containing antibiotics, the medication is switched, if possible, to another antibiotic that does not damage the balance organ. If a component of chemotherapy is damaging the balance organs, it is replaced if possible.

Physiotherapy treatment
The goal of physiotherapy is to alleviate symptoms, prevent falls, and improve mobility in daily life. Balance physiotherapy plays a crucial role here. Targeted movement exercises help the brain adapt to the new situation and simultaneously promote body stability and fine motor skills. In some cases, inpatient neurorehabilitation may be necessary. Physiotherapy also aims to develop a home exercise program tailored to the patient. If this is followed consistently (ideally daily and lifelong), it also promotes stability in standing and walking and alleviates symptoms.

Other therapeutic approaches
A balance prosthesis (the vestibular implant, analogous to cochlear implants for advanced hearing loss) is currently under investigation but is still far from being routinely used in patients with bilateral vestibulopathy. Some patients also benefit from a specially developed belt system that continuously monitors upper body deviation and alerts the wearer with vibration when a certain degree of body tilt is exceeded. This enables the affected person to correct this deviation.

Course (natural vs. under therapy) and response to treatment
A prognosis can only be made if the underlying cause has been identified. This is nowadays possible in about 75% of affected patients. While some conditions allow (rapid) recovery of vestibular function with treatment (e.g., with steroids or vitamin B1 supplementation), in other cases, prompt intervention can only prevent further deterioration (e.g., by discontinuing aminoglycoside therapy). Usually, after an initial phase of continuous deterioration, a plateau phase with persistently poor function is reached. If a hereditary disease (e.g., of the cerebellum) is present, further deterioration is usually expected. In the case of Menière's disease, the goal is to prevent further episodes and preserve remaining vestibular function. Treatment of Menière's disease is usually possible nowadays (see Sect. 4.1.3). After meningitis, even if the infection is adequately treated, a persistently reduced function of the balance organs is to be expected.

4.1.5 Recurrent Short Episodes of Vertigo Out of Nowhere (Vestibular Paroxysmia)

In Brief

Vestibular paroxysmia is a rare but well-treatable condition. It is characterized by brief, seconds- to few-minutes-long, uniformly occurring episodes of vertigo that usually arise out of nowhere without any trigger and can occur up to 100 times per day. In rare cases, tinnitus may also occur, which can sound like a typewriter. Typically, a small artery presses on the vestibular nerve, leading to a disturbance of its electrical insulation. As a result, short circuits occur, leading to brief episodes of dizziness. Important to note: Since vessel-nerve contacts can also be detected in 40%! of healthy individuals on MRI, the diagnosis must be made independently of this finding. The treatment of choice, and also necessary to confirm the diagnosis, is medication that is actually approved for the treatment of epilepsy. Nowadays, the well-tolerated drug lacosamide is used ("off-label" prescription, nowadays the treatment of choice), which reduces the excitability of the nerves. If patients respond convincingly to this treatment, the diagnosis is practically confirmed. The medium- and long-term outlook for patients is generally favorable: on the one hand, the condition is well treatable, and on the other, spontaneous improvement often occurs after several months.

A Typical Case

You have been suffering from very brief, recurrent episodes of vertigo for six months. These episodes just come out of nowhere. Initially, you had one episode per day, but in the past four weeks, you have had up to ten such episodes per day. If the episodes last a bit longer, you also experience mild nausea. You have also noticed that during more severe episodes, the images in front of your eyes jump.

Because of these symptoms, you have already seen your general practitioner, who could not find any pathological findings during his examination. He refers you to a neurologist.

The neurologist asks you about your symptoms, which you describe as above. She also wants to know how long the episodes last. You report a duration of seconds to a maximum of one minute. You are asked about triggers, and you state that you have not noticed any. Now you are asked about possible additional symptoms, especially hearing loss, headaches, sensitivity to light or noise. You say you do not have any of these.

The neurologist refers you to a radiologist to have an MRI of the head performed with and without contrast agent. This examination shows that a small artery is pressing on the right vestibular nerve (note: this is also found in 40% of healthy individuals!).

With the images and the findings, you return to your neurologist. She now explains your probable condition. She assumes that the brief episodes of dizziness are caused by the blood vessel irritating the vestibular nerve, resulting in a

kind of electrical short circuit, which you perceive as brief episodes of dizziness. This is called vestibular paroxysmia.

- **What does the treatment look like?**
  The neurologist prescribes you a medication that is normally used to treat epilepsy (nowadays treatment of choice: lacosamide, "off-label" prescription). It is important to emphasize that you do not have epilepsy. Over the next four weeks, the dose is gradually increased so that you end up taking one tablet in the morning and one in the evening. To assess the effect of the treatment, you should keep a dizziness diary in which you record how many episodes of vertigo you have per day.
- **Follow-up after four weeks**
  From week three, you have already noticed that you rarely have episodes of vertigo, i.e., at most one episode per day. Your neurologist is satisfied with the progress. You have also tolerated the medication well, except for some initial increased fatigue which was only transient. You should continue the treatment.
  After another two months, you see your neurologist again. You are pleased to report that you have not had a single episode in the past four weeks.
  The medication is continued, and after you have been episode-free for six months, the daily dose is gradually reduced. Fortunately, even after completely discontinuing the medication, you do not experience any further dizziness, so you no longer need the medication.

Case Example 2

In the video in Fig. 4.12, a patient with vestibular paroxysmia describes her symptoms before treatment, and in the video in Fig. 4.13 during treatment.

Fig. 4.12 Patient with vestibular paroxysmia, experiencing recurrent brief episodes of vertigo out of nowhere before treatment (▸ https://doi.org/10.1007/000-kxt)

Fig. 4.13 Patient with vestibular paroxysmia, experiencing recurrent brief episodes of vertigo out of nowhere during treatment (▸ https://doi.org/10.1007/000-kxv)

Patient Symptoms

In vestibular paroxysmia, sudden episodes of rotational vertigo or, more rarely, swaying dizziness occur, lasting seconds to a few minutes, sometimes a bit longer. These episodes are relatively uniform. The vertigo episodes may be accompanied by unsteady gait, rarely by tinnitus that can sound like a typewriter, or by hearing loss. The brief episodes of vertigo usually occur without a trigger. Occasionally, they are dependent on head movements or can be induced by rapid breathing (hyperventilation).

What Patients Should Pay Special Attention To

1. How long do the episodes last? They usually range from seconds to rarely a few minutes.
2. Do the episodes occur spontaneously or are there triggers? It is especially important to also consider peripheral paroxysmal positional vertigo, which, in contrast to vestibular paroxysmia, is triggered by changes in head position. Orthostatic dizziness, which typically occurs when standing up quickly from lying or sitting, is also characterized by brief transient episodes of dizziness which improve when lying or sitting down or start walking.
3. Are there any accompanying symptoms?

   - Rarely originating from the ears: impairment of hearing, tinnitus (typically so-called typewriter tinnitus)

- Signs of panic attacks would be episodes with intense anxiety and discomfort, starting abruptly with at least four of the following accompanying symptoms: palpitations, sweating, trembling, shortness of breath, feeling of choking, chest tightness, nausea, dizziness, hot flashes or chills, numbness or tingling, fear of losing control or fear of dying. Panic attacks may mimic vestibular paroxysmia so that this is an important differential diagnosis.

4. Are there any other symptoms, e.g., facial twitching (hemifacial spasm) or facial pain (trigeminal neuralgia)? In these cases, other cranial nerves may be affected.
5. How frequent are the episodes per day or week? This is important for assessing treatment success.
6. Are there any previous or existing illnesses?

- This could be a stroke in the area of the brainstem or cerebellum, which can lead to similar brief episodes of vertigo with a delay of months to years after the stroke (paroxysmal brainstem or cereellar episodes).
- Multiple sclerosis (paroxysmal brainstem or cerebellar episodes) can also cause brief episodes of vertigo.
- Patients with a benign tumor of the vestibular nerve (vestibular schwannoma) can develop such symptoms too.
- In rare cases, cardiac arrhythmias can lead to brief episodes of dizziness; however, these are often accompanied by blacking out or even loss of consciousness. Since cardiac arrhythmias are potentially serious, this needs a careful cardiological workup.

The doctor should receive this information.

What Can Patients Do If They Have Recurrent Episodes of Dizziness?

1. Patients should pay attention to the points mentioned above and keep a diary.
2. Patients should see their general practitioner first.
3. If the general practitioner is unsure or the treatment is ineffective, the patient should see a neurologist or ENT specialist. If heart problems already exist, a cardiological examination is advisable.

Cause

The cause of vestibular paroxysmia is a blood vessel (usually an artery) pressing on the vestibular nerve, which runs from the balance organ in the inner ear to the brain. The vestibular nerve is irritated by the pressure and

pulsations of the blood vessel. This leads to electrical short circuits between the nerve fibers, which patients perceive as brief episodes of vertigo.

What Does the Doctor See During Physical and Instrumental Examination?

During the physical examination, other causes of the symptoms are ruled out, especially peripheral paroxysmal positional vertigo (formerly called benign paroxysmal positional vertigo). Sometimes, the episodes of vertigo in vestibular paroxysmia can also be provoked by the doctor by telling the patient to breathe rapidly (hyperventilation).

If a patient is seen during an episode, or more commonly today, if a video recorded by the patient is viewed, twitching eye movements (nystagmus) are often observed. This supports the diagnosis.

The contact between vessel and nerve can usually be detected on MRI of the head. Important: such a vessel-nerve contact is also found in 40% of healthy individuals, so this imaging study is mainly used to rule out other causes. These include

- benign tumors of the vestibular nerve (vestibular schwannoma, formerly called acoustic neuroma),
- other types of tumors in this region,
- changes in the brainstem or cerebellum, especially previous infarcts or inflammatory changes, such as multiple sclerosis.

During the course of the disease, it may be useful to perform tests of the balance organ using the rapid head impulse test or ear irrigation with warm and cold water as well as hearing tests to check for damage to the vestibular or auditory nerves.

Brief Episodes of Dizziness—What Else Could It Be?

1. Peripheral paroxysmal positional vertigo: brief episodes of vertigo triggered by changes in position relative to gravity (see Sect. 4.1.1)
2. Space-occupying lesions in the region of the vestibular nerve: dizziness, postural imbalance, and hearing loss or tinnitus, rarely additional brief episodes of dizziness
3. Changes in the brainstem or cerebellum after a stroke or in multiple sclerosis (paroxysmal brainstem episodes)
4. Menière's disease: longer-lasting episodes of vertigo with hearing loss, tinnitus, or pressure in the affected ear (see Sect. 4.1.3)
5. Vestibular migraine: longer-lasting episodes of dizziness with headache, sensitivity to light or noise (see Sect. 4.2.2)

6. Panic attacks: episodes with anxiety, shortness of breath, palpitations (see above)
7. Transient circulatory disturbances (transient ischemic attacks): here, dizziness or vertigo can be accompanied by many central symptoms.
8. Cardiovascular diseases: cardiac arrhythmias can lead to brief episodes of dizziness or syncope with loss of consciousness. A hypersensitive carotid sinus can trigger symptoms during head turns or pressure on the neck in the area of the carotid artery.
9. Orthostatic dizziness: transient dizziness after standing up or rising, with or without blacking out, improving when lying down (see Sect. 4.4.1)
10. In rare cases: epileptic seizures or episodes of dizziness and hearing loss due to pathological pressure transmission to the inner ear (syndrome of the third mobile windows) (see Sect. 4.1.6)

Therapy

To treat vestibular paroxysmia, medications are administered that are actually used for the treatment of epilepsy, so-called anti-seizure medication (formerly called antiepileptics). Nowadays, lacosamide (dose: 100–400 mg daily, "off-label" prescription) is usually used because it is effective and much better tolerated than other, previously used anti-seizure medications.

To assess the effect of treatment, patients should keep a dizziness calendar. Follow-up visits to the doctor should take place every one to three months to adjust the treatment as needed.

Only in very rare exceptional cases, when the diagnosis is certain (patients respond convincingly to medication and the affected side can be clearly identified), in cases of unusually severe disease progression and intolerance to medication, is surgery performed to separate the blood vessel from the vestibular nerve.

Prognosis

A study on long-term outcomes shows that three-quarters of patients were symptom-free at follow-up after an average of about five years, half of them even without medication. Thus, the long-term course of the disease is also favorable in our experience.

4.1.6 Rare Cause of Peripheral Dizziness

This section describes a rare cause of peripheral dizziness. It often takes a long time for the diagnosis to be made in these cases, and such patients in particular have experienced an odyssey of doctor visits.

Third Mobile Windows Syndrome

Typical symptoms of third mobile windows syndrome, a rare disease, are brief episodes of vertigo or dizziness lasting seconds to a few minutes, triggered by changes in pressure such as lifting, coughing, straining, or even by certain sounds. These episodes may be accompanied by apparent movements in front of the eyes. In addition, many patients with this condition hear internal body sounds, such as eye movements or stomach gurgling in the affected ear; this is called autophonia.

A Typical Case

A 53-year-old patient first notices brief spinning vertigo with moving images while shoveling snow. During a cold, he subsequently experiences the same symptoms when blowing his nose. At Christmas, things get lively and quite loud: he then notices that certain loud noises also trigger these symptoms.

He first goes to his general practitioner, who finds nothing unusual and refers him to an ENT specialist. After the patient describes the above symptoms, the ENT specialist is able to trigger the symptoms by having him strain against a closed nose and closed vocal cords (straining the abdomen) and observes, using special goggles, that the eyes are jumping. When pressure is applied to the left ear, the same findings occur. The hearing test shows that low tones are perceived via the bones better than normal in the left ear, so-called negative bone conduction. He is referred to a specialized center, where further examinations, including a CT scan of the temporal bone, are performed. All these findings allow for a precise diagnosis: the bony covering over the left anterior semicircular canal of the balance organ is partially missing. The patient is advised to avoid the triggering factors. Fortunately, following this advice the symptoms improve over the following weeks, so that surgery is not necessary.

Cause

Normally, the balance and hearing organs are completely sealed within the hardest bone in the human body, except for two small bony openings to the middle ear. This bone is therefore called the petrous (this means rock in Latin) bone. If there is a bony defect, pathological pressure is transmitted to the inner ear, which can lead to vertigo, dizziness and hearing problems, so called autophonia.

Examinations

It is important to first consider this rare and still often overlooked disease at all. If this condition is suspected, a prompt referral to a specialist is advisable.

The following examinations are performed:

1. Attempt to trigger the symptoms and certain eye movements by changing pressure, including straining against a closed nose and vocal cords as well as via the middle ear as well as by certain sounds
2. Testing of hearing ability which typically shows an abnormally good hearing of lower frequencies via the bones
3. Testing the function of the otolith organs, i.e., the sensors for linear movements and gravity (linear acceleration sensors) in the inner ear which shows higher amplitudes than in healthy individuals
4. High resolution CT scan of the ear (petrous bone) to visualize the bony defect

What Does the Treatment Look Like?

1. The patient is informed about the mechanism and cause of the disease.
2. For mild and tolerable symptoms, it is recommended to avoid pressure changes as much as possible.
3. For severe symptoms and impaired quality of life, surgery to cover the defect is possible. However, this should only be performed at a specialized center by a surgeon very experienced in this field.

4.2 Central Vertigo and Dizziness Originating from the Brainstem and Cerebellum

A wide range of different causes can lead to vertigo and dizziness originating from the brain (central vertigo and dizziness). This is the case in up to a quarter of all patients who present to the emergency department with sudden onset of symptoms. Which symptoms are most prominent for those affected, what the prognosis is, and what treatment options are available depends crucially on the underlying disease. Central vertigo and dizziness can be classified according to various criteria. The time course or the underlying disease mechanism may be decisive. Accompanying symptoms and the presence of factors that promote or trigger the occurrence are also important.

The time course of central causes of dizziness can vary:

1. Acute and persistent over hours to days
2. Recurrent and transient, i.e., episodic
3. Persistent over weeks to years, i.e., persisting

Various causes can lead to central vertigo and dizziness. This includes damage to specific parts of the brain, e.g., due to a stroke, premature aging of nerve cells (neurodegeneration), or genetic and metabolic disorders.

Patients can assist the doctor in making the correct diagnosis by paying attention to the frequency, duration, and provoking factors of their vertigo or dizziness and also reporting any other symptoms they have noticed in addition.

Course of dizziness symptoms:

1. If these occur acutely and persistently, it may be an acute circulatory disorder in the brainstem or cerebellum (stroke, see Sect. 4.2.1) or a new or an overdose of a medication or, rarely, a severe vitamin B1 deficiency (see Sects. 4.1.4 und 4.2.3).
2. If the vertigo or dizziness occur repeatedly and are transient each time (episodic), vestibular migraine (see Sect. 4.2.2) should be considered in particular. However, hereditary diseases of the cerebellum with intermittent worsening (so-called episodic ataxias; see Sect. 4.2.3.1) or rarely tumors in the cerebellum or brainstem (for instance, central positional vertigo) can also present in this way.
3. If the dizziness is persistent over weeks or months, it is referred to as persisting dizziness. This also includes genetic diseases that, for example, affect not only the cerebellum and the areas involved in Parkinson's disease but also the balance organs and the nerve fibers in the feet and hands, and may be accompanied by rapid downward eye movements (downbeat nystagmus).

It is also important whether the patient has noticed any other symptoms:

1. Additional paralysis or impaired coordination of an arm and/or leg, difficulty speaking clearly, or double vision indicate a central lesion, if acute most often a stroke in the brainstem or cerebellum.
2. Recurrent episodes of headaches with nausea and vomiting, sensitivity to noise and light suggest migraine (vestibular migraine), namely if the patient is suffering from ongoing migraine or has a history of migraine.
3. If the patient has previously experienced repeated vomiting, malnutrition, or has undergone a weight reduction procedure, the possibility of acute vitamin B1 deficiency should be considered. This is potentially a very serious condition which on the other hand can be well treated.

In the following sections, important central causes of vertigo and dizziness are presented and it is described how they can be distinguished from one another. Treatment options and prognosis are also discussed.

4.2.1 Acute Stroke: Sudden Vertigo or dizziness, Tendency to Fall, Gait Disturbance, and Often More

In Brief

A stroke is an acute disorder affecting specific brain structures, with every fifth stroke involving the cerebellum or brainstem. When vertigo is the predominant symptom, affected individuals often report a sudden onset of intense, persistent spinning or swaying vertigo, dizziness, imbalance (so-called acute imbalance syndrome) a tendency to fall, and/or a gait disturbance. These symptoms are usually assessed in an emergency department. Determining the cause (stroke to due impaired circulation or bleeding) and acute treatment are the priorities. Here, a thorough examination of eye movements and the function of the vestibular organs at the bedside is also very helpful. This is supplemented by imaging of the brain and cerebral vessels. If the symptoms are correctly identified very quickly, various, often highly successful treatment options are available within the first 4.5 hours with so-called thrombolysis to a maximum of 24 hours (with so-called mechanical recanalisation). Therefore, it is very important that affected individuals seek medical attention as early as possible, ideally in an emergency department. After the acute phase, the focus shifts to identifying the underlying cause (for instance, atherosclerosis or from the heart), treating stroke-related impairments (mainly with physiotherapy) and preventing recurrent events. The prognosis is often favorable, but the recovery phase with intensive balance training can extend over several months.

Case Study 1

One evening around 10 p.m., just before going to bed, you suddenly notice significant unsteadiness while walking and a swaying dizziness as well as vertigo. There is also a strong tendency to fall to the left. A few minutes later, you feel nauseous and have to vomit. Two or three days earlier, you had noticed a new type of pain on the left side of your neck for the first time. You decide to lie down and quickly fall asleep. Around 6 a.m., you wake up feeling very thirsty. When you try to get up to drink a glass of water, you are markedly unsteady while walking and have to hold onto the walls to avoid falling. As you reach for a glass of water, you notice that you see double and that vision is blurred. When drinking, you choke severely. Because of these symptoms, you decide to call the ambulance and are taken to the emergency department of the nearest hospital.

In the emergency department, you are immediately interviewed and examined by a neurologist. You tell the doctor that you had already noticed gait unsteadiness and vertigo about five hours earlier. When asked, you also confirm pronounced and persistent nausea as well as repeated vomiting. When asked about signs of paralysis, you deny any, and you have no trouble finding the right words or understanding the neurologist during the conversation. The doctor asks about your medical history and medications. You mention a long-standing type 2 diabetes and high blood pressure; both are being treated with medication. A year ago, at the age of 75, you quit smoking due to lung disease.

You are now examined by the neurologist. The doctor asks you to look straight ahead. She notices that your eyes slowly drift downward and then quickly jerk upward, which is called upbeat nystagmus. The neurologist then asks you to follow her finger with your eyes and has you look to the side. As you try to hold this position with your eyes, you notice the image trembling. The doctor observes that your eyes move back toward the center and then jerk outward. These findings indicate a dysfunction of the cerebellum or brainstem. The neurologist also notices during the examination that your left eyelid droops and your left pupil is smaller than the right. When asked, you confirm that this is new. The doctor looks into your mouth and finds that the muscles in your throat on the left side are drooping. The neurologist now asks you to touch your nose with your left index finger while your eyes are closed. You find that you can hardly do this and repeatedly miss your nose. Since being picked up by the ambulance, you have not stood or walked. When the doctor now lets you stand without assistance, you notice a pronounced pull to the left side, you are about to fall, and have to be caught. The neurologist then tests the strength in your arms and legs, which is unremarkable. She also examines whether you can perceive touch, pain, and temperature well on both sides. It is noted that your temperature sensation is significantly reduced on the left arm and leg. Your blood pressure is currently markedly elevated at 190/100 mmHg.

- **What further examinations are performed now?**
  Immediate brain imaging is performed with computed tomography (CT) and so-called CT-angiography to visualize the arteries. This is used to look for evidence of a circulatory disorder, bleeding, or tumors in the brain. This examination is unremarkable. However, the subsequent CT angiography of your cerebral vessels shows an occlusion of an artery that is important for supplying blood to parts of the brainstem and cerebellum.
- **What is the cause of the symptoms?**
  The doctor discusses the findings with you immediately after the examination and tells you that she suspects a stroke and considers the occluded artery to be the cause. She also informs you that there is no evidence of a brain hemorrhage or brain tumor.
- **What does the treatment look like?**
  The doctor discusses treatment options with you. She clearly advises against attempting to open the occluded blood vessel with medication (thrombolytic therapy), as the symptoms have already been present for too long. Instead, a treatment with aspirin and a blood lipid lowering drug is started to reduced the risk of further strokes. You are admitted to the stroke unit for further monitoring and evaluation. Your heart rhythm, blood pressure, and pulse are continuously monitored, and you undergo regular neurological examinations. The following day, another brain imaging is performed, this time using magnetic resonance imaging. This examination reveals a strpke in the lowest part of the brainstem (so-called medulla oblongata). The cause of the vessel occlusion already detected in the CT scan is diagnosed as a hemorrhage into the vessel wall of the affected artery (so-called dissection). The doctor again discusses the findings with you and explains that the tearing of this blood vessel led to your stroke and also very well explains your new neck pain, the drooping left eyelid, and the smaller left pupil. The doctor asks you about any manipulation of the head, including chiropractic treatments or falls. You deny this. The doctor tells you that such

a tearing of a blood vessel can also occur spontaneously, i.e., without external influence. From now on you should avoid any extreme head movements.

- **What is the course of the illness?**
  Over the next few days, you notice the first, slow improvements. The nausea subsides, the blurred and double vision lessens. However, you still have difficulty swallowing, and walking and standing remain very difficult. You are now transferred to inpatient neurorehabilitation. After an three-week stay, you feel significantly better and are discharged home. You then receive regular balance training, swallowing therapy, and fine motor training for your left hand. At your follow-up examination three months later, the doctor tells you that she expects further, gradual improvements and emphasizes the importance of regular therapies.

Case Study 2

In the video in Fig. 4.14, a patient with stroke as the cause of his vertigo symptoms (Wallenberg syndrome) describes his medical history.
Figure 4.15 shows a magnetic resonance image depicting an infarct in the left brainstem.

Cause

A stroke is understood as an acute circulatory disorder affecting specific parts of the brain. Less commonly, a brain hemorrhage is the cause. An acute circulatory disorder is usually caused by a blood clot that prevents the supply of oxygen-rich blood to a defined brain area. This leads to the loss of specific brain functions. For example, an occlusion of the left middle cerebral artery leads to paralysis of the right arm and leg as well as a speech disorder. Such a clot can form in the heart in cases of atrial fibrillation and travel with the blood to the brain. It can also form locally in the affected brain vessel in patients with artherosclerosis. The latter occurs, for example, when the blood vessel is narrowed by changes in the vessel wall and deposits, and then becomes completely occluded. A dissection, as in the case described, is a rare cause of stroke but can nowadays easily be diagnosed by Doppler sonography and MRI.

Frequency

In Germany, about 200,000 patients suffer a stroke each year, with approximately one in five strokes affecting brain structures that are crucial for balance (see case studies). These include the brainstem and the cerebellum. The prognosis after a stroke varies: while about 40% of patients make a full recovery, about 35% are left with permanent damage, and about 25% die as a result of the stroke.

Fig. 4.14 Medical history of a patient with stroke as the cause of his vertigo symptoms (Wallenberg syndrome) (▸ https://doi.org/10.1007/000-kxw)

Symptoms

In the acute treatment of stroke, recognizing typical stroke symptoms and immediate presentation to the emergency department are of great importance. These include sudden onset of symptoms such as hemiparesis, speech or language disturbance, sudden visual disturbance in one or both eyes, double vision, hemisensory loss, severe headache (usually in the back of the head), or intense vertigo or dizziness. If such symptoms occur suddenly, every minute counts; ideally, the ambulance should be called promptly or the nearest emergency department visited. Acute vertigo and dizziness are one of the most common reasons for presenting to the emergency department. However, only a small percentage of those affected (3–4%) actually have a stroke. Timely identification and treatment of these patients is a priority. This can improve the prognosis after a stroke and prevent or mitigate long-term damage. The correct diagnosis is usually made very quickly if, in addition to vertigo, other typical stroke symptoms are present, such as hemiparesis or slurred speech. In cases of isolated vertigo or dizziness, i.e., in the absence of other symptoms (e.g., double vision or impaired coordination of arms or legs), the evaluation is significantly more difficult. Estimates suggest that about one in three strokes presenting as acute vertigo or dizziness is initially not recognized as such.

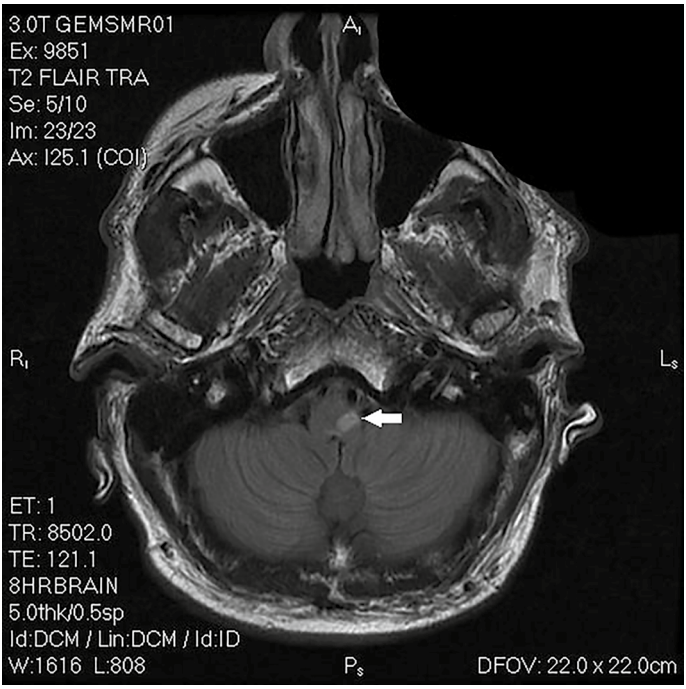

Fig. 4.15 Magnetic resonance image showing an infarct in the left brainstem (bright spot with *arrow*)

Patient Complaints

Acute spinning vertigo, swaying or imbalance, and gait disorders, which usually occur spontaneously and suddenly and last for hours to days, are one of the most common initial symptoms of a stroke of the cerebellum or brainstem. The symptoms depend greatly on which areas of the brainstem or cerebellum are affected. Patients usually avoid any movement of the head and body in general, as movement increases the symptoms. Instead, they lie quietly on their back with their eyes closed. Severe nausea is common and often leads to vomiting (especially with movement). Gait unsteadiness with a tendency to fall and jumping of the eyes (nystagmus) can also be observed. The balance disorder can be so severe that patients are unable to stand or walk independently, or would fall if they tried. The combination of these symptoms indicates an acute disturbance of the balance system (acute vestibular or acute imbalance syndrome). Other symptoms of a stroke of the brainstem or cerebellum include slurred speech (dysarthria), swallowing disorder (dysphagia), double vision (diplopia), or blurred vision or paresis, impaired coordination or sensation of arms or legs. The latter are typical for

a stroke of the cerebellum. Recognizing these additional symptoms helps to quickly and reliably diagnose stroke; patients have to be explicitely ask for such symptoms by the doctor. In some patients, the above symptoms have already partially or completely resolved by the time they arrive at the emergency department; this is referred to as a transient ischemic attack (TIA).

If acute vertigo occurs only in certain body positions, this is referred to as position-dependent vertigo. The vast majority of patients with such vertigo triggered by specific movements or positions do not have a stroke, but rather peripheral paroxysmal positional vertigo (see Sect. 4.1.1).

What Patients Should Pay Special Attention To

Stroke symptoms usually have an abrupt onset. Early recognition of symptoms and immediate presentation to an emergency department are crucial for further treatment and prognosis. Patients should therefore familiarize themselves with symptoms that may indicate a stroke. These include, in particular, new, unilateral weakness of arm, leg or face, sudden visual, speech, or language disturbance, sudden onset of new headache, or new vertigo or dizziness.

After a stroke, it is crucial that patients avoid risk factors for a recurrent stroke as much as possible. This includes consistent use of prescribed medications, especially aspirin or oral anticoagulants (depending on the cause of the stroke) and those that lower blood pressure or blood lipids and optimize blood sugar. It is also important to engage in regular physical activity (e.g., regular walking or swimming) and to eat a balanced diet. If patients smoke, they should consult their doctor for support in quitting. If the patient's family notices increased nighttime snoring and breathing pauses, this should be reported to the doctor. This can be a contributing factor to high blood pressure and should be investigated and treated accordingly.

Initial Assessment

If a patient presents to the emergency department with acute spinning vertigo or dizziness, a tendency to fall, or gait disturbance, the urgency of evaluation and treatment is determined. If symptoms of a possible stroke are present, patients are prioritized and immediate evaluation is initiated. If the emergency services bring the patient to the emergency department, the initial assessment is performed at the scene. In suspected acute stroke, specialists from various disciplines, including physicians, emergency nurses, and diagnostic staff, jointly perform the initial assessment. Evaluation and treatment of stroke patients should ideally take place in a specialized stroke unit, where standardized stroke assessment and treatment according to current

guidelines and recommendations are ensured. In cases of suspected stroke, the ambulance normally takes the patient to a hospital with a stroke unit.

For the initial assessment, it is very important to know the exact timeline of the symptoms. This includes the time onset and duration of symptoms and when the patient was last seen without these symptoms. This is particularly important if the symptoms were first noticed in the morning. The main goal of the initial assessment is to distinguish between acute vertigo or dizziness caused by a stroke or caused by a dysfunction of the vestibular organ (e.g., vestibular neuritis/acute unilateral vestibulopathy).

Physical Examination

The physical examination in suspected stroke is performed in a standardized manner. Typical findings are systematically recorded within a few minutes to assess the severity and extent of the findings. This allows an estimation of whether acute treatment is possible and necessary. In cases of acute vertigo. dizziness and/or balance or gait disturbances, special attention is paid to eye movements, coordination, and balance. By carefully examining various eye movements, the physician can determine at the bedside whether a stroke or a vestibular organ dysfunction is present. Acute unilateral hearing loss for the first time can also suggest a stroke. The gait pattern and stability while standing are also assessed, and the physician observes whether the patient can stand and walk independently. If the patient is unable to stand or sit independently, this also suggests a stroke.

Instrumental Findings

In suspected stroke-related acute vertigo, brain and cerebral vessel imaging is usually performed immediately after arrival in the emergency department, typically within the first 15–30 minutes, most often using computed tomography (CT) and CT-angiography. The advantages of CT are its universal availability, short examination time, and wide range of applications. The results are used to decide whether acute treatment is indicated. The main questions are whether there is an occlusion of a cerebral vessel and/or whether brain tissue damage is already detectable. The CT scan thus serves to detect a stroke and simultaneously search for other causes (e.g., brain hemorrhage or brain tumor; see Fig. 4.16). Using modern CT imaging and iodine-containing contrast agents, acute hypoperfusion of the affected brain areas can usually be demonstrated (CT perfusion). However, the resolution of CT is limited, and strokes that present with acute vertigo, dizzines and/or balance disturbance, i.e., are presumably located in the brainstem

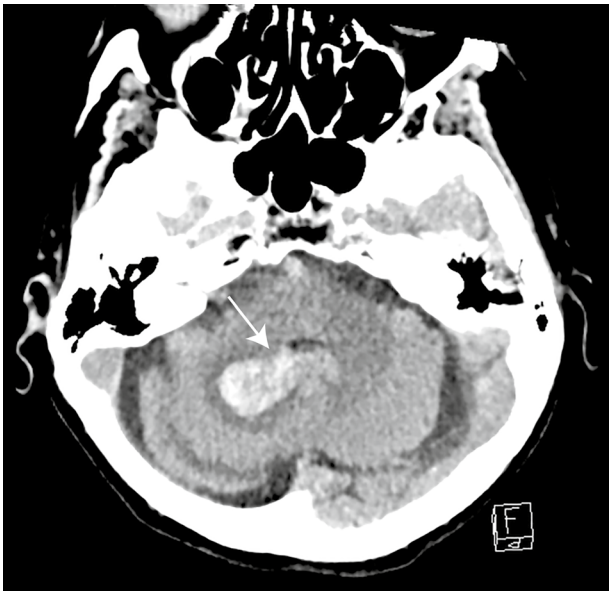

Fig. 4.16 Acute hemorrhage in the cerebellum. The 89-year-old patient noticed a sudden swallowing and speech disturbance as well as clumsiness on the right side of the body. In the emergency department, markedly elevated blood pressure was observed, jumping eye movements (nystagmus) were present both in the straight-ahead and lateral gaze, and movements with the right arm were no longer accurate. The cause was identified in the brain CT as a hemorrhage in the cerebellum (*white arrow*) under ongoing anticoagulation. Blood pressure was subsequently significantly lowered, and anticoagulation was stopped. Inpatient rehabilitation was initiated (Source of image: Radiology, Kantonsspital Baden)

or cerebellum, are often obscured by imaging artifacts. Small acute strokes in the brainstem or cerebellum are therefore usually not detected with this imaging. Alternatively, initial imaging can also be performed using magnetic resonance imaging (Fig. 4.17). This imaging technique is much more detailed; an acute circulatory disorder can be detected with high certainty after 72 hours but often not in the acute phase which can be misleading. Further MRI is more complex and time-consuming, and is not available around the clock in most hospitals.

During the hospital stay, a detailed assessment of balance is often performed (see Chap. 1).

Therapy

Acute treatment of stroke-related vertigo must be initiated quickly. Within the first 4.5 hours after symptom onset, dissolution of the blood clot can be

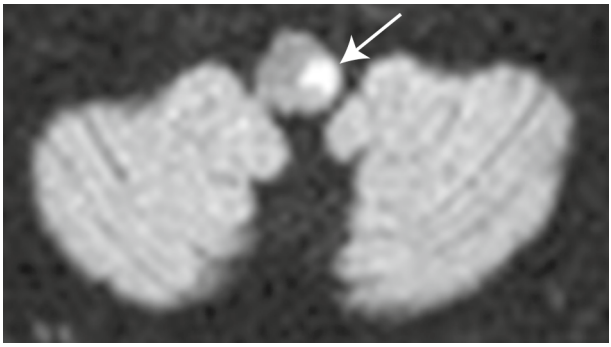

Fig. 4.17 Acute stroke in the brainstem of a 75-year-old patient with sudden onset of gait unsteadiness, vertigo, and swallowing disorder. Clinical examination revealed spontaneous nystagmus to the right; the patient was unable to stand without assistance. MRI shows an acute stroke(*white arrow*) in the lowest section of the brainstem (medulla oblongata). There was only a slow recovery with regained mobility. The cause of the stroke remained unclear (Source of image: Radiology, Kantonsspital Baden)

supported with an anticoagulant medication (intravenous thrombolysis with alteplase or tenecteplase). In cases of proven occlusion of major cerebral vessels such as the basilar artery or carotid artery, the clot can be mechanically removed using a catheter within the first 6 hours (under certain conditions up to a maximum of 24 hours) after symptom onset. Pre-existing conditions and circumstances that preclude such treatment must be known. These may include recent surgeries, treatments, or pre-existing conditions that increase the risk of bleeding. If the patient cannot provide this information, relatives must be consulted.

Course (Natural vs. Under Therapy) and Response to Therapy

The location of the circulatory disorder has a decisive influence on the symptoms and recovery. About one in seven treated patients benefits significantly from acute thrombolysis within the narrow treatment window of 4.5 hours. Mechanical removal of a clot leads to a clinically relevant improvement in one in four to five patients. With effective acute therapy, rapid improvement can be expected. Whether patients with stroke-related vertigo benefit long-term from acute therapy has not been definitively established yet but is likely. Acute therapy is recommended if there is a clinically relevant impairment (e.g., inability to stand and walk independently) and no exclusion criteria are present, provided the patient consents.

If acute treatment is not possible or desired, secondary prevention is initiated, i.e., minimizing the risk of suffering another stroke. This consists, depending of the etiology of the stroke, aspirin or clopidogrel or oral anticoagulants plus lowering blood lipid levels (cholesterol/lipid-lowering agents). Rehabilitative measures are carried out on an outpatient, day-care, or inpatient basis, depending on the extent of the impairment. In cases of vertigo and dizziness as the leading symptoms, therapeutic measures to improve balance and fine motor skills should be prioritized.

The search for the cause of the stroke follows a specific protocol. Ultrasound of the cerebral vessels and the heart as well as continuous heart rhythm monitoring over 72 hours (long-term electrocardiogram) are standard investigations. Despite these examinations, the cause of the stroke remains unclear in about one in three patients. Medication is prescribed in accordance with the examination results, e.g., oral anticoagulants in the case of proven irregular heart rhythm or atrial fibrillation. In some cases, surgery or neuroradiological interventions with a stent are performed, for example, to correct a narrowing of the carotid artery.

Complications from treatment or the stroke itself can delay or even completely thwart the recovery process. Common sequelae include pneumonia due to stroke-related swallowing disorders, development of depression, or painful abnormal postures of the affected limbs (spasticity). Worsening of stroke-related symptoms within the first 24 hours is observed in about one in seven patients. In such cases, repeat investigations and, if necessary, additional therapies are required.

Recovery from a stroke generally progresses slowly and can take weeks to months. However, it can be significantly promoted by intensive therapeutic measures (physiotherapy, occupational therapy, speech therapy). The natural course after stroke-related vertigo is usually favorable: persistent vertigo often resolves within a few days, and dizziness, imbalance and gait unsteadiness also usually improves to the extent that independent walking with or without aids becomes possible again. Nystagmus and blurred vision decrease continuously over days and can also be treated with medication. About 40% of patients make a full recovery. However, many patients require long-term regular physiotherapy, occupational therapy, and/or speech therapy, as well as regular specialist and primary care follow-up after a stroke.

4.2.2 Recurrent episodes of vertigo or dizziness with Headache, Sensitivity to Light or Noise: Vestibular Migraine

In Brief

Headaches can be accompanied by swaying dizziness or even spinning vertigo. The headache is considered migraine if it lasts for hours and is associated with nausea, sensitivity to light, or sensitivity to noise. If migraine is often accompanied by vertigo or dizziness, it is referred to as vestibular migraine. It is important to note that not every headache is a migraine, and not every headache with vertigo is a vestibular migraine. In vestibular migraine during episodes, subtle eye movement disorders (and even between the episodes) and marked balance disturbances can often be detected. For the diagnosis of vestibular migraine, careful patient history, physical examination, and functional testing of the vestibular organs and hearing are important. Another condition to consider in cases of recurrent vertigo episodes is Menière's disease. Patients may suffer from both conditions. If the cause is unclear, magnetic resonance imaging of the brain can be done to exclude other diseases. Treatment of an acute episode of vestibular migraine is identical to the usual therapy for migraine headache. The combination of pharmacological and non-pharmacological preventive treatment for vestibular migraine is recommended if episodes occur more than two to three times per month. This is also identical to other forms of migraine.

A Typical Case

Every two to three weeks, you experience sudden dizziness that usually lasts for several hours and is often accompanied by nausea and sometimes vomiting. Because of these episodes, you usually have to interrupt your current activity and lie down. Sometimes, together with the dizziness episodes, you also notice pulsating headaches, sometimes on the right side of you head, sometimes in the back of your head. An evaluation by your general practitioner did not yield a diagnosis; he is now referring you to a dizziness clinic.

At your first appointment the neurologist asks you about your symptoms. You again mention your dizziness episodes, which have been occurring since you were 48 years old and are sometimes accompanied by headaches. The doctor now asks you to describe your headaches in more detail and inquires about when you first started experiencing such headaches. You describe regular, mostly left-sided headaches lasting six to twelve hours and of high intensity since you were 15 years old. Upon further questioning, you confirm that these headaches are often accompanied by pronounced nausea as well as vomiting and a marked increased sensitivity to noise and light as well as withdrawal from physical activities, such as climbing stairs, during such episodes. You tell the doctor that acquaintances have previously suggested that you might be suffering from migraine. The doctor now asks you about other symptoms associated with the headaches. You deny accompanying visual disturbances (flickering, zigzag patterns), speech disturbances, or sensory disturbances. When asked, you mention that the headaches, but not the dizziness, respond well to painkillers.

The neurologist now examines you. At the current time, you have no headache and no dizziness. The medical examination reveals no abnormalities; in particular, balance and eye movements are normal.

- **What further examinations are now performed?**
  Because of the recurrent episodes of dizziness and headache, the doctor recommends imaging of the brain using magnetic resonance imaging. This examination reveals no abnormalities. The doctor asks you to return to the clinic after the MRI has been performed. He discusses all findings, the diagnosis, and the treatment with you.
- **What is the cause of the symptoms?**
  The doctor explains to you that the recurrent headache episodes with nausea, vomiting, sensitivity to noise and light, and withdrawal tendencies are caused by migraine. He discusses with you the connection between migraine headaches and dizziness episodes, which in your case sometimes occur simultaneously. The recurrent dizziness can also be an element of migraine. The doctor emphasizes that the unremarkable findings are consistent with vestibular migraine. During the conversation, you express relief that a cause has been found and inquire about treatment options, as you are significantly limited in your professional and private life due to the recurrent episodes.
- **What does the treatment look like?**
  The doctor recommends that, due to the regular occurrence of dizziness and headache episodes, you take a preventive medication. Various medications are possible for this purpose. The doctor first recommends a trial of therapy with a so-called beta blocker. You agree and schedule a follow-up examination in three months. Furthermore, the doctor explains that additional non-pharmacological measures are very important in migraine and recommends that you exercise regularly and also perform relaxation exercises.
- **What is the course of the condition?**
  As agreed, you now take a beta blocker, go swimming regularly, and practice autogenic training. At the follow-up examination, the doctor asks you about side effects from taking the beta blocker. Apart from increased fatigue at the beginning, you have not noticed anything negative. The doctor now asks you about the course of the headaches and dizziness. You report that the frequency, intensity, and duration of the episodes have decreased by about 75%. When asked, you deny any further illness-related absences from work. You emphasize that you are very satisfied with the course and agree to continue the existing therapies unchanged. Another follow-up in the dizziness clinic will take place in six months.

Case Example 2

In the video in Fig. 4.18, a patient with vestibular migraine reports her medical history.

What Does Migraine Feel Like and What Are the Causes?

Both recurrent episodic headaches and recurrent episodic vertigo or dizziness are among the most common health complaints overall. While the

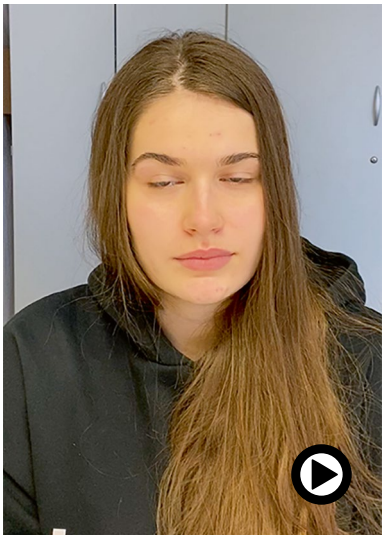

Fig. 4.18 Medical history of a patient with vestibular migraine (▸ https://doi.org/10.1007/000-kxx)

simultaneous occurrence of these symptoms can be coincidental, both symptoms can also be caused by a single condition: vestibular migraine. Remarkably, the causal relationship between recurrent vertigo or dizziness and migraine has only been recognized more widely in recent decades and has since been the subject of intensive research. The development of migraine and why dizziness episodes may additionally occur over the years are only insufficiently understood. Temporary hyperexcitability of certain brain areas are possible mechanisms that can lead to the development of migraine episodes. Neurotransmitters released in the brain and inner ear play an important role in this process.

Migraine, or more precisely, migraine headaches, affect 15–20% of all women and 8% of all men. The first symptoms of migraine usually appear in adolescence or young adulthood, but can also occur in childhood. Improvement in migraine symptoms often occurs at an older age, in women not infrequently with the onset of menopause. The likelihood that you know someone in your family or circle of friends with migraine headaches, or even suffer from them yourself, is therefore very high. The predisposition to migraine is sometimes genetically determined. Therefore, affected individuals usually have other family members with a diagnosis of migraine. Lack of sleep, stressful situations at work or in private life, e.g., exam stress or sensory overload, can promote the occurrence of a migraine episode. Those

affected can often sense the onset of a migraine episode very early. Many notice typical signs such as irritability, restlessness, or unusual fatigue. Recurrent, sometimes intensely throbbing or pounding, unilateral headaches and an increase with physical activity are characteristic. Headaches are often accompanied by pronounced sensitivity to light, noise, and/or smells, as well as nausea/vomiting. What does it feel like to live with migraine? Ask those affected! This will give you a very insightful impression of how much migraine can dominate everyday life.

How Is Migraine Diagnosed?

Diagnostic criteria help in deciding whether it is actually migraine or not. About one in three people with migraine report aura phenomena, which usually occur before the headache, e.g., a visual disturbance—also called visual aura—with flickering or gray zigzag patterns, a (migrating) sensory disturbance (e.g., extending from the shoulder to the fingers and then jumping to the face), or a speech disturbance (the right words can no longer be found or speech becomes slurred). These aura phenomena last 5–60 minutes and then subside. This is usually followed within the next hour by migraine-typical headaches. These typically last for hours to three days. Those affected withdraw, lie down, and try to sleep. Most sufferers have episodic migraine, i.e., there are symptom-free intervals between migraine episodes. The frequency can range from a few episodes per year to several per week. Chronic migraine is defined as headaches on more than 15 days per month (with migraine-typical headaches on at least eight days) and migraine symptoms persisting for at least three months. Chronic migraine is much less common than episodic migraine.

Vestibular symptoms and Migraine: Vestibular Migraine

Vestibular symptoms of various types (swaying dizziness, spinning vertigo, gait unsteadiness, or a diffuse feeling of lightheadedness) can be part of a migraine episode. The diagnosis of vestibular migraine is found in up to 3% of the population, with women being up to five times more frequently affected. In about 10% of all patients with a first diagnosis of migraine, vestibular migraine is also diagnosed at the same time. However, vestibular migraine usually occurs years or even decades after the onset of migraine headaches. The onset of vestibular migraine is more frequently observed in women during menopause. There are also diagnostic criteria for vestibular migraine, which require at least five episodes of symptoms, a criterion that may not be met at the first medical evaluation.

Other Causes of Similar Symptoms

To diagnose possible vestibular migraine, it is essential to distinguish it from other recurrent (i.e., episodic) causes of vertigo and dizziness and, in the case of first-time occurrence, also from other acute causes of dizziness. Depending on which elements of vestibular migraine are reported, the main differential diagnoses are Menière's disease, peripheral paroxysmal positional vertigo (namely because episodes of vestibular migraine can be associated with positional vertigo, and, in particular during a first episode of the combination of vertigo or dizziness with headache, a stroke (see Sects. 4.1.3, 4.1.1, and 4.2.1).

Important

Patients can simultaneously suffer from Menière's disease, vestibular migraine, and peripheral paroxysmal positional vertigo. When vertigo or dizziness with headache in the neck occurs for the first time, stroke should be considered first, as it can also be accompanied by headache.

Patients' Symptoms

Vestibular symptoms are perceived very differently by individuals. Reports include spinning vertigo or swaying dizziness, gait disturbances, but also a diffuse feeling of lightheadedness, dizziness only in certain head/body positions, nausea up to vomiting, or hypersensitivity to light, noise, or smells, as well as movements. If migraine headaches accompany the recurrent vestibular symptoms, this greatly facilitates the diagnosis of vestibular migraine. However, episodes with vertigo or dizziness in vestibular migraine can also occur without or with only mild migraine-typical accompanying phenomena.

If these symptoms are new, this often leads to presentation in the emergency department and comprehensive evaluations (see below). However, elements of migraine can often be identified in these patients through targeted questioning. This includes, for example, noise or light sensitivity accompanying the dizziness, a tendency to withdraw, or intense nausea, but also mild pressing headaches, often located at the forehead or in the neck. The duration of these vestibular episodes varies greatly from patient to patient and can range from a few seconds up to three days. Secondarly, dizziness symptoms can even be permanent; in these cases secondary functional dizziness should be considered (see Sect. 4.3). The intensity of vertigo and dizziness is also described as very variable and can range from mild to very severe. Episodes of vestibular migraine can also occur position-dependent, e.g., only when lying on the right or left side (this makes a differentiation of

peripheral paroxysmal positional vertigo sometimes difficult) or only when viewing complex and moving visual scenes. Some patients also report ear symptoms, which can include a foreign body or fullness sensation (unilateral or bilateral), but also hearing loss or a tinnitus.

Initial Evaluation

Specialist evaluation usually takes place after referral from a general practitioner, less often in the emergency department. Structured questioning about characteristic symptoms precedes the patient examination and is based on the diagnostic criteria for vestibular migraine.

Physical Examination

It is helpful to see the doctor during a episode with vertigo or dizziness. In this case, minor abnormalities of eye movements can often be detected. The examination assesses whether the patient's eyes are stable or the eyes are jumping, i.e., a nystagmus, whether they can follow objects, or whether the gaze can be shifted from one object to another. In most patients, reduced eye stability can be detected during the episode, e.g., when looking straight ahead in the sense of a spontaneous nystagmus or an inability to keep the gaze stable to the right or left (so-called gaze-evoked nystagmus). Patients usually describe reduced gaze stability as blurred vision or image shaking, which can trigger or worsen dizziness, gait unsteadiness, or nausea. When testing so-called smooth pursuit eye movements, you are asked to follow a slowly moving object. A slight jerking of the eyes can often be observed. Similarly, when testing rapid gaze shifts from one point to another (so-called saccades), inaccuracy in eye positioning may be seen. The patient's eyes miss the target and must correct their position. As a result, a saccade takes more time. During an episode many affected individuals show position-dependent instability of the eyes, typically when lying on their back or side, which is usually accompanied by position-dependent vertigo, dizziness and nausea.

During standing and walking tests, increased body sway while standing (especially with eyes closed) and while walking can be observed during the episode. In cases of pronounced gait unsteadiness, patients are no longer able to walk or stand without assistance. Hearing is also tested; sometimes a new, usually very mild impairement of hearing on one or both sides is found. The rest of the neurological examination is typically normal, except the patient has a so-called aura (see above).

If the specialist neurological evaluation takes place outside an episode, the physical examination is usually unremarkable, except that some of the above-mentioned central eye movement disorders, e.g., eye jerking when

looking to the side or in the side-lying position, can also be detected during targeted examination in the interval.

Additional Examinations

The need for additional examinations primarily depends on the course of the illness, the symptoms described, and the findings obtained. If the symptoms are new, have changed significantly in character, or are now occurring more frequently, magnetic resonance imaging of the brain is usually performed promptly. This is mainly to detect other causes of vertigo and dizziness, such as stroke or compression of the vestibular nerve. In vestibular migraine, standard MRI does not reveal any relevant findings. If the patient has aura phenomena, spots in the brain's white matter are more frequently seen on MRI then in healthy individuals. These spots have a distinctly different pattern from changes seen, for example, in multiple sclerosis. If, on the other hand, the symptoms have been known for many years and are stable over time, MRI is usually not needed.

If vestibular symptoms occur acutely and are new in this form, immediate differentiation from other causes of vertigo and dizziness, especially stroke, is necessary. In this case, a computed tomography (CT) scan and CT-angiography of the brain is usually performed immediately because the symptoms may be caused by a stroke. A cerebrospinal fluid examination (lumbar puncture)will only be required if an inflammatory or autoimmune disease is considered.

In cases of recurrent vestibular episodes, instrumental examination of the vestibular organ is often performed. This almost always takes place outside the episode. The sensors in the vestibular organ for rotational accelerations are tested via the vestibulo-ocular reflex, which transmits movement stimuli from the vestibular organ via the vestibular nerve to the brain and triggers appropriate eye movements (see Sect. 2.1). In patients with vestibular migraine, the vestibular organ is intact, so normal examination findings are expected. If testing is possible during a dizziness episode, various abnormalities may be observed. This includes (usually mild) hypofunction of the vestibular organ or hearing.

Therapy

The treatment of vestibular migraine is divided into therapy for the acute episode and preventive treatment if patients suffer from frequent episodes (two or more per month). The focus of acute treatment is on resolving migraine symptoms, especially vertigo, dizziness, nausea, vomiting and headache. Preventive treatment aims to prevent further episodes as much as possible.

Acute Treatment of a Vestibular Migraine Episode

The acute treatment of vestibular migraine largely coincides with the acute therapy of migraine headaches, especially when headache and dizziness occur simultaneously. Initially, classic painkillers such as paracetamol, ibuprofen, or acetylsalicylic acid are used. It is recommended to use substances that have previously been effective. Migraine-specific acute agents (triptans) are used if classic painkillers are not sufficiently effective and in cases of severe headache, but should only be taken at the onset of migraine. In certain conditions, including cardiovascular disease, triptans must not be used. In vestibular migraine without or with mild headache, triptans should be avoided. Classic painkillers and triptans often fail to treat dizziness. Some patients report, for example, a reduction in headache while dizziness persists. Nausea and accompanying vomiting in (vestibular) migraine often lead to significantly reduced absorption of medication in the gastrointestinal tract, i.e., medications taken for acute treatment remain ineffective. The use of preparations administered via the nasal mucosa, as suppositories, or intravenously is then very helpful. In cases of pronounced nausea and/or vomiting, the use of antiemetics is advisable, e.g., domperidone, metoclopramide, or ondansetron. Symptomatic treatment of vestibular symptoms during the episode is possible with antivertiginous agents, which include medications from various classes, e.g., dimenhydrinate or betahistine.

Important

Non-pharmacological therapeutic measures are important in the treatment of migraine: avoiding provocative factors, life-style changes, regular exercise, shielding from stimuli are very helpful, as is adequate fluid intake and sleep.

Preventive Therapy for Vestibular Migraine

If episodes of vestibular migraine occur repeatedly, preventive therapy can counteract the occurrence of future episodes. The realistic treatment goal is to reduce the frequency, duration, and intensity of migraine episodes by at least 50%. Only in a few patients do migraine episodes disappear completely. In addition to various non-pharmacological measures, different medications are available. Preventive treatment with medication is only sensible if episodes occur with a certain regularity. There should be at least two to three episodes per month to justify (depending on the preparation, daily) preventive treatment. The duration and severity of individual episodes have a significant influence on whether preventive pharmacological treatment is recommended. For example, one severe and long-lasting migraine episode

per month can have a much greater negative impact on quality of life than three short and mild episodes in the same period.

Avoidance of Triggering Factors

Certain lifestyle-modifying measures are important for the prevention of vestibular migraine, which every patient with migraine should implement, even with a low number of episodes. The behavioral recommendations are essentially based on the treatment concepts for migraine headache. Possible triggers such as stress or lack or too much of sleep should be avoided. Provoking factors, such as certain food (like chocolate, red wine, cheese or even coffee), should also be avoided. Regular physical endurance training is recommended (about 30–45 minutes of endurance sport 2–3 times per week) and, for stress reduction, daily relaxation techniques such as progressive muscle relaxation according to Jacobson, autogenic training, yoga, or tai chi. Regular, healthy meals and adequate fluid intake (about 2.5 liters per day) should be ensured. Other treatment options include acupuncture or biofeedback therapy.

Medications for Preventive Treatment

The efficacy and side effect profiles of the medication classes used for preventive treatment differ. Therefore, the choice of substance should be individualized. Medications include, for example, beta blockers and so-called sartanes, certain antidepressants, and nowadays only rarely anti-seizure medications (formerly called antiepileptics). The significance of targeted antibody therapy, which has significantly expanded treatment options for migraine headache, is getting to established more and more for vestibular migraine. Currently the so-called CGRP antagonists used are usually reserved for patients who either do not benefit from established and less expensive preventive medications or cannot tolerate them. A combination of high-dose magnesium and vitamin B2 has also become an option in the preventive treatment of vestibular migraine.

The tolerability of the individual medication classes varies, which is why individual counseling is crucial. Beta blockers (e.g., propranolol or metoprolol) are not recommended for low heart rate, and should also be avoided in known allergic bronchial asthma and a tendency to depression. Anti-seizure medications, e.g., topiramate, can be used to treat migraine headaches. However, topiramate should not be used in women who could potentially become pregnant due to proven harm to the unborn child. A relevant side effect of topiramate is increased forgetfulness. Antidepressants can also be used to treat migraine. Several substance classes have proven effective, e.g., classic tricyclic antidepressants (such as amitriptyline), selective serotonin

reuptake inhibitors (SSRIs, such as escitalopram), and serotonin-norepinephrine reuptake inhibitors (SNRIs). Dizziness, balance disorders, fatigue, and cardiac arrhythmias are among the relevant side effects of antidepressants. Calcium antagonists, e.g., flunarizine, can cause increased appetite and weight gain, but also worsen symptoms of Parkinson's disease and cause mood swings. For this reason, we do not use this class of substances anymore. All these medications require the patient's willingness to continue taking them, as slow and gradual dose escalation is necessary. A combination of high-dose magnesium (reference value about 30 mmol or 600–900 mg per day, limiting factor: diarrhea) and vitamin B2 (riboflavin, 400 mg per day) is particularly suitable as initial therapy or when only a suspected diagnosis exists due to its favorable efficacy and side effect profile.

New therapeutic approaches include the treatment of vestibular migraine with CGRP antibodies and botulinum toxin A, analogous to the treatment of chronic migraine. While treatment with botulinum toxin A is administered into the scalp every twelve weeks, the treatment interval for CGRP antagonists depends on the product, as an injection into the subcutaneous tissue either every four or twelve weeks or daily as a tablet. Antibody-based therapies have really revolutionized the treatment of migraine headaches and are characterized by very good efficacy and high tolerability. Initial placebo-controlled studies with CGRP antibodies are encouraging. Currently, due to the high costs of antibody-based therapies, it must be proven that established, more cost-effective therapies are ineffective or only insufficiently effective. This process usually takes six months or more, which can be burdensome for those affected.

Course Without and With Treatment and Response to Therapy

The natural course of vestibular migraine is usually variable. Quieter phases alternate with more active phases, with external influences (triggers) playing an important role. If the symptoms have been known for many years, medical consultation usually only occurs when exceptionally intense and long-lasting migraine episodes appear. In two-thirds of patients, a stable or improved course is observed over a period of five to ten years. The remaining third, however, experience a worsening of symptoms.

Preventive treatment is monitored by a physician at regular intervals. Tolerability and efficacy of the preventive treatment are assessed and adjustments made if necessary. A headache and dizziness diary is very helpful. If it is kept without gaps and the individual headache and/or episodes with vestibular symptoms are documented in detail, including duration, intensity, accompanying symptoms, and response to acute medications, it can be crucial in determining the effectiveness of a medication. Various smartphone

apps are now available for this purpose. However, the response to preventive treatment can only be conclusively assessed after ten to twelve weeks. Therefore, it is very important to adhere to this minimum treatment duration. If the active substance is well tolerated, premature discontinuation of therapy prevents assessment of its effectiveness. Nevertheless, early discontinuation of treatment may be necessary due to relevant side effects.

If there is no sufficient response to preventive treatment after twelve weeks, it is advisable to taper off the active substance and start with a medication from another class.

If preventive treatment is lacking despite frequent migraine episodes, this usually leads to excessive use of classic painkillers and/or triptans. If this occurs over a period of more than three months and on at least every third day, there is a risk of developing physical dependence. If the painkillers/triptans used are not taken, the headaches then become more severe and change in character. This type of headache is called medication overuse headache. Continued use of painkillers can cause damage to the liver or kidneys. When patients discontinue acute medication on their own, they usually notice a further increase in migraine symptoms, which is why they tend to continue taking it. It is important for patients and their relatives to be informed early about the possibility of dependence so that it can be avoided. If medication overuse headache nevertheless develops, (inpatient) withdrawal may be necessary. A new preventive treatment is then initiated; withdrawal symptoms are alleviated with corticosteroids.

4.2.3 Dizziness, Imbalance, Gait Unsteadiness, Impaired Coordination, and Eye Movement Disorders: Cerebellar Diseases

In Brief

The cerebellum is of great importance for the coordination of movement sequences. Balance and gait disorders, dizziness, coordination disorders of the arms and legs, eye movement disorders, and speech and swallowing disorders are possible symptoms of cerebellar dysfunction. Various causes can lead to an impairment of cerebellar function. Some of these, such as stroke, occur suddenly, while others, such as genetic diseases or autoimmune inflammations, develop gradually. The treatment of cerebellar diseases depends crucially on the cause. Fortunately, there is an increase of treatment options for cerebellar diseases, ranging from the immunosuppressive treatment in autoimmune diseases to symptomatic therapy with so-called 4-aminopyridine or acetyl-L-leucine. Treatments with life-long balance exercises also lead to a significant improvement in the overall situation.

A Typical Case

You have noticed a slowly increasing unsteadiness while walking for about 15 years. You have also observed that this becomes significantly worse in the dark. When you turn your head quickly, your vision briefly becomes blurry and you then have difficulty seeing your surroundings clearly. Your manual dexterity has also steadily deteriorated, so that you now need much more time, for example, to button up a shirt. Over the years, your speech has become somewhat quieter and less clear, making it increasingly difficult for you to express yourself and communicate. Because of your symptoms, you have withdrawn socially, which you describe as very distressing during your consultation with the doctor. Finally, when asked, you mention that you have suffered from chronic coughing and throat clearing for about 20 years. You have already seen your general practitioner, who could not find a cause for your symptoms. He is now referring you to a neurologist.

You describe the above symptoms to the neurologist. He asks whether you also have symptoms at rest, particularly whether you have noticed any swaying or oscillation of your environment. You emphasize during the conversation that there is no swaying or spinning while sitting or lying down, but that the image wobbles especially when you are looking to the right and left. Your speech and your swallowing are also impaired. In addition, there is a tingling in your feed with impaired sensation. The neurologist wants to know whether your unsteadiness while walking and your manual clumsiness increase after consuming small amounts of alcohol or when you are very tired, which you confirm. Finally, the doctor asks whether you can name any trigger for the symptoms described, which you deny.

The neurologist now examines you. First, he looks at your eye movements. The neurologist asks you to look straight ahead and observes that your eyes slowly drift upward and then jerk downward, which is called downbeat nystagmus. He then asks you to look to the side and hold that position, which you find visibly difficult and which causes you to report a wobbling of the image. The examiner notices that your eyes drift back to the center and then jerk outward. He also observes that rapid eye movements to the right or left as well as up or down are slowed and shortened, so your eyes need several steps to reach their target. You are now asked to follow the examiner's finger with your eyes, and the doctor notices that your eye movements are not smooth but jerky. Next, the neurologist holds a drum with a striped pattern in front of your eyes, rotates it, and observes that you are unable to follow this pattern with your eyes. These findings indicate a disorder of the cerebellum (or brainstem).

He then asks you to fixate on his nose and quickly turns your head to the right and left while observing your eye movements. This test examines the function of the vestibular organs. He sees that during these rapid head turns to the right and left, your eyes move with your head, and you then make a quick eye movement to see the examiner's nose again. This indicates that both vestibular organs are not functioning properly.

The neurologist now asks you to touch your nose with your outstretched index finger while your eyes are closed. You have great difficulty hitting your nose with your finger and miss it repeatedly, which points to a cerebellar disorder. He also tests your visual acuity and hearing, both of which are normal.

Next, the doctor asks you to stand upright. This allows him to assess your ability to stand. It becomes apparent that you sway significantly after closing your eyes. The doctor then asks you to walk and observes marked unsteadiness with repeated corrective steps to the side. When walking along an imaginary line, this swaying increases noticeably.

He then uses a tuning fork to test your sense of touch, which is normal in your hands but significantly reduced in your feet. Reflex testing with a reflex hammer is unremarkable in your arms, but the neurologist cannot elicit your reflexes in your legs. This shows that the function of your peripheral nerves in your legs is impaired.

- **What further examinations are now performed?**
  With the so-called video head-impulse test the speed of head and eye movements during quick head turns is measured simultaneously, thereby the function of the vestibular organs can be analyzed precisely. This reveals that both vestibular organs are almost completely nonfunctional. Further examinations are scheduled, including a magnetic resonance imaging (MRI) scan of the brainwith and without contrast agent, as well as nerve conduction studies of the arms and legs. The brain imaging showsmoderate cerebellar atrophy without evidence of a brain tumor or encephalitis. During nerve conduction studies, the neurologist notes that the function of the arm nerves is slighty and of the leg nerves is significantly reduced, and he diagnoses polyneuropathy. Various blood tests are also performed, all of which are unremarkable. After these examinations, the doctor asks you to return to his office to discuss the findings together.
- **What is the cause of the symptoms?**
  The neurologist explains that several systems important for balance, eye movements, and coordination of movements are no longer functioning properly in your case. On the one hand, hypofunction of both vestibular organs can be demonstrated. The technical term for this is bilateral vestibulopathy. At the same time, the neurologist emphasizes that cerebellar function is alsoimpaired and that the arm and leg nerves only respond weakly to stimuli. This leads to the movement-dependent swayingdizziness and gait disturbance you experience, which typically worsen in the dark and on uneven ground. The loss of cerebellar function causes you to see your environment wobble even at rest due to a so-called downbeat nystagmus, to be clumsy with your hands, and to speak somewhat indistinctly.
  The doctor explains that, based on the slow progression of symptoms and the examination results, he suspects a genetic disorder. However, the search for various hereditary diseases is initially inconclusive.
- **What does the treatment look like?**
  The most important treatment principle is lifelong training of balance and fine motor skills. This means you need to do specific exercises for about 30 minutes every day to gradually improve your balance and fine motor skills over weeks and months. It is important to note that a treatment effect often only becomes noticeable after two to three months. The same applies to deterioration if you stop training. The doctor also recommends speech therapy and mentions the possibility of using medication to alleviate the visual wobbling and unsteadiness while walking.

- **What is the course of the disease?**
  At your first follow-up with the neurologist after three months, you report a slight improvement in balance and fine motor skills with regular physical therapy, occupational therapy, and speech therapy. Your speech has also become somewhat clearer. However, the visual wobbling remains unchanged, and you decide to try a medication intended to support cerebellar function. Taking this medication (4-aminopyridine) does not improve your symptoms, so the treatment trial is stopped after six weeks. Over the years, you notice a gradual further worsening of your symptoms, especially your walking becomes more unsteady and you first need one, then soon two walking aids. You also now notice that you are increasingly choking on liquids. The doctor then suggests specifically searching for a certain hereditary disease that could match your symptoms. You agree, and aboutone monts later, you receive confirmation that a hereditary disease has been found. This is called Cerebellar ataxia, Neuronopathy and Vestibular Ataxia Syndrome (CANVAS) due to a mutation in the so-called Replication Factor C subunit 1 (RFC1-associated ataxia). The doctor encourages you to continue your therapy exercises consistently and life-long.

Case Example 2

In the video in Fig. 4.19, a patient with a combination of cerebellar disease and loss of both vestibular organs describes his medical history.
In the video in Fig. 4.20, the patient's walking ability is assessed.

Functions of the Cerebellum

The cerebellum, a part of our brain, acts as a coordination center responsible for ensuring that the interaction of individual body parts works properly. Specifically, the cerebellum ensures that movements are coordinated, smooth, and precise. This applies, for example, to movements of the arms and legs, so that an object can be grasped accurately or one step follows the next while walking and balance can be maintained. Coordination of movement sequences is also important for speaking, swallowing, and eye movements. The functions of the cerebellum often occur unconsciously. Only when the cerebellum is impaired do those affected begin to realize how extensive its functions are. In addition, the cerebellum is partly responsible for higher cognitive functions. Loss of function can also affect psychological well-being.

Cerebellar Dysfunction

The cerebellum is in constant communication with other brain areas, the brainstem, spinal cord, and cerebrum. Sensory information is processed, combined, and—if necessary—correction signals are sent to the muscles. If parts of the cerebellum or the entire cerebellum are impaired, this coordination function is no longer fully ensured. Movements of arms and legs

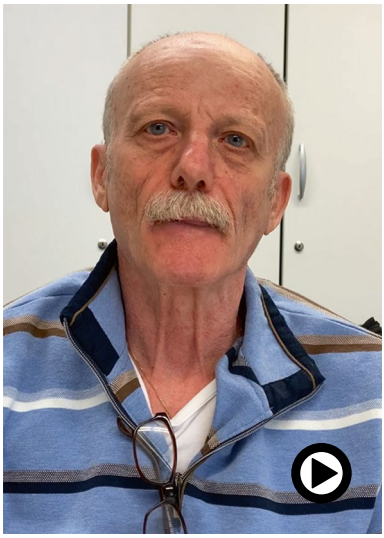

Fig. 4.19 Medical history of a patient with a combination of cerebellar disease and loss of both vestibular organs (▸ https://doi.org/10.1007/000-kxy)

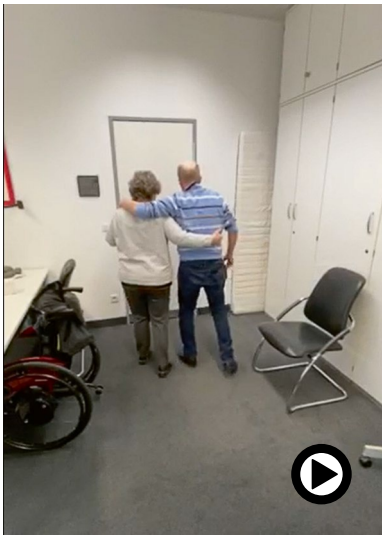

Fig. 4.20 Assessment of walking ability in a patient with a combination of cerebellar disease and loss of both vestibular organs (▸ https://doi.org/10.1007/000-kxz)

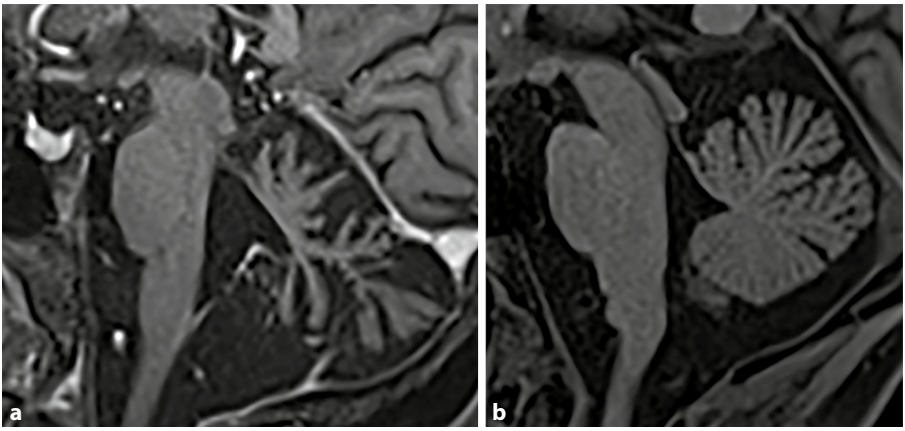

Fig. 4.21 Marked medication-induced shrinkage of the cerebellum. In this 49-year-old patient with long-standing epilepsy, MRI (**a**, lateral view) under therapy with several drugs to treat his seizures showed pronounced cerebellar atrophy. **b** For comparison, the image of the cerebellum of a healthy person (Image source: Radiology, Kantonsspital Baden)

become uncoordinated, speech becomes unclear, swallowing is impaired, walking becomes unsteady, and there is often a downbeat nystagmus and the precision of eye movements is reduced. Cerebellar dysfunction can arise in various ways. It can occur abruptly, for example, as a result of a stroke, brain hemorrhage, or traumatic brain injury. Cerebellar disorders can also progress slowly, for example, in the case of excessive alcohol consumption,in autoimmune or hereditary diseases. Various medications can also lead to cerebellar dysfunction, such as certain drugs used to treat epileptic seizures (carbamazepine, phenytoin; Fig. 4.21). A very broad range of hereditary diseases also lead to premature loss of cerebellar function—often in combination with disorders of other organ systems. Hereditary cerebellar diseases can occur in childhood or adolescence, but may also first cause symptoms in (older) adulthood. For counseling affected individuals, identifying the underlying disease by genetic testing which is nowadays possible for most of these diseases is very important. This allows much better statements regarding prognosis, comorbidities, and treatment options.

Patient Symptoms

Cerebellar dysfunction can manifest in very different ways. This depends crucially on which parts of the cerebellum are affected, what triggered the disorder, and at what stage of life the patient is. Most patients report a gradual worsening and slow onset of symptoms, but sometimes symptoms develop

within days to a few weeks as in autoimmune diseases of the cerebellum. Typical initial symptoms are gait disturbances. At first, these are often barely noticeable from the outside and not yet relevant to daily life. Over time, however, they can increase significantly and may be associated with (near-)falls. Such balance disturbances are particularly noticeable in challenging terrain, such as walking on sand or forest floor or in poor visibility. Under these conditions, the cerebellum's coordination function is especially challenged. Those affected often describe this limitation as diffuse dizziness or a feeling of lightheadedness. Eye or head movements can trigger or worsen such symptoms, especially if they are rapid. Patients with cerebellar diseases often no longer have stable vision. The image appears blurred or wobbles up and down due to a downbeat nystagmus or side to side. This is sometimes more pronounced when looking to the side and can depend on body position. Some patients report more stable vision when lying on their back and worsening in sitting or prone positions. This observation can be used specifically, for example, to read better while lying on one's back. Rapid gaze shifts are also described by those affected as impaired. For example, it takes longer to shift gaze from a nearby object to a distant one and to see it clearly. This is called delayed accommodation and is also regulated by the cerebellum. Changes in speech occur in many affected individuals: speech becomes quieter, less clear, and more monotonous or staccato. While this is usually subtle at first, over time speech can become so altered that communication is significantly impaired or even impossible. Some patients also report swallowing difficulties. These are mainly noticed when drinking liquids quickly and can lead to repeated swallowing, throat clearing, or coughing. Pneumonia can be a consequence.

Initial Assessment

The scope of assessment for patients with cerebellar dysfunction depends crucially on the time course of symptoms. Patients with slowly progressive symptoms are evaluated in a specialist neurological outpatient clinic. Less commonly, if symptoms begin suddenly, the initial assessment takes place in an emergency department or on an inpatient neurology ward. Examinations are often extensive and require repeated doctor visits and additional tests, in these days with a special focus on autoimmune and genetic diseases to make a specific diagnosis.

Physical Examination

If cerebellar dysfunction is present, typical findings can be demonstrated on clinical examination. Often, not all of the above-mentioned cerebellar functions are impaired; rather, individual subdomains predominate (e.g.,

swallowing disorder or impaired fine motor skills of the hands). The doctor systematically tests various functions, including coordination, stance and gait, as well as eye movements and speech as well as swallowing. Coordination testing includes the assessment of directed movements, such as touching the tip of the nose with the index finger, and alternating movements, such as rapidly tapping the thighs with the palms and backs of the hands. In cerebellar dysfunction, such movements are jerky and inaccurate, i.e., the target is missed or overshot. There may also be tremor during movement (so-called intention tremor when moving to a target). Movements performed more slowly are generally more accurate. These changes are usually bilateral but can also be unilateral, especially in cases of cerebellar stroke or tumor. When stance is tested, patients with cerebellar dysfunction show increased swaying. This is further exacerbated with eyes closed and on a soft surface; sometimes the patient must be caught to prevent falling. Cerebellar diseases are associated with a typical gait pattern called broad-based ataxic gait. Patients place their feet further apart when walking to improve stability. At the same time, the walking movements, i.e., lifting and rolling the feet, are no longer smooth but uncoordinated (ataxic). If there is also increased muscle tone in the legs (spasticity), it resembles walking on stilts. This indicates additional involvement of other brain areas (outside the cerebellum).

Assessment of eye movements is central in cerebellar diseases, but also in patients who present with dizziness or postural imbalance (so-called cerebellar dizziness) and very helpful for diagnosis. Various eye movement disorders can occur in cerebellar diseases: eye stability may be reduced, rapid gaze shifts or slow pursuit of an object may be impaired. Characteristic is a downward-directed, jerky eye movement when looking straight ahead. This so-called downbeat nystagmus increases with lateral gaze and may be accompanied by outward-directed nystagmus. Slow pursuit movements are typically jerky, slowed, and no longer smooth. Rapid gaze shifts (saccades) are no longer accurate, being often to long, rarely too short. Both require corrective eye movements. This significantly increases the time needed to actually see the object of interest.

Depending on the cause of cerebellar dysfunction, further investigations may be useful that are not directly related to the cerebellum. For example, there may also be damage to the brainstem or a disorder of the peripheral nervous system. In the latter case, sensory input from the soles of the feet is reduced. This can further increase gait unsteadiness. Retinal dysfunction may also be present. This negatively affects balance, as patients with cerebellar diseases rely more on visual sensory input. Therefore, a thorough clinical neurological examination is important in patients with cerebellar dysfunction.

Instrumental Examinations

For reduced cerebellar function, the necessary diagnostic workup depends on the time course of symptom development. In cases of acute cerebellar dysfunction, emergency instrumental diagnostics are required (see Sect. 4.2.1). Usually, a CT scan and CT-angiography are performed, less commonly an initial MRI of the brain. The main focus is on searching for acute causes, such as stroke or cerebellar inflammation. If medications are being taken that affect brain function, possible overdose should be checked in the blood. This applies, for example, to drugs for epilepsy, but also for various psychiatric conditions. Malnutrition or nutritional deficiencies can also lead to acute or subacute loss of cerebellar and brainstem function, especially in the case of vitamin B1 deficiency. If suspected, the vitamin B1 level in the blood should be determined and the patient thereafter treated with vitamin B1.

In patients reporting a less abrupt onset of cerebellar dysfunction, MRI of the brain plays a crucial role. This is used to specifically look for tumors as well as inflammatory changes in the cerebellum. In slowly progressive (chronic) cerebellar dysfunction, such as hereditary cerebellar diseases, MRI may reveal cerebellar atrophy. If autoimmune disorders are considered (more frequent that previously thought) lumbar puncture and examination for a broad range of autoantibodies is crucial. Assessment of vestibular function and detailed evaluation of eye movements is also recommended in cerebellar dysfunction. The goal is to detect additional vestibular organ dysfunction and to document the extent and pattern of eye movement disorders. This includes testing the sensors for rotational movements (semicircular canals) with the video head impulse test. Eye movement assessment includes evaluating eye stability in certain positions (straight ahead, lateral gaze) and the ability to follow moving objects slowly or rapidly with the eyes. This allows detection and quantification of, for example, a downbeat or gaze-evoked nystagmus. At the same time, the pattern of eye movement disorder can be described based on rapid gaze shifts, e.g., whether there is overshooting or undershooting. In cases of subacute or chronic dysfunction, a possible association with cancer is also investigated, as certain tumors, though generally rare, can preferentially cause this. These include certain lung tumors (small cell bronchial carcinoma), lymphoma, breast cancer, and thyroid cancer. As mentioned above, examination of the cerebrospinal fluid (lumbar puncture) is also part of the workup for cerebellar diseases. This is mainly used to look for inflammation, infections of the nervous system, and autoimmune disease with or without tumors.

Genetic testing is important in chronic diseases. Detecting a genetically determined cerebellar disease is crucial for counseling affected individuals

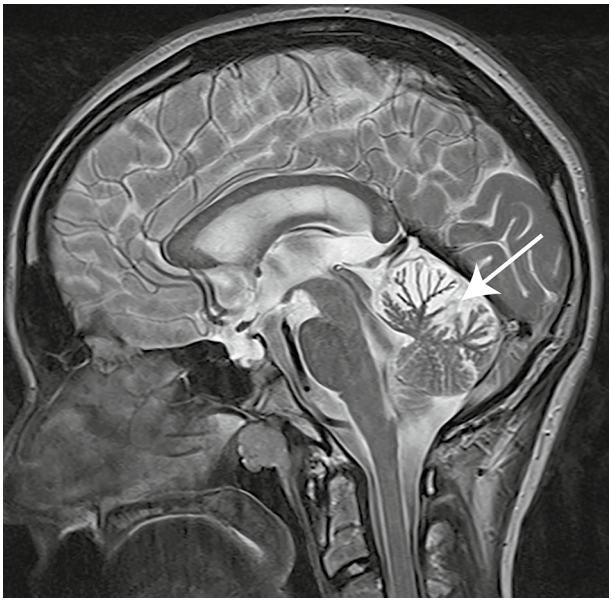

Fig. 4.22 Hereditary cerebellar disease with marked cerebellar atrophy. This 24-year-old patient has slowly increasing unsteadiness while standing and walking, slurred speech, pronounced clumsiness of the arms, and blurred vision. Clinical examination shows advanced cerebellar dysfunction. MRI of the brain (lateral view) reveals pronounced cerebellar atrophy (*white arrow*). A mutation was subsequently detected in this patient, confirming a genetically determined cerebellar disease (Image source: Radiology, Kantonsspital Baden)

and relatives, as well as for early detection of comorbidities. Important clues often arise from asking about similar diseases in the family. Genetic testing usually screens for a large number of different hereditary diseases simultaneously. This testing can take weeks. In recent years, newly discovered hereditary diseases have linked significant proportions of previously unclear cerebellar diseases to corresponding genetic changes (Fig. 4.22). These include, for example, so-called spinocerebellar ataxia 27B, in which gait disturbance and downbeat nystagmus predominate or, as mentioned, above CANVAS. The results of genetic testing can have major implications not only for the person tested but also for offspring and relatives. Therefore, adequate time for consideration and comprehensive information about the consequences of genetic testing are required.

Cerebellar diseases can also be associated with retinal dysfunction. In individual cases, an ophthalmological examination is required. If there are clinical signs of sensory disturbance in the feet, nerve conduction studies using electroneurography are recommended for further evaluation.

Therapy

- **Treatment of underlying causes**

The treatment of cerebellar disorders depends crucially on the underlying cause. In the case of a stroke affecting the cerebellum, acute management and prevention of further strokes (secondary prophylaxis) take priority (see Sect. 4.2.1). If there is a vitamin B1 deficiency, immediate intravenous administration of vitamin B1 is essential. If medication overdose is suspected, the first step is usually to pause the medication, followed by resuming at a lower dose. If the cerebellar disorder is associated with a cancer outside the brain (paraneoplastic syndromes) or a tumor is detected in the cerebellum, treatment of the cancer is crucial. Close coordination with the treating oncologists is necessary in these cases. If there is evidence of an autoimmune disease, the focus is on carefully downregulating the immune system. In addition to plasmapheresis, various classes of drugs can be used for this purpose, ranging from anti-inflammatory agents (corticosteroids and immunoglobulins) to monoclonal antibodies, such as rituximab.

- **Symptomatic treatment**

Often, it is not possible to treat the underlying cause of the disease, or persistent symptoms remain despite targeted therapy. In these cases, symptom-oriented therapy can also provide relief. Such therapies can lead to noticeable improvement, especially in the treatment of gait disturbances or visual disturbances (blurred vision) due to nystagmus. Several agents have proven effective: 4-aminopyridine (effective in downbeat nystagmus, episodic ataxia type 2 and others, acetyl-L-leucine (effective in lysosomal diseases such as Niemann-Pick Type C, GM-gangliosidoses as well as in ataxia telangiectasia and several others), chlorzoxazone (moderately effective in downbeat nystagmus) and acetazolamide (in episodic ataxia type 2). Reducing gait instability improves mobility and lowers the risk of falls. Stabilizing the eyes, for example, improves reading and makes the environment clearer while walking. The degree to which symptoms improve with these agents varies. Most of these agents are usually not approved for the treatment of these symptoms, are prescribed off-label, and sometimes must be sourced from abroad. This often complicates access and reimbursement for these therapies by health insurance providers. Whether such a pharmacotherapy should be undertaken is decided on a case-by-case basis by the physician in consultation with the patient.

- **Balance training and gait rehabilitation**

Regardless of the cause of cerebellar dysfunction, targeted physiotherapy is of great importance. The focus is on balance and coordination training. Home exercises are also taught. If more intensive therapy is required, inpatient neurorehabilitation is an option. The goals are to improve mobility, make sensible use of assistive devices, and prevent falls and fall-related injuries. Aquatic therapy or even equine therapy can be important adjunctive therapies for severely affected patients with pronounced trunk instability. If there is a swallowing or speech disorder, this is specifically treated with speech therapy. The aim is to reduce the risk of liquids and solid food entering the trachea and lungs and to improve communication.

Course without and with treatment and response to therapy

The course of a cerebellar disorder depends crucially on the underlying cause. Comprehensive evaluation is of great importance for counseling the patient and their relatives. If a causal and effective therapy is known, significant recovery of cerebellar function can be expected. Examples include cerebellar dysfunction due to deficiencies, infections, or medication overdoses. If acute vitamin B1 deficiency is promptly treated with replacement therapy, or if an overdose of an antiepileptic drug is corrected, substantial recovery is often observed. Autoimmune diseases affecting the cerebellum can be stabilized with appropriate combined therapy as mentioned above.

When there is structural brain damage, such as after a stroke, or in cases of accelerated degeneration of cerebellar neurons, the potential for recovery is significantly lower. Alcohol-induced cerebellar damage can at best be stabilized by strict abstinence from alcohol, and the same applies to cerebellar disorders triggered by tumor cells (paraneoplastic cause). The symptoms of genetically determined cerebellar disorders often progress over time, with marked differences in the rate of deterioration. Most symptomatic therapies can contribute to (temporary) stabilization.

Episodic Ataxia Type 2 and other so-called CACNA1A-related diseases

Typical symptoms of the rare genetic disorder episodic ataxia type 2 are recurrent episodes, usually lasting hours to a day, of unsteady dizziness which can be accompanied by speech, coordination, and gait disturbances

(ataxia, i.e., imprecise movements). Between episodes, most patients also experience these symptoms in a milder form. Typical triggers for episodes include exercise, stress, or alcohol. The disease usually begins in late childhood or early adulthood. The cause is a mutation in a gene that is particularly important for cerebellar function. Nowadays, the condition can usually be well managed with medication.

A typical case

A 21-year-old female patient reports that since the age of five, she has suffered from episodes of unsteadiness and dizziness several times a month. On further questioning, she states that during these phases she is harder to understand and sways more when walking. She also has more difficulty using her hands. The episodes last from an hour to a day. Between episodes, she feels somewhat unsteady. When asked about triggers, she reports that, for example, playing tennis and drinking small amounts of alcohol can trigger episodes. She also experiences more episodes when she feels stressed. She has even been examined in America because of these symptoms. There, she was told that her eyes "jump up and down."

On physical examination, the neurologist finds these eye movement abnormalities: the eyes jerk quickly downward and then drift back upward (downbeat nystagmus). The doctor also notes mild coordination disturbances in the arms and some unsteadiness when standing. To confirm the diagnosis, genetic testing is performed, which takes several weeks. Indeed, a mutation in the CACNA1A gene is found.

Treatment is initiated with a medication that improves cerebellar cell function (4-aminopyridine, a potassium channel blocker). As a result, she experiences at most one episode per month, and her symptoms between episodes improve. The doctor explains to the patient that this will be a lifelong preventive treatment.

Cause, course, and treatment

Episodic ataxia type 2 is caused by mutations in a gene important for the function of nerve cells, especially in the cerebellum. How the triggering factors (mainly physical activity, stress, or alcohol) lead to episodes is not fully understood. The course of the disease is usually benign. Fortunately, there are good treatment options with medications (especially potassium channel blockers such as 4-aminopyridine and acetyl-L-leucine) that improve the function of cerebellar nerve cells. These can usually prevent episodes and improve symptoms between episodes.

4.3 Functional Dizziness

Functional dizziness is the umbrella term for dizziness syndromes that, in addition to the feeling of fluctuating persistent unsteadiness, can also lead to gait instability, lightheadedness, fear of falling, a feeling of pressure in the head, or increased sweating, without any detectable physical, i.e., organic, cause in most patients: this is so-called primary functional dizziness.

An organic vestibular disorder, such as acute loss of a balance nerve (unilateral vestibulopathy) or peripheral paroxysmal positional vertigo, may be present at the onset or even ongoing as in vestibular migraine or Menière's disease. In this case, the symptoms are referred to as secondary functional dizziness.

Over the past decades, functional dizziness has been given various names: psychosomatic, somatoform, psychogenic, phobic, and finally functional dizziness. In the current classification, the various subtypes of phobic postural dizziness and a recently coined term in the English-speaking world, "persistent postural-perceptual dizziness" (PPPD), are listed. The two differ only slightly. Functional dizziness as a psychosomatic form of dizziness is very common: the patient perceives dizziness somatically, i.e., physically; however, the cause lies in the psychological domain. As you can see in Table 1.1 in Chap. 1, functional dizziness is the most common cause of dizziness, for instance, at the German Center for Vertigo and Balance Disorders and the Department of Neurology at LMU Munich. Most affected individuals are middle-aged; women predominate. At the onset or during the course, particular psychosocial stressors are often present.

Unsteadiness and imbalance can persist on land after prolonged passive transport in vehicles (e.g., after a sea voyage): this is referred to as Mal de Débarquement syndrome. If these symptoms do not resolve after a few weeks, there is a gradual transition to functional dizziness.

A typical case

You have noticed unsteadiness when standing and walking for two years and you are therefore afraid of falling. However, you have not actually fallen so far. Once you just managed to catch yourself, but you were not injured. Sometimes you also experience brief, completely involuntary swaying of your body and feel as if you need to take a compensatory step to stay on track while walking. In certain situations, such as shopping in a supermarket, when there are many people around or many visual stimuli, for example when walking along shelves, the dizziness becomes much worse. Over the past two years, these symptoms have generally increased. They now occur more frequently, last longer, and

interfere with your daily activities. Sometimes you also avoid evening events, such as going to the theater or meeting friends at a restaurant. Overall, you notice that the symptoms usually begin about half an hour after getting up in the morning and then increase over the course of the day. You have already seen your general practitioner, who could not find a cause for your symptoms. Now you are being referred to a neurologist.

The neurologist asks you about other situations that can trigger this unsteady vertigo. You have also noticed it in high places, on bridges, or at work. The symptoms also occur when you have to speak in front of several people. The neurologist asks whether the symptoms improve after drinking small amounts of alcohol or during physical activity, such as cycling. Both are true for you. The neurologist inquires about accompanying symptoms such as headaches, sensitivity to light or noise, hearing loss, paralysis, or loss of consciousness. None of these symptoms have ever occurred in your case.

The neurologist now examines you, looks at your eye movements, which are unremarkable. He asks you to fixate on his nose while he quickly turns your head to the right and left. The eye movements observed during this are also normal and show the doctor that your balance organs are functioning well. The subsequent neurological examination, testing eye movement and motor function, sensation, stance and gait, and fine motor movements of the hands, is normal.

The neurologist then performs some additional instrumental tests to assess your balance system, such as the video head impulse test. This examination is unremarkable in your case. He also refers you to a radiologist for a magnetic resonance imaging (MRI) scan of the head with and without contrast. This imaging study is also normal and shows no abnormalities in the brain.

With the images and findings, you return to the neurologist. He discusses all the results with you in detail, explains that all findings are normal, that your dizziness and unsteadiness when standing and walking are due to increased anxious control of stance and gait, and that this dizziness is called functional dizziness. The following explanation of how this dizziness arises from increased sensitivity regarding one's own balance is already part of the therapy. The neurologist also advises you not to avoid situations, to consciously face situations that trigger dizziness or anxiety, and to exercise regularly. He schedules a follow-up appointment in three months.

You now try to remind yourself repeatedly that the cause of your dizziness is not organic in nature, but is due to increased self-observation. During the consultation, the doctor was already able to alleviate your fear of having a serious organic disease. This also enabled you to increasingly expose yourself to situations that originally triggered dizziness. After six weeks, you notice a significant improvement: you only feel unsteady in certain situations, such as standing on a balcony at a great height with a wide view. In view of the positive course, the neurologist decides at the three-month follow-up that no further measures, such as treatment with antidepressants or psychotherapy, are necessary. Another follow-up is scheduled in five months.

Additional case examples

In the videos in Figs. 4.23, 4.24, and 4.25, patients with functional dizziness describe their symptoms.

Fig. 4.23 Patient with functional dizziness (▸ https://doi.org/10.1007/000-ky0)

Fig. 4.24 Patient with functional dizziness (▸ https://doi.org/10.1007/000-ky1)

Fig. 4.25 Female patient who developed secondary functional dizziness after unilateral loss of a balance organ (▸ https://doi.org/10.1007/000-ky2)

Patient symptoms

Affected individuals complain of lightheadedness with unsteadiness when standing and walking and experience fear of falling, i.e., they have anticipatory anxiety about falling, but do not actually fall or, if they do, are usually not injured. Brief subjective body swaying may also occur. The dizziness is usually described as unsteady or diffuse, as a feeling of emptiness in the head, and only rarely as spinning vertigo. Typical triggering situations are also found in other phobic syndromes, such as bridges or crowds, e.g., in department stores. However, dizziness episodes can also occur spontaneously. Over time, the symptoms generalize, leading to increasing avoidance of triggering stimuli. During or shortly after episodes, anxiety and autonomic symptoms such as sweating, palpitations, weak knees, trembling, inner restlessness, unpleasant sensations in the skin or abdomen, general muscle weakness are experienced. Dizziness can also occur without anxiety. Improvement is often experienced after light alcohol consumption, during physical activity, or through distraction, e.g., when talking with others. Gait may be unsteady and insecure, and patients feel the need to hold on to something to avoid falling, making grasping arm movements for support. Depending on comorbid psychiatric conditions, patients may also experience symptoms such as lack of motivation and concentration, fatigue,

reduced performance, limitations in work and daily activities, disturbances of mood and emotions, and sleep disorders.

Physical examination and personality traits of patients

The clinical neurological examination of patients with primary functional dizziness, i.e., without prior or ongoing organic disease, shows no focal abnormalities; in particular, eye movements, balance, vestibular nerve, and peripheral nerve function are normal. Although patients complain of unsteadiness when standing and walking, there is no objective evidence of instability.

If functional dizziness develops after an organic balance disorder in the sense of secondary functional dizziness, residual signs of the organic disorder may still be detectable, but these do not sufficiently explain the patient's symptoms.

Personality traits of affected individuals often include perfectionistic, compulsive, or anxious features. They tend to increased self-observation (introspection), have a need to keep everything under control, are ambitious with high self-expectations, but also easily irritable and anxious. Functional dizziness may also be accompanied by reactive depressive syndrome or an anxiety disorder.

Instrumental investigations

Instrumental diagnostics are important in this dizziness syndrome to rule out organic causes of dizziness. Balance diagnostics (video head impulse test, caloric testing, video oculography, orthoptic examination including measurement of subjective visual vertical) as well as magnetic resonance imaging of the head should be normal or may show residual signs of a previous organic dizziness disorder, but these do not explain the current symptoms. The latter constellation of findings allows the symptoms to be classified as secondary functional dizziness.

Measurement of sway patterns by posturography shows that unsteadiness when standing decreases as tasks become more difficult, in contrast to organic disorders. In tandem stance, a balancing task requiring undivided attention, affected individuals stand almost as steadily as healthy controls. Measurements of leg muscle activity show stiffening, which becomes apparent during distraction by dual tasks. Thus, body control is clearly altered in affected individuals: they show less variability and adapt more slowly to new situations. This is due to a controlling strategy that patients use not only when standing but also when walking.

In gait analysis on a gait analysis carpet, phobic dizziness is characterized by a spontaneously slow, cautious gait that normalizes when walking faster.

Step-to-step fluctuation is normal; patients are not distracted by dual tasks while walking. With eyes closed, walking speed decreases further, indicating that patients rely more on visual feedback and information. In functional dizziness, compensatory steps and fluctuations in gait are observed, and gait normalizes with distraction. These gait changes are due to a controlling strategy that healthy individuals use only under particularly challenging conditions, such as walking on ice.

Eye and head movements also change in dizziness-inducing situations, such as in crowded environments. There is a marked reduction in eye movements compared to healthy individuals, and the environment is not explored visually; instead, the gaze is almost fixed straight ahead and on the ground. Particularly anxious patients show more or less pronounced, rather nervous visual scanning of their horizontal environment.

Brain imaging studies, such as MRI or CT, show normal findings. Studies investigating both cerebral blood flow and connections between different brain structures have demonstrated reduced gray matter volume in the cerebellum with simultaneous increased volume in motor brain regions. These are signs of reduced automatic balance control (cerebellum) and increased voluntary motor activity (motor cortex), which fits well with the above-mentioned stance and gait findings. Blood flow and connectivity between different brain regions involved in vestibular, visual, emotional, and motor processing are also altered. These research findings serve to explore the underlying biological mechanisms but have no practical significance for affected individuals.

Disease model

The theory regarding the development of functional dizziness assumes that, given a certain predisposition and after resolution of a possible preceding vestibular disorder, anxious control of balance persists. This anxious self-observation leads, in response to triggers, to anxious tension of the leg and neck muscles, resulting in body stiffening. When affected individuals move, this leads to a disturbance in the perception of spatial surroundings. Active body movements send signals to various sensory systems: the vestibular, the visual, and the system that perceives the position and posture of the body and limbs (somatosensory system). The incoming signals in the brain are compared with an expectation pattern stored in the brain, calibrated by previous movement experiences. If the current sensory input during active movement matches the expectation pattern, the movement is perceived while maintaining awareness of the environment. If the incoming and expected patterns differ, as can occur with such uncalibrated movement

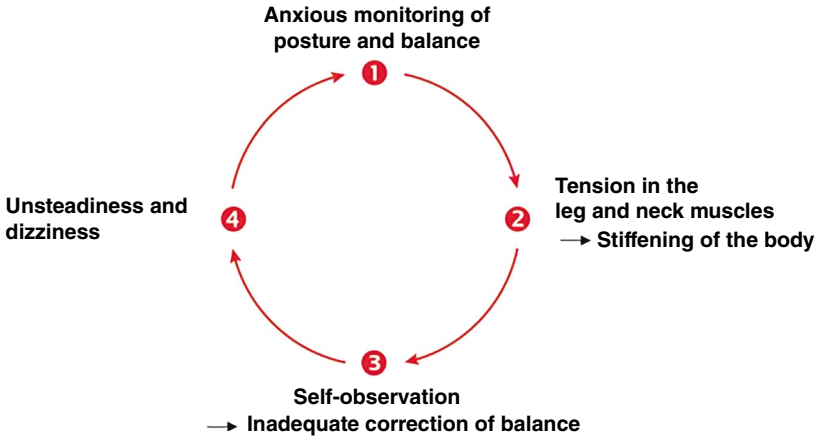

Fig. 4.26 Vicious circle of body instability

stiffening, illusory movements of the environment are perceived, resulting in an acute sensation of dizziness.

Thus, affected individuals show reduced eye and head movements, increased swaying when standing, and a slowed, cautious, or unsteady gait with compensatory steps. This is accompanied by tension in the leg, neck, and also eye muscles. These changes are not specific, but rather a nonspecific response to anxiety in general, as they also occur in height-induced dizziness, a dizziness syndrome that is also associated with anxiety. The physical changes result in a cascade of symptoms, a so-called vicious circle of body instability (Fig. 4.26).

Therapy

Therapy includes a thorough medical history by the physician, a physical and neurological examination, and vestibular clinical and instrumental diagnostics to characterize functional dizziness and exclude other dizziness disorders. Therapy also involves discussing the normal clinical and instrumental findings with the patient, in part to alleviate fears of an undetected serious illness.

Another therapeutic approach is psychoeducation, i.e., explaining to the patient how the dizziness developed. The vicious circle of anxious control of stance and gait is discussed in detail. Patients are instructed to engage in regular endurance exercise, not to avoid triggering situations, but to deliberately seek them out, since the course of the disorder often involves generalization of symptoms with increasing avoidance of triggering stimuli. If these measures alone are not successful, cognitive behavioral therapy may

be considered. There are various therapeutic techniques. Most effective initially is behaviorally guided desensitization to triggering situations. Certain disorder-related beliefs and assumptions should be analyzed by the therapist to develop and reinforce behavioral alternatives. Sometimes psychotherapy must be supported by psychopharmacological treatment, preferably with antidepressants, most often so-called selective serotonin reuptake inhibitors (SSRI). If psychiatric comorbidities, such as an anxiety disorder or depressive syndrome, are present, these must also be specifically treated.

Relaxation techniques such as autogenic training, yoga, or Jacobson's progressive muscle relaxation should be used as supportive measures. We generally recommend **not** to perform specific balance training, as it can increase attention to balance and its control, thereby exacerbating the disorder. The exception is patients with secondary functional dizziness with demonstrable relevant deficits in balance function.

Course

Long-term studies show that after the described therapeutic and educational approach, three-quarters of affected individuals are symptom-free or show significantly improved symptoms, and these positive effects are lasting. Improvement can be achieved even after symptoms have been present for a long time. Therefore, given the generally favorable prognosis, it is desirable for the diagnosis of functional dizziness to be made quickly. Early initiation of therapy can prevent many unnecessary doctor visits, instrumental diagnostics, and further uncertainty for patients due to diagnoses such as "cervical vertigo," etc.

4.4 Dizziness and Balance Disorders in Other Diseases

In addition to diseases originating from the balance organs in the inner ear and the brain, other causes may underlie the symptom of vertigo and dizziness. The most important conditions are described below. Patients may suffer from several of these causes simultaneously.

4.4.1 Dizziness Due to Blood Pressure Regulation Disorders

Typical of orthostatic dizziness, which is triggered by a drop in blood pressure when rising from lying or sitting, are lightheadedness, swaying dizziness

up to a narrowing of the visual field or blacking out. There is also a brief unsteadiness immediately after standing up from sitting or lying down. The cause may be generally low blood pressure, but it can also be a consequence of medications or of autonomic regulatory disorders in diseases such as Parkinson's disease, Parkinsonian syndromes (see also Sect. 4.4.3) or certain cerebellar diseases.

The diagnosis of orthostatic dizziness is made by the physician through blood pressure measurements while sitting and then standing (Schellong test or, if this is inconclusive, tilt table examination).

Regular endurance exercise (for 30 minutes at least three times per week), several small meals a day, adequate fluid intake (at least 2.5 L per day), as well as sleep hygiene, i.e., sufficient, preferably uninterrupted sleep, are therapeutically helpful measures. The physician should pay attention to medications that can cause orthostatic symptoms and adjust them accordingly.

Before getting up from lying or sitting, it is advisable to repeatedly tense and relax certain muscle groups such as the calf muscles, or to cross the arms and tense the muscles, and to stand up carefully and slowly.

Not only a temporary drop in blood pressure after standing up, but also repeatedly occurring high blood pressure can cause lightheadedness, swaying dizziness, and head pressure. Cardiological diagnostics using long-term blood pressure monitoring, as well as long-term electrocardiography and echocardiography, are keys for diagnosis. If medication-based regulation of blood pressure is indicated, regular self-monitoring of blood pressure should be performed to ensure that blood pressure is well controlled throughout the day.

Another important condition causing dizziness upon standing up, especially when getting out of bed, is peripheral paroxysmal positional vertigo, which typically also causes dizziness when lying down (see Sect. 4.1.1).

4.4.2 Dizziness Due to Adverse Effects of Medications

Dizziness is relatively often caused by medications. To confirm such a connection, a careful medical history by the physician is required. Particular attention is paid to the temporal relationship between the start or dose adjustment of a medication and the onset of dizziness. Medication-induced dizziness is not precisely defined in the classification systems of dizziness disorders. There are three types of dizziness-inducing medications:

1. Medications that damage the sensory cells located in the balance organs. These include antibiotics, e.g., aminoglycosides (gentamicin, streptomycin), cancer treatment drugs (cytostatics), diuretics (loop diuretics), and

a drug for cardiac arrhythmias (amiodarone). The damage to the balance organs is bilateral (bilateral vestibulopathy) and typically presents as swaying dizziness during movement with gait unsteadiness and visual blurring while walking (oscillopsia; see Sect. 4.1.4).

2. Medications that lead to swaying dizziness, which increases with higher doses. These medications affect the function of the balance organs, brainstem, or cerebellum and can also cause disturbances of eye movements, e.g., a saccadic pursuit or a gaze-holding defect with gaze-evoked nystagmus in the end positions of eye movements (horizontal or vertical). Patients perceive this as an unstable gaze, which, in addition to swaying dizziness, also leads to gait unsteadiness. There are numerous medications with such adverse effects. These include antiseizure medications (formerly called antiepileptics, e.g., carbamazepine, phenytoin), sedatives (barbiturates, benzodiazepines), antidepressants (e.g., amitriptyline), hormones (corticosteroids, antidiabetics, sex hormones, contraceptives), anti-inflammatory agents (antibiotics, antifungal and antihelminthic agents, tuberculosis medications), as well as drugs for the treatment of Parkinson's disease (L-dopa and dopamine agonists). Alcohol, incidentally, has similar effects. Nevertheless, patients should not be overly alarmed by package inserts, which often list dizziness as a side effect. Mild swaying dizziness usually subsides after a few days of taking the medication. In older people and when taking multiple medications, dizziness as an adverse effect of certain drugs occurs more frequently, sometimes also because of interactions between individual medications. Here, patients should ask their doctor which medications are truly necessary.
3. Medications that cause a blood pressure regulation disorder, which mainly manifests as swaying dizziness with lightheadedness after rapid standing up from lying or sitting (orthostatic dysregulation; see Sect. 4.4.1). Falls are also possible here, especially if there is loss of consciousness. Medications that induce orthostatic symptoms are those prescribed for high blood pressure (antihypertensives, diuretics).

4.4.3 Dizziness in Forms of Parkinson's Disease

Movement disorders originating from dysfunction of the basal ganglia in the brain (Parkinson's disease and Parkinsonian syndromes) are characterized by motor symptoms and very often dizziness and postural imbalance if patients are asked for. These include a shuffling, unsteady gait, reduced facial expression, slowing and disturbance of hand and arm movements, which

Fig. 4.27 Illustration of Parkinson's disease. (From: Handbook of Diseases of the Nervous System, W.R. Gowers, 1886)

may manifest as progressively smaller handwriting, increased muscle tone, and tremor of the arms and legs, preferably at rest (Fig. 4.27). Additional symptoms occur depending on the type of Parkinsonian syndrome, e.g., disturbances of eye movements, sleep and smell disorders, gastrointestinal disturbances, and depressive syndromes. The underlying causes of these disorders are various diseases resulting from the loss of nerve cells in motor and other brain regions. In addition, deposits of protein structures are found in various brain areas, differing in composition and distribution depending on the disease (e.g., alpha-synuclein, Lewy bodies, tau aggregates). The most common group is Parkinson's disease. There are also Parkinsonian syndromes caused, for example, by poisoning or by certain medications. Another group consits of atypical Parkinsonian syndromes, in which the above-mentioned symptoms occur as part of other degenerative (neurodegenerative) diseases of the brain.

A common accompanying symptom of Parkinsonian syndromes is orthostatic regulatory disorder, i.e., after standing up or during prolonged standing and walking, swaying dizziness with lightheadedness occurs. In addition to the above-mentioned therapeutic measures, physical approaches such as

wearing compression stockings, a salt-rich diet, and sleeping with the upper body slightly elevated at night are recommended. In some cases, a change of medications used to treat the Parkinsonian syndrome is necessary.

In cases of very pronounced orthostatic symptoms, medications (including ephedrine, midodrine) can be prescribed by a physician. Of course, therapy includes the treatment of the underlying Parkinsonian syndrome with specific medications after causal classification.

Atypical Parkinsonian Syndromes

Many movement disorders, such as Parkinson's disease or diseases with similarly slowed movements when walking and of the arms, begin with swaying dizziness or gait disturbances as the first symptoms. Therefore, slowly increasing swaying dizziness should also prompt consideration of such movement disorders. Parkinson's disease can be well treated for many years with medications and later also with deep brain stimulation. However, there are other forms that are not yet well treatable.

A Typical Case

A 73-year-old female patient presents to a neurologist because of increasing swaying dizziness over the past year, slowing of gait and movements of both arms and legs. The family fears she may have Parkinson's disease. On further questioning, she and her husband report that she has also fallen at least once a week in the past three months.

The neurologist finds on physical examination of eye movements that the patient can hardly look up or down. The patient's facial expression is reduced. There is marked rigidity in the arms and legs, and movements are slowed. Gait is also slowed with small steps. The doctor now tests postural stability by pulling the patient backward while standing: she almost falls backward.

The neurologist orders a magnetic resonance imaging scan. This shows that the diameter of the upper part of the brainstem (midbrain) is reduced.

The doctor now discusses the cause of the disease with the patient and her family. It is a premature aging of nerve cells due to deposits of certain proteins in these brain areas. This is called progressive supranuclear gaze palsy. The treatment of choice is intensive physiotherapy with gait training, also to prevent further falls. The neurologist explains that there is currently no effective drug treatment for this disease. Unfortunately, in most patients, it progresses significantly within a few years, so that care and alleviation of symptoms become the main focus.

Case Example 2

In the video in Fig. 4.28, a patient with progressive supranuclear gaze palsy and her husband recount the medical history. Examination of the patient's eye movements shows that especially upward and downward eye movements are slowed (video in Fig. 4.29).

Fig. 4.28 Medical history of a patient with progressive supranuclear gaze palsy (▸ https://doi.org/10.1007/000-kxf)

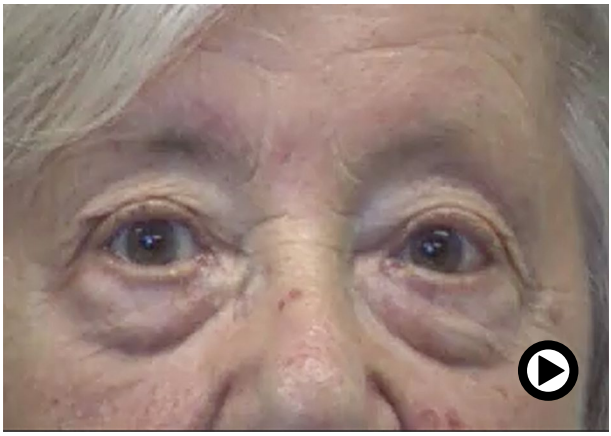

Fig. 4.29 Patient with progressive supranuclear gaze palsy. Examination of eye movements, which are particularly slowed upward and downward (video without sound) (▸ https://doi.org/10.1007/000-ky4)

Cause, Course, and Treatment

The cause of many diseases involving premature aging of brain cells (neurodegenerative diseases) is the deposition of certain proteins in these cells. This is also the case in Alzheimer's and Parkinson's disease as well as many similar conditions. The underlying cause of these deposits is unknown in most cases. In the case described above of progressive supranuclear gaze palsy, this leads to patients no longer being able to look up and down properly, and later also not to the right and left. Otherwise, the outward symptoms of these patients at first glance resemble those with Parkinson's disease: they walk with small steps, arm movements while walking are reduced. However, they typically fall increasingly often as the disease progresses, as the reflexes for postural regulation are impaired. Unfortunately, progressive supranuclear gaze palsy often progresses significantly within a few years, and there is still no drug treatment. Therefore, physiotherapy with balance training and gait training is the mainstay, also to reduce the risk of falls. As the disease progresses, the focus is on alleviating symptoms and ensuring patient care.

4.4.4 Dizziness with Impaired Memory

Dizziness and impairments of concentration, memory, and thinking ability, so-called cognitive deficits, frequently occur together in older people and are among the most common neurological complaints in the elderly. The prevalence in people aged 70 and over increases with age and ranges from 3 to 25%. Although a causal relationship between the two complaints has not been clearly proven, current studies suggest such a connection: Central vestibular and cognitive systems share common networks in the brain, which are jointly affected by the loss of nerve cells or by vascular-related nerve cell changes. Recent research also shows that reduced function of the balance organs in old age is associated with poorer cognitive abilities.

Affected individuals report as their main complaint a persistent or even progressive form of non-directional dizziness, i.e., swaying dizziness and lightheadedness, as well as gait unsteadiness. In those with marked cognitive deficits, there are more central nervous symptoms, e.g., coordination disorders of the limbs, poorer control of trunk motor function (truncal ataxia), and increased muscle tone. Medical history and neurological examination can reveal cognitive impairment (if pronounced and persistent for more than six months, the diagnosis of dementia can be made). In addition to neurological examination, cognitive screening tests are important in the routine assessment of older people with dizziness. Brain imaging in those with

dizziness and cognitive impairment shows patterns of degeneration in specific balance regions. For dizziness complaints, it is important to carry out the diagnostic measures mentioned in this guide to classify the dizziness. If these examinations do not reveal a cause, unexplained, persistent, progressive dizziness may also be a prodromal stage of an impending neurodegenerative disease. Further classification is (still) not possible, as the degeneration is presumably heterogeneous and involves various deposits of protein structures (amyloid, tau, and other protein aggregates).

4.4.5 Dizziness in Acute and Chronic Cervical Syndrome

It remains still controversial whether dizziness can originate from the cervical spine. This has been debated among medical specialties for many years. Proponents consider this dizziness a possible cause of dizziness with balance disorders. Opponents reject the diagnosis because there is no reliable clinical test that can distinguish this dizziness from other pathological dizziness syndromes. The interaction of eye, head, and body movements in space is controlled by the interplay of stimuli from neck sensors and vestibular and visual signals. This interaction ensures that we can perceive the direction of our movements and that our center of gravity is finely tuned, i.e., that the body is stable while walking and standing. Studies investigating whether there is dizziness originating from the cervical spine have found brief episodes of dizziness in patients with acute neck pain syndrome with markedly restricted neck mobility, triggered only by rapid, not slow, head movements, and resolving within a few days as neck mobility improves. Similarly, such brief episodes of dizziness occur in patients with chronic neck pain that restricts neck mobility, again only with rapid, not slow, head movements. Gait analysis shows a slowed and cautious gait pattern, which is even more pronounced with rapid head turns. The disease model for this form of dizziness corresponds to that of functional dizziness. The extremely rare cervical second-long dizziness also arises from a discrepancy between incoming signals from vestibular, visual, and somatosensory sensory stimuli and the movement patterns stored in the brain. Due to restricted neck mobility, a stimulus pattern is sent to the brain that differs from the stored pattern, which assumes normal cervical spine mobility. Whether there are transitions to functional dizziness in chronic cervical spine syndrome is not yet clarified.

Given the controversial views, it should be emphasized once again that acute or chronic cervical syndrome is only very rarely the cause of dizziness; much more often, other dizziness disorders are responsible. It is the other

way round: the increased muscle tension and pain in the neck is secondary because patients with vertigo and dizziness often experience motion intolerance. Therefore, they try to minimize head movements by contraction of neck muscles. These symptoms, however, have to be treated, of course.

5

What Can Those Affected Do Themselves?

In cases of very severe vertigo or dizziness, prolonged complaints, or frequently recurring symptoms, affected individuals should definitely consult a physician to determine the cause of the dizziness, especially since, although less commonly, a serious illness such as a stroke may be behind the symptom. This also applies to the sudden onset of balance or gait disturbances. As a rule of thumb: the more sudden and intense the onset of vertigo, dizziness or balance disturbance occurs, the more urgently affected individuals should seek medical evaluation. It is helpful to become familiar with possible warning signs of a stroke.

In terms of acute onset of symptoms, the Face-Arms-Speech-Time Test (FAST test) is a self-test developed for laypersons that summarizes the most important stroke symptoms—drooping mouth corner ("face"), arm paralysis ("arms"), speech or language disturbance ("speech")—and emphasizes the urgency of clarification ("time") should one or more of these symptoms occur.

For all questions concerning the underlying disease, the treating physician should be the first point of contact for the patient. It is helpful to bring your own medical records to the doctor's appointment and, in particular, to ensure you have an up-to-date list of medications. If such a list is not available, it is advisable to bring the medication packages with you.

Affected individuals are often confronted with all kinds of well-intentioned but not always professional advice, which may promise a quicker improvement. Such advice often leads to uncertainty. After a diagnosis has been made, further, often unhelpful or even counterproductive diagnostic

M. Strupp et al., *Patient Guide to Dizziness and Balance Disorders*,
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measures should be avoided. These can also unsettle the patient and negatively affect the course of therapy. Patients should consistently follow medical recommendations or inform the treating physician if implementation is not possible. For example, if certain medications are not tolerated, alternative preparations can be sought, or an ineffective treatment can be replaced by another. Smartphone apps now play an important role in the self-management of vertigo, dizziness and balance disorders. Patients should pay attention to medical guidance to ensure the correct therapeutic measures are selected. In many diseases, taking an active role in managing symptoms gives patients the feeling that they can support the healing process through their own efforts. This active participation often has a positive effect on the course of the illness.

In Germany, there are also various self-help groups for dizziness and balance disorders, such as the German Hereditary Ataxia Society e. V. (DHAG). Likewise, the Vestibular Disorders Association (VeDA) in the US and the Menière's and Vestibular UK patient organization are valuable resources. These offer affected individuals and their relatives the opportunity to exchange ideas with others who are directly or indirectly affected and to learn more about the clinical picture and treatment options.

A healthy lifestyle—that is, regular and sufficient sleep ("sleep hygiene"), avoiding irregular daily routines, drinking an adequate amount of fluids daily, eating regular, healthy meals, not consuming alcohol indiscriminately, reducing stress through relaxation techniques, and daily physical activity ("The more you walk, the longer you walk!")—are important measures for many dizziness disorders to reduce episodes of vertigo and dizziness and better cope with dizziness symptoms.

Nevertheless, constant self-monitoring of vestibular symptoms can often be counterproductive if insignificant changes in one's own body are overemphasized. This can be, for example, a single misstep or a brief episode of swaying dizziness when standing up, or even a momentarily irregular heartbeat, which in turn can lead to increased self-awareness.

Patients and their relatives understandably have great hope that the doctor can make the correct diagnosis based on the existing symptoms. Such a diagnosis allows for counseling of the patient and relatives regarding the expected course of the disease and available treatment options. Sometimes, an explanatory conversation about the cause of the symptoms is already sufficient, and no further treatment is necessary. At the same time, patients can expect open communication from their treating physician. For many conditions, the underlying causes are only partially or not at all known. This also applies to some vestibular and balance disorders. The same is true for

effective treatment methods: for certain diseases, they are (as yet) unknown. The treating physician should actively address this.

Finally, consistently keeping a vertigo and dizziness and, if applicable, headache diary is an important component in the care and treatment of patients with these complaints. Patients should bring this diary to their doctor's appointments. Such a diary can also be kept via a smartphone app and helps the physician assess the frequency and severity of symptoms and adjust treatment accordingly.