Introduction: How to approach the patient with neurological disease

The keystone of neurological diagnosis is the patient’s history. The neurological examination is most valuable when it is guided by hypotheses that the physician formulates as the history is elicited. For example, if a right-handed patient reports an episode of difficulty in speaking, the examiner will search for signs of left hemisphere dysfunction: right hemiparesis, hemivisual and hemisensory disturbance. In a patient who reports wrist-drop since awakening from an alcoholic stupor, the examiner will look for signs of a radial nerve palsy. This hypothesis-driven approach should not, however, blind the examiner to the occasional surprise on the exam that entirely changes the nature of the diagnosis: for example, discovery of up-going toes or of mental status changes in a patient who complains of muscle fatigue. Indeed, the initial evaluation of all patients demands a thorough and systematic evaluation.

Formulating the neurological diagnosis usually consists of three main steps. First, define the functional deficits, based on the findings from the history and examination (e.g., the patient has a left hemiparesis, double vision and right ptosis). Second, localize the site and extent of the lesion within the nervous system, based on the examination (e.g., the right cerebral peduncle and left oculomotor nerve, which traverses it — Weber’s syndrome). Attempt to account for functional deficits by as few lesions as possible, applying Occam’s Razor, which states that the simplest of all possible answers is usually correct. Note, however, that involvement may be focal, multifocal or diffuse. Third, identify the pathological process from the history of how the symptoms evolved (e.g., onset over the course of minutes, suggesting acute ischemia). Disease processes may be static or progressive, with an acute, subacute or chronic course. Beware that some conditions, such as metabolic diseases of the brain, epilepsy and migraine have no circumscribed anatomical substrate, and are best described in pathophysiological rather than anatomical terms.

One great aid to recognizing common neurological disorders is a basic knowledge of neuroanatomy. Review the visual pathways, and the major descending and ascending tracts. Such knowledge makes it easy to understand why homonymous visual field defects are due to lesions behind the optic chiasm. Similarly, a patient who presents with weakness and spasticity of the right leg and right arm, loss of position and vibration sense in the right leg and arm, and decreased pain and temperature sense in the left arm and leg must have a lesion in the right cervical cord. Such a lesion would affect the ipsilaterally descending motor tracks and ascending dorsal column proprioception tracks, but also the crossed spinothalamic ascending pain and temperature fibers (syndrome of Brown-Sequard).

Also, learn to recognize common neurological syndromes. Thus, weakness accompanied by spasticity, hyperreflexia, and Babinski responses (up-going toes) suggests pyramidal tract dysfunction. Proximal muscle weakness and fatigue with intact sensation characterizes disorders of muscle or the neuromuscular junction. Loss of sensation and paraesthesias (tingling sensations) in a “stocking-glove” distribution suggests a peripheral neuropathy (e.g., in association with diabetes). Alternatively, numbness and weakness may correspond to the anatomical region supplied by a spinal nerve root or a peripheral nerve. For example, loss of sensation restricted to
one little finger and the adjacent half of the ring finger is most likely due to an ulnar nerve disorder; numbness in this distribution would not likely be central. Whenever a patient presents with a cranial nerve palsy, attempt to determine whether there are associated “long-tract” signs, such as pyramidal tract dysfunction; as in the example of Weber’s syndrome provided above, the combination accurately localizes the lesion. Alternatively, the cranial nerves may be affected after they have left the brainstem, for example, by meningeal disease.

Avoid the mistake of regarding the neurological examination as being a fixed entity. The time-honored tests of the neurological examination have survived by eliciting reliable clinical signs. However, powerful techniques employed by neuroscientists are now providing a firm physiological basis to some of the signs that we elicit at the bedside, but also requiring us to re-evaluate the significance of others. Current tests of the nervous system are incomplete and there is still plenty of room for imagination and originality in the bedside clinical evaluation.

**Examination tools**

In an emergency, a neurological examination can be performed without any examination tools. Indeed, a good deal can learned simply by careful observation of how the patient talks, spontaneously moves, and walks. Nevertheless, it is worthwhile investing in a few simple instruments.

Always carry a card to test visual acuity, and a flashlight to test the pupils. Many modern facilities provide wall-mounted ophthalmoscopes, but it becomes easier to use a familiar, personal instrument. A reflex hammer is essential and although the tomahawk design remains popular, it lacks the lever-arm provided by Queen Square or Mayo Clinic hammers, which are superior in eliciting hypoactive reflexes. Learn to tap lightly on your finger as it is pressed against the tendon of the muscle being tested. A 256 cps tuning fork is useful to test hearing and a 128 cps tuning fork to test vibration sensation. A tuning fork also provides a convenient object to assay the thermal sensation, especially if cooled, or warmed, by tap water. Maintain a pocket supply of cotton-tipped applicators and tongue blades. The former are used to test the corneal reflex, posterior pharyngeal sensation and, when broken, sharp sensation. The tongue blades are also useful during testing of pharyngeal sensation. An open safety pin provides a sharp and a dull testing stimulus, but these pins should only be used once to avoid transmitting disease. Do not penetrate the skin or produce blood spots. The sensory pinwheel with pins on rolling spokes is obsolete because of the possibility of transmitting disease. A stethoscope can be applied to detect cervical or cranial bruits. Cue cards of dermatomes, peripheral nerve innervations, and other neuroanatomical facts are often helpful.
TABLE 1. AN OUTLINE OF THE NEUROLOGICAL EXAMINATION

Aspects of the General Exam

Measure pulse and blood pressure, supine and standing
Neck: listen for bruits; define range of movements; palpate for tenderness
Head; note tilt or turn; measure circumference (infants); note symmetry; palpate for tenderness and masses, listen for bruits
Back: note spinal curvatures; palpate and percuss for tenderness; determine mobility

Mental Status Testing

Determine level of education; present and previous employment
Alertness and attention
Orientation
Language
Memory: short-term, long-term, and fund of general knowledge
Calculations
Spatial perception
Reasoning and abstractions

Cranial Nerves

CN I  Smell coffee or vanilla

CN II  Visual acuity
Visual fields
Optic fundi

CN III, CN IV, CN VI
Range of movement and alignment of the visual axes
Fixation in center and eccentric gaze positions
Saccades, smooth-pursuit, vestibulo-ocular reflex, vergence
Pupils
Lids

CN V  Sensory testing of all 3 divisions
Corneal reflex
Muscles of mastication
Jaw jerk

CN VII  Facial symmetry and strength
Taste over anterior 2/3 tongue

CN VIII
Sensory threshold (compared with examiner)
Air conduction vs. bone conduction
Weber lateralization test
(Vestibular reflexes are considering under eye movements and balance)

CN IX, CN X
  Phonation and enunciation
  Posterior pharyngeal sensation
  Symmetry of palate elevation
  Gag reflex

CN XI
  Sternomastoid and trapezius muscles: bulk and strength

CN XII
  Tongue bulk, strength, adventitious movements

**Motor Function**

Screening tests for motor imbalance: pronator drift and forearm-rolling test
Gait, station,
Bulk
Tone
Power
Coordination
Adventitious and abnormal movements

**Sensation**
Elementary Modalities:
  
  Pain
  Light touch
  Vibration
  Joint position sense
  Thermal Sense

Discriminative Modalities:
  Two point discrimination
  Stereognosis
  Graphesthesia
  Baresthesia
  Double simultaneous stimulation

**Reflexes**
Upper limbs: Biceps, triceps, supinator, finger jerks
Lower limbs: Patellar, ankle jerks

0 = absent
+ = hypoactive
2+ = normal
3+ = hyperactive
4+ = hyperactive with clonus

Plantar responses
Also test Hoffmann's sign, grasp, snout, and palmomental reflexes.

Autonomic Function

Blood pressure and pulse
Sweating
Rectal tone
ASPECTS OF THE GENERAL EXAMINATION

Much of the neurological examination overlaps what an internist would consider to be general examination. A few features are especially important for neurology, including: blood pressure and pulse (measured both supine and after standing for two minutes), temperature and respiratory rate, auscultation of the heart for murmurs or irregularities that might be a source of emboli or syncopal episodes, auscultation of the neck for carotid or vertebral artery bruits. Occasionally, careful skin examination reveals signs of septic emboli or neuroectodermal changes such as might occur in tuberous sclerosis or neurofibromatosis. If a neurology student is admitting a patient to the inpatient neurology service, a complete general examination is usually performed in addition to the neurological examination, since that team is serving as the overall physician for the patient. In clinic or on a consultation, the examination is usually limited to a neurological exam with certain important components of the general physical examination, especially the vital signs. In patients who complain of pain in the head, neck, or back, determine the location of the pain and whether palpation or percussion exacerbates it.

The sequence in which the neurological examination is performed is largely a matter of personal preference and experience. In the beginning, students usually start by evaluating mental status, then test the cranial nerves, motor function, reflexes, sensation, and gait (Table 1). With more experience, different portions of the examination are selected and grouped for efficiency. In this manner, the patient has less need to lay down, stand up, and roll over repeatedly. The discussion below follows the outline in Table 1.

MENTAL STATUS

The mental status examination evaluates how well a patient thinks and communicates. It should not be omitted, even in a screening neurological examination. An initial impression is possible during history taking, when it is important to assess the patient’s level of education and inquire about current employment and the “best” job ever held. Such information is important for interpretation of the patient’s performance on cognitive tests. If there is a question of declining cognition, then the account of family or friends (by a telephone call, if necessary) is often decisive. Tact is always required in testing mental functions.

When screening mental status, assess orientation, short term and long term memory, ability to speak spontaneously, to repeat, to comprehend and follow commands, to read, to perform a constructional task, and to calculate. A standardized approach, such as Minimental or MoCA Tests, provides the advantage that it can be repeated at a later date, and scores compared.

Alertness and attention
Before starting testing, note the patient’s level of alertness and attention, mood, and any evidence of thought disorder, delusions, or hallucinations. Ask the patient to spell “world” forwards and then backwards. If they cannot do this, try “hand” and “cat”. In patients with limited education, ask them to count down from 20 to 0. Bear in mind that impaired performance may reflect impaired concentration or disturbance of language (aphasia).

Orientation
Ask the patient their name, their city, the month, day and year. If they can not report the precise date, ask them the day of the week or what holidays are in proximity to the current date. The phrase “Oriented x4” usually adds a point for awareness of reason for hospitalization.
**Language**
Before assessing language, determine if the patient is right- or left-handed. Listen to the patient’s speech, noting rate, rhythm, inflection, articulation, pronunciation, and effort of initiation of speech. Listen for paraphrasic errors (including neologisms), the word content (substantive words compared with filler words), echolalia (repeating the examiner, without reason or instruction to do so), and perseveration (repeating the same words inappropriately, having once used them appropriately). Give the patient a command with some complexity, such as “close your eyes, open you mouth and put your left thumb on you right ear.” This command requires both memory, ability to perform motor tasks (praxis) and good speech comprehension. Give the patient a sentence or phrase to repeat, such as “no ifs, no ands, no buts”, or “The only thing we have to fear is fear itself.” Provided their eye sight allows, ask the patient to read aloud, and perform, the printed command: “Make a fist with your left hand”. Pick up some large print material at hand, and ask the patient to read the material. In cases of questionable aphasia, give patients a more challenging task, such as “name all animals that begin with the letter A”. A normal person should be able to come up with at least 10 animals in 30 seconds.

Distinguish between dysarthria and aphasia. Dysarthria is a slurring or mumbling of words, whereas aphasia reflects a problem with language itself. With aphasia the words are either used excessively sparingly, or words are substituted, or there is a lack of normal grammatical flow and rhythm (prosody) to normal speech. So-called anterior aphasia (or motor or Broca’s aphasia) presents with difficulty in speaking words, very sparse speech, occasional word substitutions and frustration on the part of the patient. Posterior aphasia (sensory aphasia, receptive aphasia, Wernicke’s aphasia) presents with fluent sounding speech from a distance, but with much of the speech content being nonsense when listened to up close. There will be many word substitutions. A patient with pure Broca’s aphasia can comprehend and follow commands correctly, where as patients with Wernicke’s aphasia can not comprehend verbal commands.

Broca’s area is located in the inferior lateral frontal gyrus and Wernicke’s at the superior posterior portion of the temporal lobe, usually in the left hemisphere of right handed people. Interruption of the arcuate fasciculus causes inability to repeat. This is called a conduction aphasia. An aphasia like a Broca’s, but with preserved ability to repeat is referred to as a transcortical motor aphasia. A Wernicke-like aphasia with preserved ability to repeat is called transcortical sensory aphasia. Inability to read is dyslexia, and to write dysgraphia. It is important to realize, however, that current concepts of language consider the brain as a parallel processor, and the conventional view presented above, although useful clinically, may be simplistic.

**Memory**
At the start of the examination, give patients three words or brief phrases to remember, mixing verbs, nouns, and adjectives, and unrelated to each other. Pick your own favorite phrases that you can use over the years (e.g., blue, bicycle, Ringo). Ask the patient to repeat them immediately after you give them to them, verifying registration of material. At approximately five minutes, ask them the three phrases. If they cannot remember all three, give the phrases to them again and ask five minutes later. Normals should be able to remember all three phrases without difficulty.

If memory is a significant complaint or appears to be abnormal, give strings of numbers, spoken steadily without grouping (i.e., not as telephone numbers). People with normal short-
term memory should be able to recall 8 digits forward.

Ask a few remote memory questions, which may be preserved when short-term memory is impaired, such as where a person was born. Assay memory for current events, especially disasters such as earthquakes and acts of war. Test their fund of knowledge by asking them to name the current President and his predecessors, in order. For more detailed testing of memory, formal neuropsychological tests were standardized stories with key elements can be given. Some patients have impaired visual memory with intact verbal memory. This can be tested by drawing a complex figure, showing it to them and asking them to reproduce it from memory.

Calculations
Ask the patient to subtract 7 from 100 and then subtract 7 from the result, repeated 5 times. Write down their answers. Ask: “How many nickels in a dollar, twenty-five”. If they seem unable to perform these calculations, ask them: “If I gave you a dollar and took back 7 cents, how much would I have?” and “if I took 7 more cents from that?”.

Spatial Perception
The simplest maneuver is to ask a person to draw an imaginary circle clock in the air and point to where 12 o’clock, 5 o’clock and 9 o’clock would be in the circle. If pencil and paper are handy, the patient can draw the clock and put on the numbers. People with parietal deficits tend to crowd all the numbers into the right side of the drawing (assuming that the right hemisphere is the non-dominant hemisphere). Ask the patient to copy two overlapping boxes or pentagons. Draw about 20 horizontal lines on a sheet of paper and ask the patient to draw a vertical line through the middle of each. If there is visual neglect, then the patient tends to bisect the lines to the right of the true center mark, and may ignore the lines on the left-hand side of the sheet. If the examiner omits a test that’s spatial perception during the screening neurological examination, sooner or later a large non-dominant hemisphere parietal tumor, stroke or hemorrhage will be missed.

Reasoning
Although some judgement of the patient’s ability to reason is usually possible based on the history of illness they provide, it is sometimes useful to test reasoning. For example, ask what “people who live in glass houses shouldn’t throw stones” means, or “why is it a bad idea to shout fire in a crowded movie theater?”, or “what would you do if you found a wallet lying on the street?”. Depending upon the context of the problem, patients may need to be asked specifically whether they are having hallucinations, delusions, strange thoughts or ideas, a sense that people are talking about them behind their backs or putting thoughts into their heads. Such inquiries may provide insights about underlying schizophrenia or organic psychoses. Ask the patient to rank their mood on a scale of 1-10. If the patient is significantly depressed, remember to explicitly ask about whether they are planning asks of suicide. Asking will not make it so, and may uncover the route to preventing a lethal act.

THE CRANIAL NERVES

CN I
The sense of smell depends on chemoreceptors located on the superior nasal concha and facing nasal septum. The central processes of these cells project through the cribiform plate of the ethmoid bone to the ipsilateral olfactory bulb, on the under surface of the frontal lobe. The olfactory tract runs beneath the orbital surface of the frontal lobe in the olfactory groove and terminates in piriform cortex of the medial temporal lobe (primary olfactory cortex), the amygdala, and septal nuclei.

Test smell one nostril at a time using coffee. Avoid using substances with pungent odors. Ask the patient to close his or her eyes, warn them that you are going to close one nostril (otherwise it is a bit alarming to be poked in the nose) and ask them if they can identify the smell. Use the same object or a different object to test the other nostril. Loss of smell is usually due to nasal allergies, upper respiratory infections or consequences of smoking. Loss of smell may also be a consequence of head injury, and of advancing age. Rarely, subfrontal tumors impair smell by pressing on the olfactory tracts.

CN II

To accurately localize disturbances of the visual pathways, it is useful to review certain important anatomical points. The optic nerve is formed by the central processes of the retinal ganglion cells, which converge towards the optic disc. In the optic nerve, fibers from the upper retinal quadrants occur the upper parts of the nerve, fibers from the lower retina in the lower parts, and macular fibers in the center. At the chiasm the fibers undergo a partial decussation: fibers emanating from the temporal halves of the retina continue without crossing, while fibers from the nasal halves cross in the chiasm to the optic tract of the opposite side. The optic chiasm lies above the pituitary gland, below the suprachiasmatic recess of the third ventricle and the hypothalamus, and anterior (in most cases) to the pituitary stalk. From the optic chiasm, most axons continue as the optic tract to reach the lateral geniculate body-- the visual thalamus. From lateral geniculate nucleus, axon sweep posteriorly around the lateral aspects of the lateral ventricles as the optic radiation, which projects to striate cortex (primary visual cortex, V1) of the occipital lobe. Fiber representing superior retina turn posteriorly and run through the deep parietal white matter, but fibers representing lower retina loop forward around the temporal horn of the lateral ventricles forming the "Meyer's loop". Fibers carrying vision from the upper and lower retina terminate in the upper and lower bank of the calcarine fissure respectively, with the horizontal meridian running along the base of the fissure.

First test visual acuity; this is an essential part of every screening neurological examination. Test each eye separately with refractive correction. If the patient lacks spectacles, use a pinhole to correct refractive errors (you can make one by making some small holes in a piece of paper). Use a Rosenbaum card or equivalent at a viewing distance of 14 inches. If vision is poor, ask the patient to count your fingers, or detect a flashlight, presented centrally and in the four quadrants of the visual field.

Second, screen for visual field defects by confrontation. Sit facing the patient, at the same level. Test each eye separately, using yourself as a control. First, cover the patient's left eye while the patient fixes on your left eye; close your right eye. Briefly present 1-3 fingers from each hand simultaneously in opposite quadrants of the visual field (e.g., upper nasal and lower temporal). During each presentation, ensure that the patient is fixed on the pupil of your left eye. Most patients soon understand the nature of the test, which is superior to presentation of a wagging finger, which does not demand a quantitative judgement. In patients who have a visual
field defect, map out the extent of the field defect using a red test object such as a match; this technique is especially helpful with central field defects and in comparing the size of the patient's blind spot with your own. Classically, ischemia of the retina and anterior optic nerve cause altitudinal defects (above or below the horizon), optic nerve demyelination causes a central scotoma (visual defect), optic chiasm lesions cause bitemporal field defects, and retrochiasmal lesions cause homonymous (same-sided) hemianopia. Optic tract lesions cause defects that are very dissimilar in the two eye (incongruous defects) where as striate cortex lesions cause defects that are very congruous. Temporal lobe lesions causes superior homonymous defects (“pie in the sky”), whereas parietal lobe lesions cause more inferior homonymous defects. Retrochiasmal lesions should not cause impairment of visual acuity.

Third, examine the optic fundus with an ophthalmoscope. A certain amount of practice is necessary to learn to see the fundus, particularly in poorly cooperative patients. The patient is instructed to look, at a point, over the examiner’s shoulders and continue to stare in that direction without moving the eyes. The ophthalmoscope light is usually adjusted to a medium sized bright round white light. The examiner moves closer to the eyes, with a slightly extended finger to avoid bumping in to the patient’s eye, all the time looking through the ophthalmoscope. The redness of the retina becomes visible. The focus nob of the ophthalmoscope is then turned until the vessels are in clear focus. The examiner then looks around to find the optic disk. A judgement is made about the sharpness and color of the disk, the caliber and smoothness of the veins and arteries, and presence or absence of any unusual pigmentation, blood or puffy white/yellow exudates. If abnormalities are present, they are recorded by description and location such as a “flame shaped hemorrhage 2 disks diameters lateral at approximately 10:00 in the left eye”. Particularly important in neurology is the presence of papilloedema. This indicates pressure in the optic nerve sheath, which is usually reflective of increased intracranial pressure. Papilledema may not develop until 12-24 hours after increased intracranial pressure, so absence of papilledema is not reassuring in the acute situation. Early papilledema may present only with engorgement of the retinal vein (the darker, larger structures). Fine pulsations can be seen in the normal retinal veins. Presence of these pulsations argue strongly against papilloedema. Absence of pulsations, however, does not necessarily indicate presence of papilloedema. The next stage includes blurring of the disk margin. The disk margin may bulge out instead of being its usual concavity in the center. This may be perceived by having to turn the focus of the ophthalmoscope more than 2 diopters when focusing on the edge and the center of the disk. Presence of hemorrhages or exudates, in conjunction with blurred disk margins is a serious neurological sign and call for an urgent evaluation. Loss of vasculature in the retinal fields is suggestive of retinal vascular disease, which sometimes may co-exist with cerebral vascular disease. Pallor of the optic disk is a sign of optic atrophy, either on an ischemic basis or in association with demyelinating disease: optic neuritis or multiple sclerosis.

**CN III, CN IV, and CN VI**

The sixth (abducens) cranial nerve innervates the lateral rectus muscle, which turns the eye out (abduction). The fourth (trochlear) cranial nerve innervates the superior oblique muscle, which helps to pull down the abducted eye. The third (oculomotor) cranial nerve innervates the remaining muscles, the pupil and the levator palpebrae superioris, which elevate the lids.

First test the range of movement of movements of each eye, taking it into each of the 6 directions of gaze. Observe if the eye appears to move fully or is restricted. Then ask the patient to follow with both eyes and report when double vision is noted; this direction of gaze
corresponds to activity of the weak muscle. Abducens nerve palsy causes horizontal diplopia on ipsilateral gaze and when viewing distant objects. Trochlear nerve palsy causes vertical diplopia when the subject looks down and inward with the affected eye. Complete oculomotor nerve palsy causes ptosis, a dilated (mydriatic) pupil, and an eye that is “down and out” (like an intern who has been up all night); however, the defect may be partial.

Next, test the ability of the patient to sustain steady fixation of a pen-tip with the eyes close to central position and then looking eccentrically – far laterally, up or down. Note any nystagmus (oscillations of the eyes). Unsustained “end-point” nystagmus is common in normal subjects. Nystagmus is a significant finding when it is present with the eyes close to central position, when it is sustained, and when it is vertical in direction (especially down-beating); such nystagmus often points to cerebellar disease, unless the patient is taking medicines such as anticonvulsant.

Then test saccadic eye movements by asking the patient to look between your nose and the pen tip, which is held to one side (for horizontal saccades) or above or below (for vertical saccades). Note if the movements are initiated promptly, if they are fast, and if they are accurate. Difficulty in initiating saccades occurs with cortical lesions, slow saccades occur with degenerative diseases (such as progressive supranuclear palsy) and inaccurate saccades occur with cerebellar disease.

Test smooth-pursuit movements as the patient attempts to track your pen-tip smoothly moved horizontally and then vertically. Older subjects, patients on sedative medications, and cerebellar disorders impair smooth pursuit movements.

Test vestibular eye movements either with rapid head turns or with caloric stimulation. In alert patients, ask then to fix their gaze on a distant target and then make a sudden, small, rapid head turn. Only the vestibulo-ocular reflex can compensate for such head rotations and, if deficient, a visually mediated saccade follows -- it being the sign that the vestibular movement was lacking. Alternatively, after first checking that the tympanic membranes are intact, irrigate 1 ml of iced water into one ear, and then position the supine patient with the head 30 degrees up. A small-amplitude nystagmus should be evoked. After several minutes, repeat by stimulating the other side. This “minimal ice-water caloric test” compares the threshold for a responses, which is best evaluated behind magnifying (Frenzel) goggles. The stimulating dose can be increased if no response is obtained.

Then, test vergence eye movements by asking the patient to fix upon their thumb nail as it is brought towards them along the mid-sagittal plane. This evokes the near triad: convergence, pupillary constriction, and accommodation of the lens.

Observe the resting size and shape of the pupils in room light, as the patient fixes upon as letter or other symbol at a distance to impose a fixed state of accommodation, and measure pupillary size. Then test the pupillary responses to light. Move a penlight in from the temporal side to shine the beam into one eye and test the direct pupillary light reaction. Remove the beam and repeat for the other eye. It is most convenient to compare the direct and consensual responses by rapidly moving the pen light from one eye to the other, and holding it at its new location for about 5 seconds. If the pupil dilates when the flashlight is shone into it, then the prior consensual response was greater than the present direct response; this is "relative afferent pupillary defect" and indicates an abnormality of the anterior visual pathway of that eye. Horner’s syndrome consists of miosis, ptosis and anhydrosis. The most reliable component is the miosis, and this is best demonstrated by putting the patient in a dark room. Some patients with Horner’s syndrome have an underlying apical lung cancer that disrupts the sympathetic
innervation of the pupil. Adie’s pupil is usually mydriatic, and responds poorly to light. Diagnosis rests on demonstrating denervation hypersensitivity with 0.125% pilocarpine. Forty-five minutes after applying two drops to each eye, only the tonic pupil will have constricted. Unilateral pupillary dilatation may be the first sign of compression of the oculomotor nerve by aneurysm or during uncal herniation, because the parasympathetic fibers lie peripherally in nerve. Often, however, anisocoria results from the effects of mydriatic drugs such as atropine. To determine whether a fixed, dilated pupil is due to CN III palsy or mydriatic agents, apply pilocarpine 1% eye drops; they will cause the former to constrict, but not the latter. Argyll-Robertson pupils due to neurosyphilis are now rare; however small, irregular pupils that constrict with a near stimulus but not with light are encountered in other disorders, such as with diabetes.

Observe the resting position of the eyelids with the eyes in mid position and in each direction of gaze, noting whether the lids follow vertical eye movements. When the eyes are in mid position, the upper lid normally just covers the upper cornea but the lower lid lies below the cornea. Compare the two palpebral fissures and measure any differences. Observe if there is any contraction of the frontalis muscles, which may be seen when patients with partial ptosis attempt to open their eyes. In such cases, the patient may be unable to open the eyes if the frontalis muscle is fixed with a finger. Also note any proptosis.

Ptosis -- drooping of the eyelid -- is caused by several processes. Lid dehiscence is common in the elderly and is of no neurological significance. Disease affecting the extraocular muscles and lids, such as ocular myopathies, usually produces symmetrical limitation of the lids and vertical gaze. Neuromuscular disease, especially myasthenia, often produces variable, asymmetrical ptosis that worsens during sustained upgaze. Third nerve palsy commonly causes monocular ptosis except when the location is nuclear, in which case ptosis is bilateral. Supranuclear lesions cause bilateral ptosis; most frequently this occurs with right hemispheric lesions. Eyelid opening apraxia describes patients who can open their eyes spontaneously, but cannot do so on command. Blepharospasm consists of paroxysmal, involuntary contractions of the orbicularis oculi; it is a form of focal dystonia and may reflect a dopaminergic disorder in the basal ganglia.

**CN V**

The trigeminal nerve mediates sensation over the skin of the face, teeth, and the mucous membranes of the mouth and nasal cavities. It also innervated the muscles of mastication (chewing). An important anatomical point is that the fibers conveying painful (nociceptive) sensation project caudally in the spinal cord tract of CN V, before synapsing in the lower medulla and cervical cord. On the other hand, fine touch is mediated by neurons that synapse in principal nucleus in the pons, which is also the location of the motoneurons. The motor and sensory roots emerge together on the ventrolateral surface of the mid-pons. The trigeminal nerve passes through the pontine cistern and the sensory portion swells to form the trigeminal ganglion, which rests in a small groove on the apex of the temporal pyramid bone, empouched in dura. After piercing the dura, the trigeminal nerve splits into three division: the ophthalmic (V1), the maxillary (V2), and the mandibular (V3). The motor portion of the nerve runs with V3. The ophthalmic division passes through the lateral wall of the cavernous sinus and, after passing through the superior orbital fissure, it supplies the lacrimal gland, the skin of the forehead to the vertex, cornea, conjunctiva, sphenoid sinus and mucous membrane of the anterior part of the nose. The maxillary division runs below the cavernous sinus, passes through the foramen rotundum, and emerges through the infraorbital foramen to innervate the skin of the maxilla,
anterior part of the temple and teeth of the upper jaw, and mucous membrane of the posterior nasal cavity and hard palate. The mandibular division exits the skull through the foramen ovale where its splits into auriculotemporal, lingual, long buccal, inferior dental, and masticatory branches. The last of these supplies the muscles of mastication: temporalis, masseter, external and internal pterygoids. The distribution of the cutaneous branches is summarized in Figure.

In examining the sensory functions of the trigeminal nerve, it is most important to test pain and the corneal reflex. Use a sterile pin and ask the patient to compare the sharpness of the stimulus when it is applied to the right and then the left side of each of the three divisional dermatomes. Note that V1 extends up to the vertex, but that the angle of the jaw is not supplied by the trigeminal nerve. Test the corneal reflex by touching the cornea (not the sclera) with a wisp of cotton while the patient looks up. Observe both the direct and consensual reflexes, and ask the patient if the sensation is the same on both sides.

Test the motor function of the trigeminal nerve by palpating the masseter and temporalis muscles as the patient clenches the teeth. Ask the patient to open the mouth and look for a deviation; the jaw will deviate laterally to the side of the weak pterygoid muscle. Attempt to elicit the jaw jerk by striking your finger placed on the patient's chin with the mouth slightly opened. Many normal individuals lack a jaw jerk but, when it is increased, it may help determine whether a corticospinal lesion lies above the level of the cervical spine.

Disorders of facial sensation may be due to local processes within the face and paranasal sinuses, or within the cavernous sinus (e.g. by tumor, aneurysm or inflammation). Sustained facial pain (typically in V1) may be accompanied by diplopia -- "painful ophthalmoplegia". If the trigeminal ganglion is involved by herpes zoster, facial pain is accompanied by skin rash. Inflammation or tumor of the temporal bone (e.g. nasopharyngeal carcinoma) may involve the trigeminal ganglion causing facial pain accompanied by ipsilateral sixth nerve palsy (Gradenigo's syndrome). Trigeminal neuralgia or tic doloureux is characterized by lancinating pains that usually affect one divisions (often V2), and are triggered by cold or shaving. The trigeminal nerve is also often affected by brainstem disorders, because of the long rostrocaudal extent of its spinal nucleus. Thus, loss of facial sensation is a feature of Wallenberg's syndrome, in which lateral medullary infarction involves the spinal tract of CN V and its nucleus.

**CN VII**

The facial nerve supplies the muscles of facial expression. The intermediate nerve, which runs with it, carries visceral efferent and afferent fibers. From the facial nucleus, which lies in the caudal pons, axons pass dorsomedially, hooking over the abducens nucleus, just beneath the fourth ventricle, and then run ventrolaterally to leave the brainstem between the olive and the inferior cerebellar peduncle, in the cerebellopontine angle. The facial nerve and intermediate nerve enter the internal auditory meatus with CN VIII and at this point, the intermediate nerve gives off the greater superficial petrosal nerve, which supplies the lacrimal gland. In the facial canal, CN VII runs close to the wall of the tympanic cavity, where it gives off fibers to supply the stapedius muscle. Subsequently, the intermediate nerve gives off the chorda tympani branch, which innervates the submandibular and sublingual salivary glands, and taste over the anterior two thirds of the tongue. The facial nerve exits the skull from the stylomastoid foramen, penetrates the parotid gland where it forms a plexus, and splits into branches to innervate the facial muscles. An important point about the innervation of facial nerve motoneurons is that neurons innervating muscles above the palpebral fissure appear to receive bilateral corticopontine projections, whereas muscles below the palpebral fissure receive crossed
projections. The facial muscles are involved in the blink response to corneal stimulation (the corneal reflex).

In examining the facial nerve, two main judgements must be made: (1) Is there any facial weakness; and (2) if there is facial weakness, is it due to an upper motor lesion (e.g., following a hemispheric stroke causing hemiplegia) or a lower motor lesion (peripheral facial palsy). The first question can be addressed by looking for asymmetry of the patient’s nasolabial folds (from the edge of each nostril to the corner of the mouth) at rest and when asked to "show me your teeth". Asymmetry, especially of the movement, indicates facial weakness. The second question can be answered best by noting the symmetry of eyebrow raising. Weakness of the frontalis muscle indicates a lower motor neuron lesion. In addition, routinely test eye closure (orbicularis oculi), blowing out the cheeks (buccinator), pursing the lips (orbicularis oris), and movements of the skin of the neck (platysma). Ask the patient to blink repetitively, and note if synkinetic movements of other facial muscles occur such as "jaw-winking". This Marcus Gunn sign reflects from aberrant regeneration (“misrouting”) of the facial nerve following a prior palsy. Note if tearing is present; not only is this helpful in localization, but it is also important for managing eye care if the patient cannot close the lid adequately. Test taste by holding the protruded tongue with cotton gauze and placing test solutions on one or the other side of the anterior two thirds of the tongue using cotton-tip applicators. Make up separate solutions with salt, sugar, sour (lemon) and bitter crushed acetaminophen tablets. Instruct the patient to indicate the taste of the stimulus by pointing to written labels.

In addition to the corneal reflex, test the glabellar reflex by tapping the forehead between the eyes; the orbicularis oculi muscles reflexly contract. In normal subjects, this response habituates with repeated stimuli but persists in some patients with Parkinson’s disease. Light percussion over the lips may induce contraction of the levator anguli oris muscles (the "snout" or "pout" reflex). This and the suck reflex (elicited by stroking the lips with a tongue depressor) are normally present only during infancy, but return in patients with bihemispheric or degenerative disorders.

Facial palsy is probably the commonest of the cranial nerve disorders. Sometimes this is due to injuries to the face or the parotid gland, but more commonly it is due to Bell's palsy. The latter often occurs with an upper respiratory tract illness, probably due to nerve compression in the facial canal after the greater superficial petrosal nerve comes off, since tearing is usually preserved. Infections involving the middle ear and temporal bone, and temporal bone fractures may cause facial palsy. Lesions in the cerebellopontine angle -- classically schwannoma of CN VIII (“acoustic neuroma”)-- may lead to deafness, vertigo and facial palsy. Such lesions, which are central to the point where the greater superficial petrosal nerve comes off, also cause loss of tearing. Lesions central to the point where the branch to tensor tympani is given off cause hyperacusis – sounds seem too loud. Bilateral peripheral facial palsy indicates an infectious or carcinomatous meningitic process, sarcoid, Lyme disease, or the Guillain-Barre syndrome. Within the brainstem, lesions in the pontine tegmentum affect the fascicles CN VII as they hook over the abducens nucleus; the result is a horizontal gaze palsy and ipsilateral facial palsy.

Unilateral facial weakness is a common feature of disease affecting the pyramidal tracts as they descend from cortex to brainstem, such as stroke. Such patients with "central facial weakness" show sparing of the frontalis muscles and relative sparing of the orbicularis oculi, but the muscles of the lower face are weak. Other descending pathways project bilaterally to all the facial muscles so that a dissociation between the voluntary and emotional facial movements may be observed. So, for example, patients with central facial weakness may be able to smile
symmetrically in response to a joke, but not when they mimic the examiner's smile.

**CN VIII**

The vestibulo-cochlear nerve mediates balance and hearing. The vestibular nerve conveys signals from the receptors (hair cells) in the semicircular canals, which sense head rotation, and the utricle and saccule, which sense linear motion (including gravity). The vestibular nerve synapses in the medulla in the vestibular nuclear complex. Although vestibular inputs do project to cerebral cortex, they are mainly important in generating two types of reflex. The vestibulo-ocular reflex moves the eyes so that the line of sight is not perturbed during body movements. The vestibulospinal reflexes make rapid changes in the tone of the muscles of the trunk and lower limbs to maintain equilibrium. An important physiological factor is that, even with the head stationary, there is a resting discharge in the primary vestibular afferents. When the head rotates, the discharge from afferents from one ear increases whereas the inputs from the other decrease. Thus, the brain looks for differences between the discharge of the vestibular afferents to detect head movements.

The organ of Corti consists of a spiral array of hair cells that lie on a basilar membrane covered by a tectorial membrane. Mechanical vibrations of the basilar membrane generate neural impulses. The cochlear nerve runs with the vestibular and facial nerves through the cerebellopontine angle to enter the brainstem at the lower border of the pons, dorsolateral to the inferior cerebellar peduncle, and reach the dorsal and ventral cochlear nuclei. Secondary auditory neurons project through the contralateral lateral lemniscus to the inferior colliculus, and then via the medial geniculate nucleus of the thalamus to terminate in the upper bank of the temporal lobe (primary auditory cortex).

The cardinal symptom of vestibular disease is vertigo -- the illusion of movement (usually spinning of self or the environment). The brain misinterprets a difference in the discharge from the two vestibular nerves as being due to rotation, while it is really due to a unilateral loss of function. Vertigo should be differentiated from other causes of "dizziness", including presyncope, postural unsteadiness, or psychological feelings such as panic. In patients who complain of vertigo at the time that they are evaluated, look for nystagmus and note its characteristics. Determine whether the nystagmus increases when visual fixation is prevented by transiently covering the viewing eye during ophthalmoscopy in an otherwise dark room; this sign implies a disturbance of the peripheral (rather than central) vestibular system.

Evaluate the vestibulo-ocular reflex with rapid head-turns; if eye movements are not promptly generated, a corrective saccadic movement is required to hold the eye on the target. The vestibulo-ocular reflex can also be tested by ophthalmoscopy, observing the optic nerve and retinal vessels as the patient shakes the head from side to side in small movements at about 2 cycles per second and fixates a distant, stationary target. If the patient wears glasses, look at the retina through the spectacle lenses. If the reflex is appropriate, eye and head movements are equal and opposite, and the retinal vessels and optic disc appear stationary. Patients with an impaired vestibulo-ocular reflex experience a decline in their visual acuity while they rotate their heads from side-to-side at about 2 cycles per second while they attempt to read from a Rosenbaum card. The contribution of each ear to the vestibulo-ocular reflex is best tested by caloric stimulation which can be performed at the bedside. First, check that the tympanic membrane is intact and that wax is absent, then elevate the patient's head 30 degrees on a pillow to place the lateral semicircular canal in a vertical position. A normal response -- nystagmus with quick phases directed away from the side of stimulation -- can be elicited with as little as
0.2ml of ice-cold water.

The vestibulospinal reflexes can be best evaluated by asking the patient to stand with feet together, arms flexed at the elbows and pulled taught against the fingers of each hand, which are hooked under each other against the chest. Three factors contribute to balance: vision, proprioception (largely at the ankle), and the vestibulo-spinal reactions. The effects of vision can be excluded by asking the patient to close the eyes. The effects of proprioception can be largely negated if the patient stands on a soft pillow. In this way, the vestibulo-spinal contributions to steady posture can be tested separately. Test for past-pointing of the limbs by asking the patient to bring the outstretched upper limb from an initial position above the head rapidly downwards so that the index finger meets the examiner's index finger. After a trial run with the eyes open, the test is performed with eyes closed.

Before testing hearing, check that the external auditory meatus is clear and the tympanic membrane is intact. In patients with no complaints, screen hearing by detection of finger rub, or test by using a 256 Hz tuning fork, comparing their threshold with yours. If there is hearing loss, compare air with bone conduction by alternately placing the vibrating tuning fork on the mastoid process (bone conduction) and holding it in front of the external auditory meatus (air condition); this is Rinne's test. In a normal individual, the tuning fork is heard louder and longer by air than by bone conduction. In conduction deafness (e.g., due to middle ear disease), bone conduction is better. In sensorineural deafness both air and bone conduction are reduced but air conduction is superior. Then place the vibrating tuning fork over the middle of the forehead and inquire whether the vibration is heard more in one ear; this is Weber's test. Normal Subjects cannot lateralize the vibration to either ear. In conductive deafness, the stimulus is heard better in the affected ear, but in sensorineural deafness, the stimulus is heard better in the normal ear.

Decreased hearing may be due to wax in the external ear, perforation of the tympanic membrane, and middle ear disease, such as effusion. These are all cause conductive hearing loss. Patients with a facial palsy may report hyperacusis if the nerve to the stapedius is involved. Sensorineural hearing loss occurs with either disease of the cochlea or of the cochlear nerve and its central connection ("retrocochlear"). Disorders of the cochlea such as Ménière's disease produce deafness that is greatest for lower frequencies, tinnitus, and the phenomenon of recruitment, in which loud sound are perceived as too loud, distorted and discomforting. Patients with cochlear disease show similar loss of acuity for pure tones and speech discrimination. Disease of the cochlear nerve and its central projections causes tinnitus and hearing loss that is more pronounced for higher tones. Loss of speech discrimination is greater than for pure tones. The cochlear nerve may be damaged with temporal bone fractures. Progressive, insidious, sensorineural hearing loss is a classic presentation of “acoustic neuroma” – schwannoma of CN VIII.

**CN IX**

The glossopharyngeal nerve supplies the tongue and pharynx. It is closely related to the vagus nerve (CN X) and contains motor, sensory and visceral fibers. The motoneurons of the glossopharyngeal nerve arise from the nucleus ambiguus, situated in the medulla. Its visceral efferent fibers arise from the inferior salivatory nucleus, which is just rostral to the dorsal motor nucleus of the vagus. Visceral afferents synapse in the nucleus of the solitary tract. The somatic sensory fibers synapse in the spinal trigeminal nucleus. The glossopharyngeal nerve leaves the medulla just dorsal to the inferior olive, exit the skull through the jugular foramen, and then arches down and forward, on the lateral wall of the pharynx, to reach the base of the tongue. The
motor fibers supply the stylopharyngeus muscle, which helps to elevate the larynx during swallowing. The visceral efferent fibers pass via the lesser superficial petrosal nerve to reach the otic ganglion; postganglionic fibers innervate the parotid gland and salivary glands in the posterior tongue. The visceral afferents mediate taste from the posterior one third of the tongue. Just below the skull, a branch leaves descends to the carotid bifurcation where it innervates the carotid sinus and body. The somatic afferent fibers supply sensation over the posterior tongue, palate and pharynx, where they anastomose with vagal fibers in the pharyngeal plexus.

The most reliable test of glossopharyngeal nerve function is taste over the posterior third of the tongue (see testing of taste under CN VII). Sensation in the posterior pharyngeal wall and the gag reflex probably depend mostly on the vagal contribution to the pharyngeal plexus. Similarly, isolated lesions of the glossopharyngeal nerve are unusual; more commonly the vagus nerve is also involved. Glossopharyngeal neuralgia causes a lancinating pain similar to tic douloureux, but located in the throat or tonsillar region, with radiation into the ear. Sometimes syncope or cardiac arrest has been associated.

**CN X**

The vagus nerve contains motor, visceral and sensory fibers. The motor fibers originate in the nucleus ambiguus in the medulla. The visceral efferent fibers originate in the dorsal nucleus of the vagus, which is located lateral to the hypoglossal nucleus under the fourth ventricle. The visceral sensory fibers terminate in the nucleus of the solitary tract. The somatic sensory fibers terminate in the spinal trigeminal nucleus. The vagus emerges from the medulla just dorsal to the inferior olive and exits the skull -- along with CN IX, CN XI, and the jugular vein -- through the jugular foramen. A pharyngeal branch supplies the pharyngeal constrictors and muscles of the palate, and carries visceral afferent fibers mediating taste on the posterior third of the tongue. The superior laryngeal nerve innervates the cricothyroid muscle and the mucous membranes of the larynx. The vagus descends in the neck in the carotid sheath, medial to the jugular vein. When the right vagus enters the thorax, it descends anterior to the subclavian artery close to the posterior aspect of the right main bronchus; it hooks under the subclavian artery, and ascends close to the thyroid gland to innervate all laryngeal muscles except for the cricothyroid. The left vagus descends through the thoracic aperture anterior to the subclavian artery and aortic arch to reach the left mainstem bronchus. The left recurrent laryngeal nerve arises from the left vagus at the level of the aortic arch and hooks under this vessel before ascending to the larynx. The vagus innervates the heart, lungs and esophagus; terminal branches pierce the diaphragm to innervate the stomach, intestines, kidneys, pancreas, spleen and liver.

Clinical testing of CN X largely depends on testing somatic motor functions. Observe the soft palate and uvula in the resting position; deviation of the uvula may be due to old tonsillar scarring. Pay attention to whether the palate elevates symmetrically when the patient says "aah". The palate will elevate less on the side of weakness, and the uvula will deviate towards the intact side. Bilateral palatal weakness causes difficulty in closing the posterior pharynx, causing a nasal quality to speech so that "k" becomes "ng". Test **pharyngeal sensation** over the posterior wall of the pharynx by touching each side with a cotton-tip applicator and asking the subject if the stimulus is perceived on each side. Note, however, that the glossopharyngeal nerve also contributes to pharyngeal sensation. The pharyngeal ("gag") reflex is induced by stimulating the posterior pharyngeal wall; its threshold varies idiosyncratically, but asymmetries of response are more reliable. It is often more useful to watch the patient swallow liquids. Test **laryngeal function** by ask the patient to speak (listening for hoarseness), to cough, and to imitate a high-
pitch "ee" sound (which requires apposition of the vocal cords). A simple bedside test of autonomic functions supplied by the vagus nerve is to monitor change in pulse rate during a Valsalva maneuver (forced expiration against a closed glotis). Slowing of the pulse occurs after the maneuver implies intact vagal cardiac innervation.

Because of its long course, the branches of the vagus nerve are susceptible to many disease processes. Brainstem strokes may cause dysphagia, and lead to aspiration pneumonia. Neck trauma tumor, and thyroid gland surgery may affect CN X and cause hoarseness. In patients with swallowing difficulties, consideration should be also given to neuromuscular disorders such as myasthenia gravis, botulism, and organophosphate poisoning.

**CN XI**

The accessory nerve arises from cranial and spinal roots. The cranial root originates in the nucleus ambiguus and, after joining the spinal root, soon leaves to join the vagus nerve. Thus, the cranial root can viewed as an aberrant portion of the vagus nerve. The accessory nerve proper depends on the spinal root, which emerges from the lateral funiculus of the spinal cord from the second to the sixth cervical segments and is purely motor. The spinal roots ascend through the vertebral canal to enter the cranial cavity through the foramen magnum and join the cranial root. The fused cranial and spinal roots leave the skull through the jugular foramen. After the cranial fibers leave to join the vagus nerve, the spinal accessory nerve descends behind the internal jugular vein to reach the sternocleidomastoid and trapezius muscles, which it innervates.

Note the size and symmetry of the sternocleidomastoid and trapezius muscles and look for any abnormal neck postures such as torticollis (wry-neck). Test the right sternocleidomastoid muscle by asking the patient to turn the chin forcefully to the left (horizontal head rotation) against your hand; observe and feel contraction of the muscle belly. Similarly, test the left sternocleidomastoid during rightward head turn. With bilateral involvement, the patient may have difficulty lifting the head off the pillow. Look for asymmetry of the trapezius muscles and shoulder droop. Test the trapezius by asking the patient to shrug and retract the shoulders against your resistance. The trapezius muscles can also be tested by asking the patient to extend the neck against resistance. With severe, bilateral trapezius weakness, the patient's head falls forward. Isolated lesion of the spinal accessory nerve are rarely occur when tumor or infection involves the cervical lymph nodes in the posterior triangle of the neck. When the CN XI is involved in along with the vagus and glossopharyngeal nerves, the location is usually at the jugular foramen, due to skull base tumor or trauma. Bilateral weakness of the sternocleidomastoid and trapezius muscles occurs in muscle disorders, such as myotonic dystrophy (in which there is atrophy of the sternocleidomastoid), polymyositis, and myasthenia gravis. Another disorder affecting the sternocleidomastoid is torticollis or wryneck, in which the head and occiput are pulled to the side of the affected muscle and the face is turned in the opposite direction. Persistent torticollis is a manifestation of hereditary dystonia.

**CN XII**

The hypoglossal nerve supplies the muscle of the tongue. Its nucleus lies in the medulla, just lateral to midline, under the floor of the fourth ventricle. Each hypoglossal nerve innervates the ipsilateral side of the tongue. The hypoglossal nerve emerge on the ventral surface of the medulla between the pyramid and the inferior olive, and leaves the skull through the anterior condyloid foramen (the hypoglossal canal) in the occipital bone. The nerve passes between the vagus and internal carotid artery towards the root of the tongue to innervate the genioglossus (intrinsic tongue muscle).
First, observe the tongue as it rests on the floor of the mouth. Look for a atrophy and fasciculations. Reduced volume and a wrinkled appearance to the mucosa indicate atrophy. Fasciculations are fine involuntary movements that are usually detected at the sides of the tongue, posteriorly. Bear in mind that normal subjects cannot hold their tongues perfectly still, but are able to prevent protrusion from the mouth and sustain protrusion. Ask the patient to protrude the tongue and look for any lateral deviation. In patients with unilateral facial weakness, it is important to use a reliable reference of the midline or else the tongue will be misinterpreted as deviating laterally. In such patients, lift the weak side of the face with one hand, and place the index finger of the other hand at the middle of the lower lip; then ask the patient to protrude the tongue. Test the mobility of the tongue by asking the patient to make rapid, side-to-side movements, illustrating by your own example. Test the strength of the tongue by asking the patient to push the tongue into each cheek in turn, feeling the force generated. The contribution of the tongue to speech can be evaluated by asking the patient to say "La-la-la-la" repetitively.

With a unilateral lesion of the hypoglossal nerve, the tongue shows hemiatrophy and, on attempted protrusion, deviates towards the affected side (because the intact genioglossus muscle pulls its half of the tongue forward). Unilateral paralysis of the tongue does not usually interfere with swallowing, but bilateral paralysis does. Fasciculation of the tongue suggest a nuclear process, usually amyotrophic lateral sclerosis (ALS). When impaired tongue movements are due bilateral pyramidal tract disease (pseudobulbar palsy), the tongue appears small and patients cannot make rapid side-to-side movements; they often also show “emotional incontinence.”. Tremors or involuntary movements of the tongue occur in parkinsonian, chorea and oral-facial dyskinesia (e.g., following neuroleptic drug administration).

**MOTOR EXAMINATION**

The motor system is encompasses motor cortex, pyramidal and extrapyramidal (basal ganglia) pathways, spinal cord, peripheral nerve, neuromuscular junction and muscle. Elucidating motor problems requires a systematic approach: muscle bulk, spontaneous (adventitial) movements, tone, power, and coordination.

**Screening tests of motor function**

Before starting the examinations of the limbs, it is worthwhile carrying out a few simple tests to determine if there is any asymmetry of motor function. These are especially helpful in patients who have reported an episode of weakness or clumsiness, but appear to have recovered. First, ask the patient to hold out their upper limbs in front of them, with their palms facing the ceiling. Having established that they can do this, ask them to close their eyes but hold their hands steady. Look for any downward drift, accompanied by pronation. Another useful screening test is to ask them to hold their arms, flexed at the elbows and rotate one forearm rapidly around the other. Look for any “orbiting” of one forearm around the other; the arm that moves less is usually the abnormal side. Third, ask the patient to tap one index finger against the interphalangeal joint of the adjacent thumb as quickly as possible. Some normal subjects show a slight asymmetry, being faster with the dominant side.

**Evaluation of gait and posture (station)**

Always attempt to test the patient’s ability to stand and walk, unless there is a compelling
reason not to (e.g. recent surgery). First, ask the patient to stand with their feet together, and arms by their sides. If they are able to do this, ask them to close their eyes (Romberg’s test), being ready to offer support should they become unsteady. Ask them to walk as quickly as possible in the hall. In sick patients, be ready to provide assistance. Note if they walk with a normal base (feet close together), or broadened (implying gait ataxia). Look for any asymmetry, such as stiffness of one lower extremity with the toes pointed down and circumduction – the spastic leg is brought around to the side to the front so that the toes does not catch on the ground. Bilateral spasticity causes a “scissors gait” with legs crossing in front. Look for the ataxic gait which occurs from imbalance. Note any excessive hip movement – a waddling gait. Also pay attention to arm swing; loss of such “associated movements”, especially asymmetrically, is encountered in Parkinson’s disease. Finally, in the patient who complains of falls but shows no tendency to broaden the base of their gait (feet still kept close together) and rocks from side to side, always saving themselves, consider psychological factors.

Next, if the patient seems able, encourage them to take a few paces on their toes and then on their heels. This supplements strength testing in the lower extremities. Ask them to walk a line, the heel of one foot placed in front of the toe of the other (tandem gait). Patients with gait ataxia are unable to do this. Ask the patient to turn and walk back the other way, since postural instability may become evident upon turning. Although ataxic gait is often taken to imply cerebellar disease, it can also occur following loss of proprioception or vestibular function. The loss of proprioception becomes more noticeable if visual input is eliminated. This is the basis of the Romberg test for proprioceptive ataxia. In this test, the patient is asked to stand with feet together and eyes open. The amount of sway is noted by the examiner. The patient’s eyes are then closed. The Romberg is positive if the sway increases markedly when the eyes are closed. The Romberg is not positive if instability is present both with eyes open and with eyes closed.

A sensory deficient gait occurs when patients can not sense the proprioceptive and tactile feedback from their feet and joints. In order to increase the tactile feedback, they may slap their foot hard against the ground. In classical neurology this gait was referred to as tabetic, since it occurred with destruction of sensory neurons from syphilis. As syphilis has become much more rare, other conditions such as diabetes, complications of medications, paraneoplastic syndromes and the many other causes of chronic neuropathies have replaced syphilis as a cause of a sensory deficit gait.

Gait apraxia is an interesting disturbance which occurs with injury to the frontal lobe and its projections. It is encountered in hydrocephalus, when medial frontal lobe fibers are stretched around the enlarged ventricles. Patient with apraxia appear to have “forgotten” how to walk, but may improve if they are given a series of marks on the floor on which to place their feet. Sometimes, if the examiner taps out a rhythm, the gait can be improved.

In parkinsonism, the initiation of movement may be difficult, and the patient is slow in getting started. Once started, however, it may be difficult for them to stop or change direction. Movements are not fluid, and balance is poor. This gait has been called shuffling, festinating, or “marche a petit pas”. Some patients retropulse if given a gentle push (always protect the patient from falling). Whenever an elderly patient complains of falling backwards, always consider the parkinsonian disorders.

Gait difficulties from muscle weakness are usually evident. Confusion may arise if the examiner forgets that anti-gravity muscles are much stronger than most other muscles, so weakness may not be apparent if the patient is tested in a bed or chair. Foot drop from tibialis anterior weakness, inability to walk on the toes from gastrocnemius and soleus weakness,
instability at the knee from quadriceps weakness or weakness of any of the glutei muscles (the strongest muscles in the body) will cause a recognizable pattern of abnormal gait. With upper motor neuron lesions patients usually show a combination of weakness and spasticity.

Astasia-abasia gait is, in its extreme, a vaudeville reeling gait side to side, with exaggerated tilts, twists and turns, but always managing to catch balance at the very last moment. It is actually a difficult act to perform, and requires considerable coordination. Astasia-abasia is a marker for a conversion reaction. It is sometimes called “a hysterical gait”, although the term hysterical is these days taken to be somewhat pejorative. The more common instances of astasia-abasia are not highly dramatic, but still can be recognized as conscience or sub conscience attempts to exaggerate imbalance. Always consider the possibility that patients who have unphysiological or exaggerated aspects to their examination do in fact have underline neurological disorders. They simply want to make sure the examiner notices the problem.

**Muscle Bulk**

Any patient complaining of weakness should be examined for evidence of muscle wasting. In the head, note any temporal hollowing. In the upper limbs, look for asymmetry of the shoulders or arms, and the intrinsic hand muscles: the first interosseus space, the thenar and hypothenar eminences. In the lower limbs, look especially for quadriceps wasting and prominence of the anterior tibial surface. Muscle mass varies considerably between different normal subjects; however, asymmetries of bulk are often significant. When in doubt, measure the circumference of the limbs at corresponding points (measured at similar points above or below the elbow or patella.

**Adventitial Movements**

Even during history-taking it may be apparent that the patient shows abnormal, involuntary movements. When the patient is undressed, ask them first to sit quietly and then lie at rest on the couch. Look for fine, spontaneous flickering movements of the muscles, especially in the tongue, around the shoulders, in the thighs, calves, and the interosseous muscles of the hands. Oblique lighting sometimes facilitates detection of fasciculations. Tapping muscles may also occasionally bring out fasciculations, but responses to such tapping are less specific as an abnormality than are truly spontaneous fasciculations. Such fasciculations are spontaneous contractions of motor units (a motoneuron plus the muscle fibers it supplies), and implies reinnervation of a denervated muscle, often in motoneuron disease (ALS). Fasciculation can also result from fatigue and metabolic factors such as low potassium or calcium, or from hyperthyroidism. Spontaneous, repetitive fasciculations, typically around the eyes over the course of a week or two are common in normal individuals, and do not indicate pathology.

Note any tremor (a repetitive movement of the extremities or head). A tremor present at rest implies Parkinson’s disease (often with a three per second pill rolling character at the fingers). A tremor while sustaining posture (hands and arms held outstretched) is usually essential. Some normal individuals have a slight tremor, accentuated by fatigue, which is called physiological tremor. Essential tremor may be familial; it gets worse with age. Thyroid disease, stimulant medications (e.g. for bronchospasm) and lithium commonly cause or accentuate postural tremor. Alcohol and beta- blocker medications reduce postural tremor. Essential tremor can also involve the head with either a side to side movement (in Parkinson’s disease, it tends to be vertical), and occasionally imparts a quavering quality to the voice. The student should be aware that different types of tremors may overlap; for example, elderly patients with essential
tremor may develop Parkinson’s disease. Chorea consists of fleeting, brief involuntary movements, while myoclonus is shock-like. Dystonia is an abnormal sustained posture of the head or limbs.

**Tone**

Tone may be defined as the resistance of a muscle to passive stretch. Encourage the patient to relax and stop voluntary movements or isometric tensing up of the muscles (which some patients find difficult). Tone changes can be mechanically or neurologically. Mechanical increases of tone are usually caused by contractures which make it difficult to move a muscle against the direction of the shortened tendon. Rarely infiltrated diseases of muscle may increase muscle tone itself. Neurological increases of tone are divided into three broad categories: spasticity, cog-wheel (or lead-pipe) rigidity, and paratonia (gegenhalten).

**Spastic tone** is characteristic of pyramidal tract lesions. Loss of input from upper motoneurons, stretch reflexes become uninhibited or hyperactive. A sudden passive movement of a limb, such as extension at the elbow, will initially be opposed (a “catch”) but then suddenly the muscle tone gives way. This catch and release phenomenon has been called a “clasp-knife” action, in analogy for example, to the way that a Switz Army knife is hard to close at the start but then after a short distance slides close easily. Spasticity is both velocity and length dependent. The more quickly the muscle is moved, the more apparent is the resistance. Similarly, the resistance is most obvious at the start of the movement when the muscle is shortened, and becomes less obvious as the movement progresses. Spasticity results from anything that separates the spinal reflex loops from the upper motor neuron. This could include spinal cord damage, demyelination of the descending motor axons, destructive lesions of cortex, cortical outflow white matter tracks or white matter in the brainstem by stroke, tumor, trauma, infection, congenital or developmental or degenerative conditions or other destructive processes. For reasons that are unclear, spasticity does not develop immediately but may be delayed a few hours to a few weeks after a destructive lesion. Prior to the development of spasticity muscle tone may actually be decreased. This should not mislead the student who has taken a careful history of the time course of the events.

Test for spasticity in the upper limbs by asking the patient to relax and rapidly extending the elbow. Test the lower limbs with the patient supine and relaxed; attempt to lift the leg off the bed by support it slightly rostral to the knee. With spasticity, it will be possible to lift the heel off the bed. Often, chronic spasticity coexist with contractures that make the clasp-knife aspect of spasticity difficult to discern. In such cases spasticity can be diagnosed by the company it keeps, such as clonus, hyperreflexia, and positive Babinski signs (see section of reflexes below)

**Cog-wheel or lead pipe rigidity** is an increase of tone characteristically accompanied by basal ganglia disease, such as Parkinson’s disease. Like a lead pipe or mechanical rachet, the resistance to passive motion is uniform across the range tested. Test for lead pipe rigidity at the wrists, holding the patient’s wrist with one of your hands and rotating the wrist with the other. First ask the patient to relax and then draw a circle with the index finger of the other hand; this “reinforcement” will often bring out the tone increase. Cog-wheel rigidity can often also be appreciated at the elbow, but less readily in the lower extremities.

**Paratonia**, also called gegenhalten, is increased tone from variable resistance to movement. It results essentially from poor patient cooperation. Individuals with defuse cerebral disturbances or frontal lobe disorders may be unable to relax their muscles, producing paratonia. It can be distinguished from spasticity or lead pipe rigidity by the variable nature of the increased tone and a tendency for it to be worse when the arm is moved faster or when the patient is
alarmed by a sudden examiner movement.

Hypotonia, occurs acutely following injury to the pyramidal tracts (as in cord transection) and chronically with injury to the lower motor neuron, neuromuscular junction or muscle. Cerebellar disease can also result in decreased tone. Hypotonia is often difficult to demonstrate, but an asymmetry of tone can be demonstrated by asking the patient to rest their elbows on a bedside table, with the wrists flexed; the arm with reduced tone shows greater flexion of the wrist.

COORDINATION

Although normal coordination requires intact muscle strength and normal proprioception, dexterity of all movements depends on the cerebellum. Test cerebellar function during the examination of eye movements (see above), looking for saccadic dysmetria, impaired ability to hold eccentric gaze (gaze-evoked nystagmus), and downbeat nystagmus. During testing of speech, ask the patient to repeat “kitty-kitty...” and sing a single note; observe any irregularity of the effort. In the limbs, observe coordination during finger-to-nose and heel-to-knee-to-shin testing. During rapid repetitive movements, such as tapping each side of the hand in turn against the thigh, note irregularity of the movement, which is called dysdiadochokinesia. During gait testing, observe whether the base is broadened, whether the patient can perform rapid turns without losing balance, and whether they can walk in tandem. A patient with severe truncal ataxia up in bed without aid.

SENSORY EXAM

An informative sensory examination requires an attentive patient and thoughtful examination. In patients with no sensory symptoms, it is essential only to screen sensation in the toes; vibration and joint position sense are most sensitive. In addition, all patients should be tested for sensory extinction with double-simultaneous stimulation to identify any neglect. In patients with sensory complaints, a systematic examination is called for. The single most useful maneuver is to ask a patient, presuming that they are competent to respond, whether they have numbness or unusual sensations anywhere over their head or body. If they do, they should outline these sensations. Sometimes even putting temporary pin marks on the borders can be helpful to guide the examination. Always note whether the patient is attentive to testing; be prepared to take a break if they are tired.

Modalities tested in the screening neurological examination include pain and temperature, joint position sense, vibration, discriminative sensation such as graphesthesia, and occasionally other specialized testing. Pain and temperature is carried in the lateral and anterior spinothalamic tracks. It may be assayed by pin sensation (asking whether sharp or dull), or temperature sensation (warm or cool). Remember not to reuse a pin on different patients, in order to avoid the potential of spreading disease, and there is never need to draw blood in the testing of pain. Only the recognition of sharp sensation gives information to the examiner, since sharp will be perceived as dull and dull will be perceived as dull in most areas or relative anesthesia. Therefore, the examiner should make most of the test sensations with a sharp point, and only throw in a few dull sensations as control stimuli. Deep pain can be ascertained by squeezing a tendon, but this is not usually done in the alert patient. It may be useful in comatose patients. Temperature sensation can most easily be assayed by holding the usually cool tuning fork up against the skin of the distal extremities and asking if it feels cool. Decrease of
cool sense or of pain sense should be drawn on a sketch or described in terms clear enough to allow a reader to transfer the words to a drawing of his or her own. A distribution of numbness in a stocking-glove pattern suggests a diffuse polyneuropathy, such as might occur with some forms of diabetes mellitus. If the patient is numb in the little finger and half of the adjacent ring finger and the corresponding part of the palm of the hand, then an ulnar neuropathy should be considered. Numbness of the thumb, index, and middle finger will suggest a median neuropathy. Radial nerve lesions caused by compression over the back of the arm (“Saturday night palsy”) cause wrist-drop accompanied by a small area of numbness over the first interosseus space. Numbness on the anterior and lateral thigh may indicate a problem with the lateral cutaneous branch of the femoral nerve. Patterns of numbness in a dermatome are also of importance.

Hemibody numbness below a certain level, for example mid thoracic or cervical, would suggest spinal cord lesion. Numbness involving the face cranial nerve problems, and numbness involving face and body on one side would point to problems in the somatosensory system of the brain, including thalamus, hemispheric white matter or parietal cortex on the side contralateral to the numbness. Position and vibration sense travel together in the dorsal column of the spinal cord through the brainstem, to cross at the medial lemniscus, to ascend to ventral posterior thalamus, and from there to parietal cortex. Test vibration sense by applying a large (128Hz) tuning fork against the toenail or the bone of the large toe on each side. Use yourself (or the patient’s hand sensation) as a control before applying the tuning fork to the toes. If vibration cannot be perceived at the toes, the examiner should test vibration at the malleolus of the ankle, and then the knee, and then the iliac bone at the hip. If there are symptoms referable to the arms or hands, or vibrations senses abnormal in the feet, then testing vibration on the bones of the fingers or wrist should be performed. Vibration testing on the forehead or on the sternum is useful in suspected cases of functional (hysterical) numbness. A sharp division of vibration perception at the midline of these bones is unphysiological, since the vibration carries through the bone for a considerable distance.

Position sense may be impaired with peripheral nerve injuries, rude or posterior spinal cord processes, brainstem, white matter or parietal cortical diseases. People should be able to feel fine movements up and down of the toe. The toe should be held at the side when moving it in order to eliminate cues from pressure on the top or bottom of the toe. If position sense is absent in the toes, then it should be assayed in the ankles and knees. Position sense may also be tested by moving the distal knuckle of the finger joints up and down. The normal individual feels any visible movement. An indirect measure of position sense is the finger to nose test with the eyes closed and the attempt to maintain palms upward without drift, with the eyes closed. Patients with position sense abnormality may find their finger missing their nose, but with no difficulty powering the finger to the nose, or may find their hand drifting out in to space. Complete anesthesia to any form of touch is a rarity in the neurological examination, but sometimes does occur with complete nerve or root lesions. These areas should be assayed with a light touch with a wisp of cotton, heavy touch with a finger as well as pin sensation. Be careful to not overlook a spinal sensory level with anesthesia from one point on the body downward. This may be an indication of a serious condition such as spinal cord compression by metastatic tumor. There should be corresponding motor and sphincter signs.

A dysesthesia is an unpleasant sensation in a region. A paresthesia is an abnormal sensation, though not necessarily unpleasant. Hyperesthesia is a rare entity, denoting increased degree of a normal sensation. Hypesthesia indicates partial reduction of a sensory modality and anesthesia absence of the sensory modality, not to be confused with surgical anesthesia which
can mean loss of consciousness as well as loss of pain sensation. Sensitivity of skin from irritation such as sunburn, a partial peripheral nerve injury or a central process, usually at the level of the thalamus can all cause dysesthesia. Pain clinics invest a great deal of effort in diagnosis and therapy of syndromes of dysesthesia. The student should refer to textbooks for discussion of causalgia, or reflex sympathetic dystrophy.

**REFLEXES**

Deep tendon reflexes (muscle stretch reflexes) are among the most useful of neurological signs, although they must be interpreted in the context of the whole history and examination. A tap on the tendon actually serves to stretch the muscle. The muscle contains fibers called intrafusal fibers, which provide a reflex loop signal via the spinal cord when they are stretched. The result of the reflex is contraction of the stretched muscle. This quick reflex loop is useful in real life for stabilizing a muscle position in space. A quick unexpected movement of the joint, will stretch the muscle, leading to an immediate contraction and restitution of muscle length and joint position. A special type of intrafusal muscle fiber is called a gamma fiber. The gamma fiber resets the bias on the reflex loop to make it more or less active, on the basis of instruction received from involuntary cortical centers.

Traditionally tested muscle stretch reflexes include the biceps, triceps, brachial radialis, patellar and Achilles. Others, such as finger flexor reflexes, pectoral reflexes, hamstring, jaw jerk, may be tested in special circumstances. To test the reflex, the muscle is usually put in mid position with the involved joint partly flexed and the left and right side limbs positioned symmetrical. A soft brisk tap is then given to the examiner’s finger, while pressing some tension onto the appropriate tendon. It should not usually be necessary to hit hard. Reflexes can be assessed as easily by the threshold of tapping force required to illicit them as the amount of joint movement from a hard tap. The briskness of the reflex is recorded on a subjective 4 point scale 1-4. Tradition involved writing of + marks next to a stick figure to indicate the degree of reflexes and a “2” reflex therefore came to be known as 2+. In this context (unlike the MRC strength grading scale) the 2+ has no significance of being more than 2. A normal reflex is graded 2 in the midpoint of the scale (again unlike the MRC grading system for muscle strength, in which the highest grade of 5 is normal). A present but decreased reflex is graded 1, an absent reflex 0, a more than usually active but normal reflex 3 and an abnormally brisk reflex 4. Neurologist often debate what makes a reflex 4, since some normal individuals simply have very brisk reflexes. If the reflex spreads to other muscle groups and activates their contraction as well, then it is abnormally brisk. If it produces clonus which is repetitive contraction of the muscle to a single tap, then the reflex is considered 4+. A 4+ reflex may also be one which simply produces a very pronounced movement to a very slight tap. Such reflexes might not require a reflex hammer at all, but only a light tap of the examiner’s finger on the tendon.

In a written note, the stick figure method is a good method for recording reflexes, with either pluses or numbers written next to biceps, triceps, wrists (for brachial radialis) knee and ankle. If reflexes are 0 then the examiner should attempt to potentiate the reflexes with a maneuver named after Dr. Jendrasic. The examiner asks the patient to partially flex the fingers of each hand, invert one and hook his own fingers. On the count of three the patient pulls one hand against the other and at the same time the examiner taps the knees or ankles. Muscular efforts impart a general tone to all of the muscles, which may bring out a reflex. The upper extremities can be tested by having the patient tighten the legs or the jaw.
Special reflexes are usually recorded in the reflex section. The most important of these is the Babinski reflex. A purist reports the sign of Dr. Babinski to be either present or absent, rather than positive or negative: however, purist are becoming rare. To test the Babinski, or planter reflex as it is also known, the examiner holds the foot so that the sole is accessible, and runs a medium sharp object such as a key, along the lateral under surface of the foot, coming up towards the toes and then crossing at right ankles towards the base of the big toe. It is important to start at the lateral edge rather than the medial edge, in order to maximally involve the S1 dermatome. The normal response is for the great toe to flex down. An absence of response can also be considered normal if it is bilaterally symmetric. The abnormal response is an upgoing toe. A biphasic movement with toe up and then down is also abnormal. The full Babinski response consists of an initially upgoing toe, a fanning of the toes and a seemingly involuntary withdrawal of the leg with tensing of the quadriceps muscle. If the Babinski is absent a number of other named maneuvers can be performed, such as applying sliding pressure to the sheen while testing the planter response (Oppenheim), squeezing the calf (Gordon or Chaddock-check this), or repeatedly applying a pin to the dorsal surface of the toe (Bing). These accessory maneuvers are only marginally useful when the planter response is equivocal. Presence of the Babinski sign (positive Babinski and common parlance) suggests an upper motor neuron lesion. The practical issue in testing this reflex involves judgement about whether the patient is simply withdrawing because of ticklishness. Whether withdrawal is voluntary or involuntary can best be learned by experience and testing, but sometimes remains unclear even to the expert examiner.

Frontal lobe “release” signs are tested in patients who exhibit decrease mental status. They are usually not tested in apparently cognitively normal individuals. They are, in fact, more a sign of diffuse cerebral disturbance. The grasp reflex involves the patient gripping the examiner’s hand as it brushes their palm. A grip once or twice may be taken semivoluntary, but a forced grasp, by which the patient can not inhibit grasping the hand even when told to do so, is clearly abnormal. Tapping on the mouth may produce a puckering of the lips called a snout reflex. A rooting reflex is elicited by lightly scratching the mouth, and finding the patient attempting to reach over to align the finger with their mouth. These are primitive reflexes which are useful in hungry babies, but are generally suppressed in normal adults. The glabellar reflex is elicited by tapping the midpoint of the forehead, usually from above the head so as not to present a visual threat. The normal response is not to blink or to blink once or twice and then stop blinking. The abnormal response is to blink repeatedly every time the forehead just above the bridge of the nose is tapped. The palmomental reflex consists of twitching of the muscles of the lower jaw ipsilateral to a hand which has been briskly scrapped on the thenar eminence. All of these reflexes reflect disinhibition. Abnormal reflexes are most diagnostically most useful when they are combined with other abnormal findings.