NSI HANDBOOK OF CLINICAL EXAMINATION

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EXAMINATION OF HIGHER MENTAL FUNCTIONS

Dr. P. Sarat Chandra, Dr. Manjari Tripathi

"*This chapter is dedicated to all neurosurgery residents, for most of whom, examination of higher mental functions is limited to just saying that the "patient is conscious, alert and oriented."*

"Also to all Neurology residents, who may know more but still fall short of practicing this important component of Neurology."

Definition:

The examination of higher mental status involves a structured assessment of the patient's behavioral and cognitive functioning. It provides an objective insight to the examiner on the patient's level of cognitive functioning, his/her ability to interact with environment and adjust to the existing social structures. It is not a *single* function, but a constellation of multiple capabilities localized to various parts of the brain. Some functions have a definite localization while other functions cannot be localized to any single area of the brain and occur as a result of interaction of multiple neuronal circuits.

Historical Background:

Historically, there has been a paradigm shift in our understanding of localization of various cognitive functions. In the medieval times, people could not fully grasp the importance of the brain. Ancient Egyptians thought that the cranium contained "lymph". They removed it prior to mummification from a hole made through the nose. Even though ancient Greek scholars understood the basic functions of the brain, it was only in the 19^{th} century, when Paul Broca discovered the "expressive language" area that the scientists began to understand how higher mental functions such as language could be localized in the brain. Paul Broca, Carl Wernicke, and Hughlings Jackson developed different models of brain function, and each contributed important insights to the study of aphasia. Broca's contributions were influenced by the fundamental question of whether higher mental function could be localized in the

brain at all; Wernicke's contributions were influenced by an attempt to unite more mechanistic and physiological principles to a model of higher brain functions; and Jackson's contributions were influenced by British association psychology.

Norman Geschwind (1926-84) made an important contribution towards understanding of dyslexias, the neuroanatomy of cerebral lateral asymmetries, and other areas of neurological dysfunction. He is especially known to have provided an anatomical insight towards disconnection syndromes.

Scope Of The Chapter And Importance Of Clinical Examination Of Higher Mental Functions:

The current scope of the chapter is to provide "practical guidelines" for clinical examination at bed-side for residents using simple yet objective methods.

While examination of higher mental functions (HMF) currently is the field of clinical psychologists, it is still important for the residents to be aware of this aspect of examination for the reasons given below

1. HMF, pre-operatively, prior to cranial surgery provides baseline information for the cognitive status. While the clinical psychologists provide a very extensive examination, an examination by the operating surgeon will provide him/her an idea of the patient's cognitive status before and after the surgery
2. HMF examination provides an idea to the examiner for the areas of the cognition that need re-training. For e.g, in a patient with aphasia, post operative speech therapy would be more useful.
3. Most of the formal examinations e.g aphasia batteries are quite extensive. Hence a brief clinical examination is many times more useful providing the examiner a better idea of the patient's cognitive status.

Method of Examination:

There have been many methods described in various textbooks. About 2 decades earlier, the emphasis was to localize every cognitive function. Today, it is understood that only some functions may be localized adequately. Many others do not have a focal anatomical substrate or have a diffuse involvement. For the sake of simplicity the authors have divided the cognitive function examination into 2 parts. The first part has 7 parts (Basic examination) and the next part has 3 parts (Advanced examination).

In his Treatise on Insanity, published in 1801, Pinel, one of the fathers of modern

psychiatry, gave some advice to his contemporary colleagues.

"*To seize the true character of mental derangement in a given case, and to pronounce an infallible prognosis of the event, is often a task of particular delicacy, and requires the united exertion of great discernment, of extensive knowledge and of incorruptible integrity*".

This cannot be truer even today, as higher mental functions are a highly subjective examination. Hence, this chapter is only an attempt to provide practical working guidelines and invoke further interest in the intelligent resident to read more about it.

Table 1:

FIRST 7 QUESTIONS	**PART I: BASIC**
1.	Appearance, Grooming and Educational Status
2.	Handedness
3.	Mood, Affect, Emotions
4.	Conscious, alert, oriented to time, place and persons
5.	Attention and Vigilance
6.	Memory (immediate, recent, remote) & Language
7.	Insight and Judgment
NEXT 3 QUESTIONS	**PART II: ADVANCED**
8.	Specific lobar function
9.	General fund of knowledge
10.	Specific conditions: apraxias, alexias, agraphia, autotopogosia
ALSO INCLUDED	
11.	Mini mental score examination (MMSE)

PART I: Basic examination

1. **Appearance, Grooming and educational status:**
    - **a.** Is the patient well kept? Yes/No
    - **b.** Is he/she dressed appropriately? Yes/No
    - **c.** Is he/she well groomed and clean? Yes/No
    - **d.** Is he/she dressed as per his/her socio-economic status? Yes/No
    - **e.** Posture: Straight/ stooping
    - **f.** Eye contact: present/ avoids
    - **g.** Mention the educational status of the patient
        - **i.** School only/graduate/post-graduate
        - **ii.** Salaried/ self employed
        - **iii.** English speaking also/ local language only
    - **h. Pearls**:
        - i. These variables give the examiner an overall impression of the patient.
        - ii. Certain specific syndromes such as unilateral spatial neglect and the disinhibited behavior of the frontal lobe syndrome are readily appreciated through observation of behavior.
2. **Handedness**: (Luria's tests)
    - **a.** Writing**:**writes with right/left hand
    - **b.** Pin-hole: sees through the right or left eye
    - **c.** Kicking: kicks a ball with right or left leg
    - **d.** Pearls: In right handed individuals, the left hemisphere is dominant in 90% of cases, while in left handed individuals, the right hemisphere is dominant in only 40% of cases.
3. **Mood and Affect:**
    - a. Affect: is the patient's immediate expression of emotion. It is also described as the mood of the patient as perceived by the examiner.
    - b. Mood**:** Refers to the more sustained emotional makeup of the patient's personality.
    - c. Pearls**:**
        - i. Affect is inappropriate when there is no correlation between what the patient is experiencing or describing and the emotion he is showing at the same time (e.g., laughing when relating the recent death of a loved one).

ii. Both affect and mood can be described as dysphoric (depression, anxiety, guilt), euthymic (normal), or euphoric (implying a pathologically elevated sense of well-being).

iii. Affect must be judged in the context of the setting and those observations that have gone before. For example, the startled-looking patient with eyes wide open and perspiration beading out on the forehead is soon recognized as someone suffering from a brain tumor.

4. Conscious, alert, oriented to time/ place and person

a. Is conscious? Yes/No

b. Looks alert? Yes/No

c. Oriented to

i. Time? Ask the time

ii. Place? Ask the name of city and the place where the patient is at ?

iii. Person? Ask relationship between at 2 persons, one known other unknown? E.g Doctor (he/she may say the generic word like doctor)

iv. Pearls**:**For the level of consciousness, the scheme by Plum and Posner (1980) is well accepted, even though GCS score is in use widely

1. A normal level of consciousness is one in which the patient is able to respond to stimuli at the same lower level of strength as most people who are functioning without neurologic abnormality.
2. Clouded consciousness is a state of reduced awareness whose main deficit is one of inattention. Stimuli may be perceived at a conscious level but are easily ignored or misinterpreted.
3. Delirium is an acute or subacute (hours to days) onset of a grossly abnormal mental state often exhibiting fluctuating consciousness, disorientation, heightened irritability, and hallucinations. It is often associated with

toxic, infectious, or metabolic disorders of the central nervous system.

4. Obtundation refers to moderate reduction in the patient's level of awareness such that stimuli of mild to moderate intensity fail to arouse; when arousal does occur, the patient is slow to respond.
5. Stupor may be defined as unresponsiveness to all but the most vigorous of stimuli. The patient quickly drifts back into a deep sleep-like state on cessation of the stimulation.
6. Coma is unarousable unresponsiveness. The most vigorous of noxious stimuli may or may not elicit reflex motor responses.

5. Attention and Vigilance

a. Attention: (same as immediate memory): It is defined as an ability of a patient to repeat a task immediately after being asked to do the same. It is tested by forward and reverse digit span.

i. Forward digit span:

1. Ask the patient to repeat non-sequential numbers (e.g not even numbers or not in any known pattern)
2. Start with 1 digit and after the patient successfully repeats it, is asked to repeat increasing number of digits
3. Repetition upto 5 digits is considered as normal

ii. Reverse digit span:

1. Ask the patient to repeat non-sequential numbers (e.g not even numbers or not in any known pattern)
2. Start with 1 digit and after the patient successfully repeats it, is asked to repeat increasing number of digits *in a reverse manner*.
3. Repetition upto 3 digits is considered as normal

b. Vigilance**:** is defined as a sustained attention over a period of time. It is tested by "tapping tested"

i. Patient is provided non serial letters e.g "N..K..L..N..O..R..P..I..U..N..Y..N

ii. The patient should tap on the table with a pencil whenever he/she hears a specific letter e.g N

iii. It is better to write down a set of letters and then spell them out to avoid pauses by the examiner

iv. Pearls:

1. Patients may perform "commission" or "omission" errors i.e tapping too many times or may not tap at all
2. Patients with frontal lobe dysfunction may keep on tapping due to perservation.

6. Memory and Language:

a. Memory**:** This may be tested as immediate, recent and remote memory

i. Immediate memory**:** Same as attention

ii. Recent memory**:** This can be visual or verbal memory

1. Visual memory (non-dominant temporal lobe)**:**

a. Show the patient 4 unrelated objects. e.g: pen, key chain, coin, purse.

b. Hide each of these items in a *nearby different* place e.g. under the mattress, in the shelf etc.

c. Before hiding each item, the patient has to be clearly shown where the item is being hidden and he/she *should repeat the same after this has been hidden to ensure that immediate memory registration has taken place.*

d. The examiner should then continue with the examination and after 15 minutes, ask the patient to remember *all the items* and *where they are hidden.*

e. The findings should be noted as given in the example "The patient was able to remember 2 objects and was able to retrieve them from the respective hiding places. He was not able to remember the 3rd object (name the object) but was able to identify the hiding place. He was

neither able to remember the 4th object nor its hiding place".

2. Verbal memory (dominant temporal):
    - **a.** Tell the patient names of 4 unrelated things e.g. suresh (one proper noun), car (one noun), red (one color) and anger (one abstract).
    - **b.** The patient *should repeat the same after this has been told to him to ensure registration of immediate memory.*
    - **c.** The examiner should then continue with the examination and after 15 minutes, ask the patient to remember *all the words*.
    - **d.** The findings should be noted as given in the example "The patient was able to remember 2 words. He was not able to remember the 3rd word but was able to describe it (e.g. name of a color). He was not able to remember the 4th word.

iii. Remote memory**:**This may include

1. Names of family members
2. Name of the home town
3. Names of colleagues at work

b. Language**:** This is tested under the following headings.

i. Spontaneous speech (severely impaired in Broca's)**:**

1. Ask the patient to describe for about 2-3 minutes all about his day's schedule
2. Avoid asking questions like "what is your name?"
3. Examine:
    a. Pronunciation: Normal/abnormal
    b. Word/sentence formation: Normal/abnormal
    c. Fluency (volume of speech output, normal: 100-115 words/min; phrase length <7): Normal/abnormal.
    d. Prosody: Emotional intonation

4. Look for**:**
    - **a.** Paraphasias: "word replacement"
        - **i.** Phonemic: e.g. says "bow" instead of "cow"
        - **ii.** Semantic: e.g. saying "horse" instead of "cow"
    - **b.** Word finding difficulty
    - **c.** Neologisms: totally new and meaningless words

ii. Comprehension(severely impaired in Wernicke's)**:**

1. Pointing commands: Single step commands to point at named objects e.g. fan, door etc
2. Yes/No questions:
    - **a.** E.g**.** is your name Rajesh (false name),is your name (real name)?
    - *b. Ask at least 5 questions*
3. Simple commands:
    - *a.* E.*g.* Close your eyes, raise your hands
    - *b.* Touch your right ear with your hand

iii. Repetition(severely impaired in Conduction and preserved or overactive [e.g. echolalia] in trans cortical aphasia):

1. Start with simple words and proceed to more complex commands

iv. Reading:

1. Following written command
2. Read aloud

v. Writing: Writing to dictation

1. Writing spontaneous sample

vi. Pearls*:* See table 2

Table 2: Summary of various aphasias

Type of Aphasia	**Etiology**	**Spontaneous speech**	**Paraphasias**	**Comprehension**	**Repetition**	**Naming**

Broca's	Lesion in the left postero-inferior convolution, area 44	Non-fluent	-	Good	Poor	Poor
Global	Diffuse pathology e.g large stroke	Non-fluent	-	Poor	Poor	Poor
Transcortical motor	Water shed infarct-anterior	Non-fluent	-	Good	Good	Poor
Wernicke's Aphasia	Lesion in the left postero-superior temporal gyrus, area 22	Fluent	+	Poor	Poor	Poor
Trans-cortical sensory	Water shed infarct-posterior	Fluent	+	Poor	Good, sometimes "echolalia"	Poor
Conduction	Lesion of the arcuate fasciculus connecting Wernicke's and Broca's	Fluent	+	Good	Poor	Poor
Anomic	Left temporal, no known exactly	Fluent	+	Good	Good	Poor

7. **Insight and Judgment:**
    - **a.** Does the patient have insight about his/her disease? Yes/No
    - **b.** Real Life question: E.g. What would the patient do if he/she sees a house on fire?

PART II: Advanced examination

8. **Specific lobar functions:** A brief description of certain cognitive dysfunction arising due to lobar pathologies are described here:
    a. Frontal lobe
        - **i.** Cognition:
            1. Loss of inhibition
            2. Social incontinence
            3. Apathy
            4. Loss of Judgment
            5. Lack of planning
        - ii. Perservation: Is the lack of ability to perform alternating motor or visual tasks or inability to detect a change in the pattern of motor or visual tasks
            1. Motor: Ask the patient to show fist, ring, and palm alternatively. The patient will usually repeatedly perform a single task
            2. Visual: Ask the patient to draw the following pattern below: 1 semi-circle, 1 triangle, 1semi-circle, 2 triangles, 1 semi-circle, 3 triangles. A patient with frontal lobe dysfunction will fail to notice the pattern

iii. Others: These have been described in the respective sections and include

1. Motor/Broca's aphasia
2. Primitive reflexes: Snout, glabellar tap, and mentalis
3. Tip: The following is sentence has been taken from Dejong's textbook for Clinical neurological examination and very aptly describes a patient with frontal lobe dysfunction with a frontal lobe meningioma: "*He invested and lost his life's savings in an ill-advised business venture. He was fired from a succession of jobs because of tardiness and disorganization. His wife divorced him, and, unemployed, he moved back in with his parents. He required two hours to prepare for work each morning. He took a job 100 miles from his home but was fired for lack of punctuality. He spent entire days shaving and washing his hair. Minor decisions were scrutinized ad infinitum, including simple purchases and deciding where to eat. He collected outdated and useless items including dead houseplants, old phone books, six broken fans, five broken television sets, three bags of empty orange juice cans, 15 cigarette lighters, and countless stacks of old newspapers"*.

b. Temporal Lobe**:**

i. Bilateral: KluverBucy"like" syndrome (originally described in monkeys): Rare but may occur in herpes encephalitis due to bilateral temporal pole involvement. It generally consists of:

1. Amnesia. Characterized by an inability to recall memories. Its nature is both anterograde and retrograde, meaning new memories cannot be formed and old

memories cannot be recalled. The level of amnesia is considered to be profound.

2. Docility. Characterized by exhibiting diminished fear responses or reacting with unusually low aggression. This has also been termed "placidity" or "tameness".
3. Dietary changes and/or Hyperphagia. Characterized by eating inappropriate objects (pica) and/or overeating.
4. Hyperorality. "an oral tendency, or compulsion to examine objects by mouth".
5. Hypersexuality
6. Visual agnosia. Characterized by an inability to recognize familiar objects or people.

ii. Others: already described, include Wernike's aphasia, loss of recent memory (both visual for right temporal and verbal for left temporal lobe)

c. Parietal lobe

i. Right (non-dominant) parietal lobe:

1. Hemi-neglect
    a. Ignores the left side of the body
    b. May even say that the left side belongs to someone else or his "brother"
2. Geographical disorientation:
    a. Cannot find his way to familiar places e.g. toilet, home etc
    b. Cannot mark locations on maps
3. Dressing apraxia: "Cannot put on the shirt if one of the sleeves is pulled out"

ii. Left parietal lobe:

1. Gerstman's syndrome:
    a. Dysgraphia**:** Inability to write
    b. Dyscalculia**:** Inability to calculate
    c. Finger agnosia**:** Cannot name fingers
    d. Right-left disorientation**:** "Touch the right ear with the left hand"

2. Autopognosia**:** *agnosis, meaning "without knowledge", and topos, meaning "place", autotopagnosia virtually translates to the "lack of knowledge about one's own space…,"*
    a. Cannot orient or localize body parts
    b. E.g. when asked to point to elbow, may point to the knee
    c. It is important to rule out aphasia before doing this testing

iii. Occipital lobe:

1. Anton's syndrome**:** (denial of blindness)
    a. A form of visual agnosia, patient is blind but is unaware of this
    b. Due to lesion over the bilateral primary visual cortex and the surrounding areas
2. Balint's syndrome:
    a. Occurs due to bilateral lesions over the parieto-occipital area
    b. Characterized by
        i. Simultognosia: or "functional tunnel vision". The patient can only see one object and not the entire picture. This occurs due to loss of ability of the brain to "compose and reconstruct" the entire picture.
        ii. Optic ataxia: Ataxia which occurs when eyes are kept open but disappears on closing the eyes
        iii. Ocular apraxia: cannot move eye ball on command but can move it involuntarily

9. General fund of knowledge:

a. These may include various question pertaining to general knowledge

b. For illiterate persons/farmers especially in Indian setting, more relevant questions may be "when was the last major monsoon?", "when are the crops sown in winter?"

10. **Apraxias, Alexias, Agraphia, Agnosias, and Disconnection syndromes**: Many have been already discussed but will mentioned again for the sake of better comprehension of the subject:
    - **a.** Apraxia: Is the inability to perform a coordinated motor task in absence of motor/sensory and cerebellar symptoms
        - i. Ideomotor apraxia (Single task)
            1. Usually due to damage to contralateral pre motor cortex
            2. Oro-buccal**:** Cannot show tongue or blow cheeks on command but can do it involuntarily
            3. Limb kinetic**:** Cannot move limbs to command but can do it involuntarily
        - ii. Ideational apraxia (Multiple task)
            1. Occurs due to diffuse brain damage
            2. Cannot perform multiple task commands even though may obey single task commands
            3. A simple way to examine this at bed side is to
                - **a.** "dismantle a ball point pen and ask the patient to put it back"
                - **b.** "write a letter, fold it and put it in envelope"
        - **iii.** Other apraxias**:**
            1. Dressing apraxia
            2. Ocular apraxia
            3. Construction apraxia: Ask the patient to draw a clock with number
    - b. Alexias and agraphias:
        - i. Alexia: inability to read
        - ii. Agraphia: inability to write
        - iii. Alexia without agraphia: a disconnection syndrome due to a infarct involving the left occipital lobe involving the splenium (**Fig 1**)
    - c. Agnosia: "Lack of knowledge or insight"

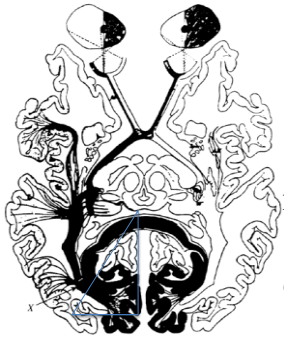
i. Tactile or astereoagnosis: due to parietal sensory cortex involvement
ii. Visual agnosia: Also called Anton's syndrome
iii. Finger agnosia: part of Gerstman's syndrome
iv. Autotopognosia: see above

d. Disconnection syndromes: These could be inter-hemispheric or intra hemispheric
    i. Inter-hemispheric:
        1. Usually due to acute corpus callosal sectioning e.g infarct. Some attribute this to cingulate gyrus involvement
        2. Also called "alien hand"
        3. Here the left hand is autonomous as the right hemisphere is now disconnected from the left hemisphere and it cannot express itself as it does not have any language function.
    ii. Intra-hemispheric:
        1. Alexia without agraphia
        2. Conduction aphasia
        3. Trans-cortical aphasia

11. **Mini Mental State examination (MMSE) (Fig 2)**
    a. Originally devised by Folstein (1975) for psychiatric residents
    b. Called "mini" as it does not test mood or thought disorders
    c. Was never meant for diagnosis of dementia but has become the most common screening test
    d. Its sensitivity is 86% and specificity is 92% and has a test-retest reliability of 0.86 (1 year, $p<0.001$).
    e. This has also become a very popular for patients who require a cognitive testing on follow up or testing even on phone

Fig 1:

Agraphia (inability to write) and alexia (inability to read)



left

right

Visual word system

Left occipital infarct: interruption of fibres from the right occipital lobe: **alexia without agraphia (pure word blindness)**

Fig 2:

Mini-Mental State Examination

Question	Score for each correct answer
Orientation	
What is the year, season, date, month, day	1 point
Where are we (country, county/district, town, hospital/building, floor/room)?	1 point
Registration	
Name 3 objects, taking 1 second to say each. Then ask the patient to repeat them. Repeat this until the patient remembers all 3.	1 point
Attention and calculation	
Serial sevens - subtract 7 from 100 and 7 from the result, etc (93, 86, 79, 72, 65).1 point for each correct answer. Alternatively ask the patient to spell "world" backwards.	Stop after 5 answers
Recall	
Ask for the name of the 3 objects in Question 3.	1 point
Language	
Point to a pencil and a watch. Have the patient name them for you.	1 point
Have the patient repeat "No ifs, ands or buts".	1 point
Have the patient follow a 3-stage command: "Take the paper in your right hand, fold the paper in half, put the paper on the floor."	3 points
Have the patient read and obey the following: "Close your eyes" (this should be written in large letters).	1 point
Have the patient write a sentance of his or her own choice (it must contain a subject and an object and make some sense). Ignore spelling errors.	1 point
Enlarge the design below to 1-5 cm per side and have the patient copy it.  *Maximum score 30 points*	Give 1 point if all the sides and angles are present and if the intersecting sides form a quadrangle

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EXAMINATION OF I & XII CRANIAL NERVES

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OLFACTORY NERVE

ANATOMY

- The olfactory membrane lies in the superior part of each nostril. Medially, the olfactory membrane folds downward along the surface of the superior septum laterally, it folds over the superior turbinate and upper surface of the middle turbinate. In each nostril, the olfactory membrane has a surface area of about 2.4 square centimeters

- **The receptor olfactory cells** - bipolar nerve cells (100 Million).The mucosal end of the olfactory cell forms a knob each with 4 to 25 olfactory hairs (also called olfactory cilia), measuring 0.3 micrometer in diameter and up to 200 micrometers in length. These cilia react to odors in the air and stimulate the olfactory cells. Activation of the receptor protein by the odorant substance activates the G-protein complex.cAMP opens many more sodium ion channels. Sodium ions increase the electrical potential thus exciting the olfactory neuron and transmitting action potentials through olfactory nerve. Therefore, even the most minute concentration of a specific odorant initiates a cascading effect that opens extremely large numbers of sodium channels.

- **Olfactory nerves** arise from olfactory receptor neurons in the olfactory mucosa. The axons collect into approximately 20 bundles and enter the anterior cranial fossa by passing through the foramina in the cribriform plate.They attach to the inferior surface of the **olfactory bulb.**Apparently unique in the nervous system, olfactory receptor neurons are continually replaced throughout life by differentiation of stem cells in the olfactory mucosa. Secondary sensory axons then form the olfactory tract.The olfactory bulb is continuous posteriorly with the **olfactory tract**, through which the output of the bulb passes directly to the olfactory cortex. The olfactory bulb is the rostral enlargement of the olfactory tract. The olfactory bulbs and tracts are parts of the brain that evaginated from the telencephalon in early development. Each bulb has

several thousand glomeruli**.**Each of which is the terminus for about 25,000 axons from olfactory cells. Each glomerulus also is the terminus for dendrites from about 25 large mitral cells and about 60 smaller tufted cells, the cell bodies of which lie in the olfactory bulb superior to the glomeruli.

- Laminar structure in the olfactory bulb from the surface inward:

1) The olfactory nerve layer – unmyelinated axons of the olfactory neuronesat different stages of growth, maturity or degeneration

2) Glomerular layer – incoming olfactory axons divide and synapse on terminal dendrites of secondary olfactory neurons that is, mitral, tufted and periglomerular cells

3) External plexiform layer – principal and secondary dendrites of mitral and tufted cells

4) Mitral cell layer – mitral cells and few granule cell bodies

5) Internal plexiform layer – axons, recurrent and deep collaterals of mitral and tufted cells and granule cell bodies

6) Granule cell layer – centripetal and centrifugal nerve fibers. The granule cell is likely to be a powerful inhibitory influence on the output neurons of the olfactory bulb

Olfactory Nucleus:--

- The olfactory tract leaves the posterior pole of the olfactory bulb to run along the olfactory sulcus on the orbital surface of the frontal lobe. The granule cell layer of the bulb is extended into the olfactory tract as scattered medium-sized multipolar neurons that constitute the anterior olfactory nucleus.
- As the olfactory tract approaches the anterior perforated substance, it divides into medial and lateral olfactorystriae.An intermediate striasometimes passes from the centre of thetrigone to end in a small olfactory tubercle.
- The olfactory tract divides into two pathways, one passing medially into the medial olfactory area (MOA) of the brain which represents a very old olfactory system, whereas the lateral olfactory area(LOA) is the input to (1) a less old olfactory system and (2) a newer system.

- **Medial olfactoryarea**: Most conspicuous are the septal nuclei, which are midline nuclei that feed into the hypothalamus, paraolfactory area, subcallosal gyrus and other primitive portions of the brain's limbic system. Lesion here will affect primitive emotional drives associated with smell like licking the lips, salivation, and other feeding responses caused by the smell of food.

- **Lateral olfactory area**: Composed mainly of the prepyriform and pyriform cortex, uncus, anterior hippocampal gyrus, entorhinal cortex and amygdaloid nuclei.

- Signal pathways pass into almost all portions of the limbic system, especially into less primitive portions such as the hippocampus. It also feed directly into an older part of the cerebral cortex called the paleocortex in the anteromedial portion of the temporal lobe.

Primary Olfactory Cortex consists of:-

- Anterior olfactory nucleus, pyriform cortex, amygdala nucleus, periamygdala complex, entorhinal cortex

CLINICAL EXAMINATION

- **History:** Past head injury, smoking, recent URTI, systemic illness, exposure to toxins, medications, unilateral loss of smell is more significant than bilateral.

- **Method:** Ask the patient to close his eyes while presenting a series of nonirritating, familiar olfactory stimuli such as coffee or chocolate .The aromatic stimulus should be placed under one nostril while the other nostril is occluded. The patient is asked to sniff the substance and then identify it. The procedure is repeated for the other nostril. If the patient can name or describe the substance, it is assumed that the olfactory tract is intact. Small bottles containing essences of familiar odors - Coffee, almonds, chocolate, oil of lemon and peppermint, soap, camphur. Patient is askedi) if he can smell anything, ii) if he can identify the odour, iii) If the odour is same in each nostril.

- **DOES**:Close eyes, One nostril to be closed

- **DONTS**:Don't use irritating stimuli like ammonia

- **The Newer Pathway**:- A newer olfactory pathway has now beenfound that passes through the thalamus, passing to the dorsomedial thalamic nucleus and then to the lateroposterior quadrant of the orbitofrontale cortex. Based on studies in monkeys, this newer system probably helps in the conscious analysis of odor.

Olfactory disease:-

- **Anosmia** - upper respiratory tract infection, head injury, nose and sinus disease, idiopathic.
- **Hyposmia**-Parkinson disease, Alzheimer disease, and HIV–dementia complex.
- **Hyperosmia** - migraine headache.
- The perversion of smell (**parosmia)** and unpleasant odors (cacosmia) occur in psychiatric disease and develop after head trauma.
- **Olfactory hallucinations** - aura in temporal lobe seizures because of involvement of primary olfactory cortex

Important points:

- **Foster Kennedy Syndrome** includes anosmia, ipsilateral optic atrophy, contralateral papilloedemadue to a large tumor involving the orbitofrontal region
- **Kallmann's Syndrome** is a X linked hereditary disorder manifesting with hypogonadism and anosmia
- Chronic intranasal cocaine use may cause anosmia.
- Lesions of the olfactory cortex does not cause anosmia due to bilateral innervations

HYPOGLOSSAL NERVE

The hypoglossal nerve is a purely motor nerve. It supplies to all the muscles of the tongue, except the palatoglossus**.**

ANATOMY

- **Nucleus :**

Hypoglossal nuclei are upward extensions of the anterior gray columns of the spinal cord. The paired nuclei extend almost the entire length of the medulla just beneath the floor of the fourth ventricle close to midline.

- **Course:**

  It emerges from the medulla in the sulcus between the pyramid and inferior olive as a series of 10 to 15 rootlets on each side anterior to the rootlets of IX, X, XI. The hypoglossal rootlets run laterally behind the vertebral artery, collected into two bundles that perforate the dura mater separately then enter the hypoglossal canal in the occipital bone. The nerve emerges from the canal in a plane medial to the internal jugular vein, internal carotid artery and ninth, tenth and eleventh cranial nerves and passes inferolaterally behind the internal carotid artery and glossopharyngeal and vagus nerves to the interval between the artery and the internal jugular vein. There it makes a half-spiral turn around the inferior vagal ganglion. It then descends almost vertically between the vessels and anterior to the vagus to a point level with the angle of the mandible, becoming superficial below the posterior belly of the digastric and emerging between the internal jugular vein and internal carotid artery. It loops around the inferior sternocleidomastoid branch of the occipital artery and crosses lateral to both the internal and external carotid arteries and the loop of the lingual artery a little above the tip of the greater cornu of the hyoid.It is crossed itself by the facial vein.

➢ The hypoglossal nerve communicates with:

  - First and second cervical nerves
  - The sympathetic trunk
  - Vagus (inferior vagal nuclei)
  - Lingual nerve

- Near the atlas it is joined by branches from the superior cervical sympathetic ganglion and by a filament from the loop between the first and second cervical nerves, which leaves the hypoglossal as the upper root of the ansa cervicalis.The vagal connections occur close to the skull, and numerous filaments pass between the hypoglossal nerve

and the inferior vagal ganglion in the connective tissue. As the hypoglossal nerve curves around the occipital artery, it receives the ramus lingualis vagi from the pharyngeal plexus. Near the anterior border of the hyoglossus, it is connected with the lingual nerve by many filaments that ascend on the muscle.

- The branches of distribution of the hypoglossal nerve aremeningeal, descending, thyrohyoid and muscular nerves.

CLINICAL EXAMINATION

- Evaluate for any tongue deviation, strength, bulk and abnormal movements. Note the position and appearance of the tongue at rest in the mouth.
- Patient is then asked to protrude it out, move it in and out, side to side (inside mouth) upward downward (inside mouth)both slowly and rapidly. Motor power can be tested by having the patient press the tip against each cheek as the examiner tries to dislodge it with finger pressure. For more precise testing press firmly with a tongue blade against the side of the protruded tongue comparing the strength on the two sides.When unilateral weakness is present the tongue deviates towards the weak side on protrusion because of the action of the normal genioglossus which protrudes the tip of the tongue by drawing the root forward. In case of unilateral weakness there is impairment of the ability to deviate the protruded tongue toward the non paretic side and the ability to push it against the cheek on the sound side, but the patient is able to push it against the cheek on the paralyzed side.
- If paralysis is bilateral, the tongue is motionless. Taste and tactile sensibility are unaffected, but articulation is slow and swallowing is very difficult.
- Abnormal movements of the tongue are tremors and fasciculations.Tremors will usually disappear when the tongue is lying at rest in the mouth whereas fasciculationspersist. Coarse tremors of the tongue can occur in Parkinsonism and alcoholism. Fine tremors occur in thyrotoxicosis.

- Tongue weakness may be :
    - Supranuclear
    - Nuclear
    - Infranuclear

- **Supranuclear lesions** cause weakness but no atrophy and the weakness is less severe.Genioglossus has crossed supranuclear innervation. Tongue protrudes toward the weak side but to the side opposite the supranuclear lesioncaused by a destructive lesion of the cerebral cortex or the corticobulbar tract in the internal capsule, cerebral peduncle or pons.In nuclear and infranuclear lesions there is atrophy of the involved side. In addition to weaknessprogressive nuclear lesions like motor neuron disease cause fasciculations in addition to weakness. Nuclear lesions are commonly caused by neoplasms, vascular lesions and motor neuron disease. Rare causes of nuclear lesions are syringobulbia, abscess, granuloma, syphilis, and poliomyelitis.

- **Infranuclear involvement** involving intracranial course is seen in infectious and neoplastic meningitis, subarachnoid hemorrhage, neoplasms like schwannoma, inflammation and trauma.Infranuclear involment involving the extracranial course is seen in penetrating wounds to neck, neck dissection, surgery, carotid aneurysms, tumors or infections in the retroparotid or retropharyngeal spaces, deep cervical adenopathy, cranial irradiation and tumors of the salivary glands.

FUNDUS EXAMINATION

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Fundus examination is an integral part of routine neurosurgery / neurological examination. A neurophysician / neurosurgeon should be well versed with basic Neuro-ophthalmological workup as well as fundus examination as many a times it obviates ophthalmology references and saves patient's and surgeon's time.

Fundus examination can be done by:

1. Direct ophthalmoscopy
2. Indirect ophthalmoscopy
3. Slit Lamp biomicroscopy
4. Fundus Camera

Direct ophthalmoscopy

Direct ophthalmoscope was originally invented by Charles Babbage in 1857.It was reinvented by Hermann Von Helmholtz. In 1951 Josh Zele and Jon Palumbo invented the world's first hand held direct illuminating ophthalmoscope.

We are not able to see the fundus with naked eye when we throw torch light into the subject's eye because:

1. 1/10000 to 1/100000 of light entering the eye is reflected back.
2. Observer's eye blocks the rays of the subject's retina
3. The pupil limits the view

To view the retina, by illuminating subject's retina 3 things needs to be fulfilled:

1. Light source should be extremely bright.
2. Light source needs to be placed very close to the observer's eye(maintaining the illumination axis and viewing axis as close as possible).**(Fig 1)**
3. Subject's should be accommodated to infinity (subject should be looking at far distance).

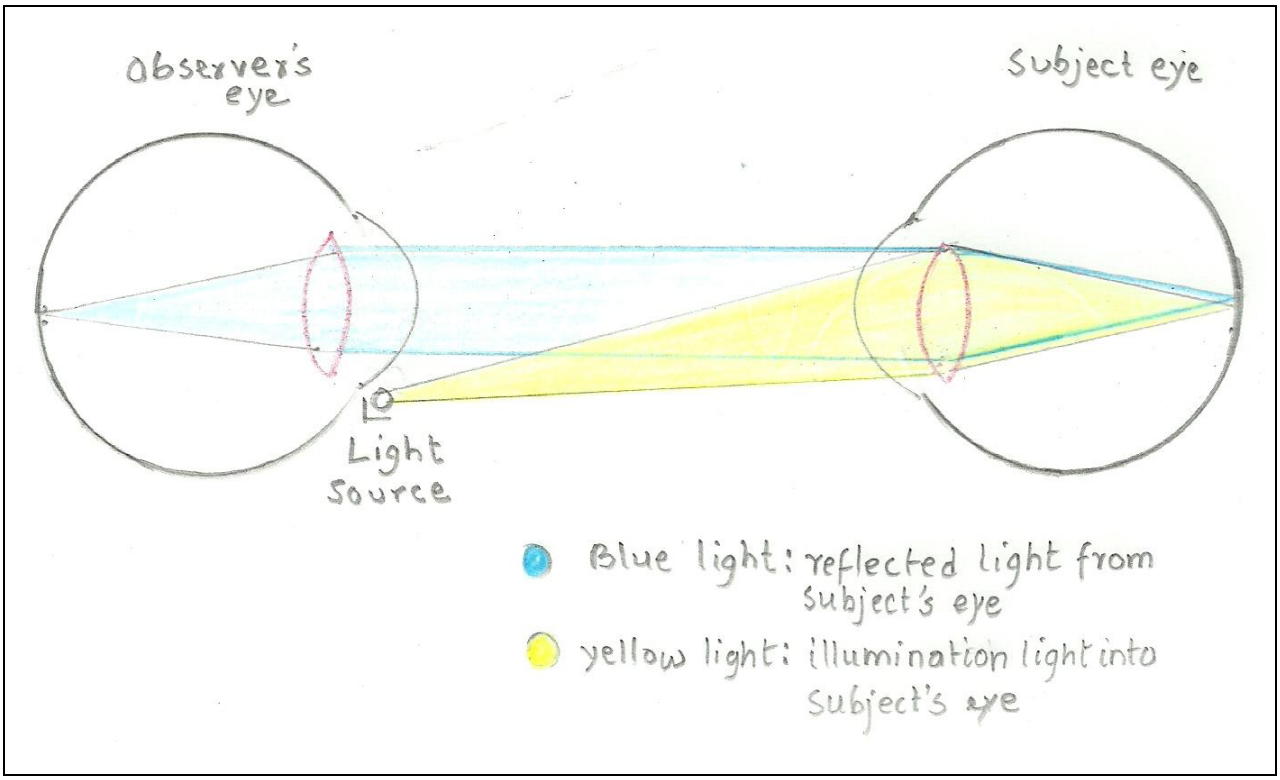

Fig:1 Making the human eye appear luminous,light source shielded from observer,s eye

A hand held direct ophthalmoscope has following features:**(Fig 2)**

1. Illumination bulb and illumination aperture adjustment
2. Viewing aperture
3. A 45º tilted mirror
4. Compensating lens section
5. Cone of Illuminating light rays

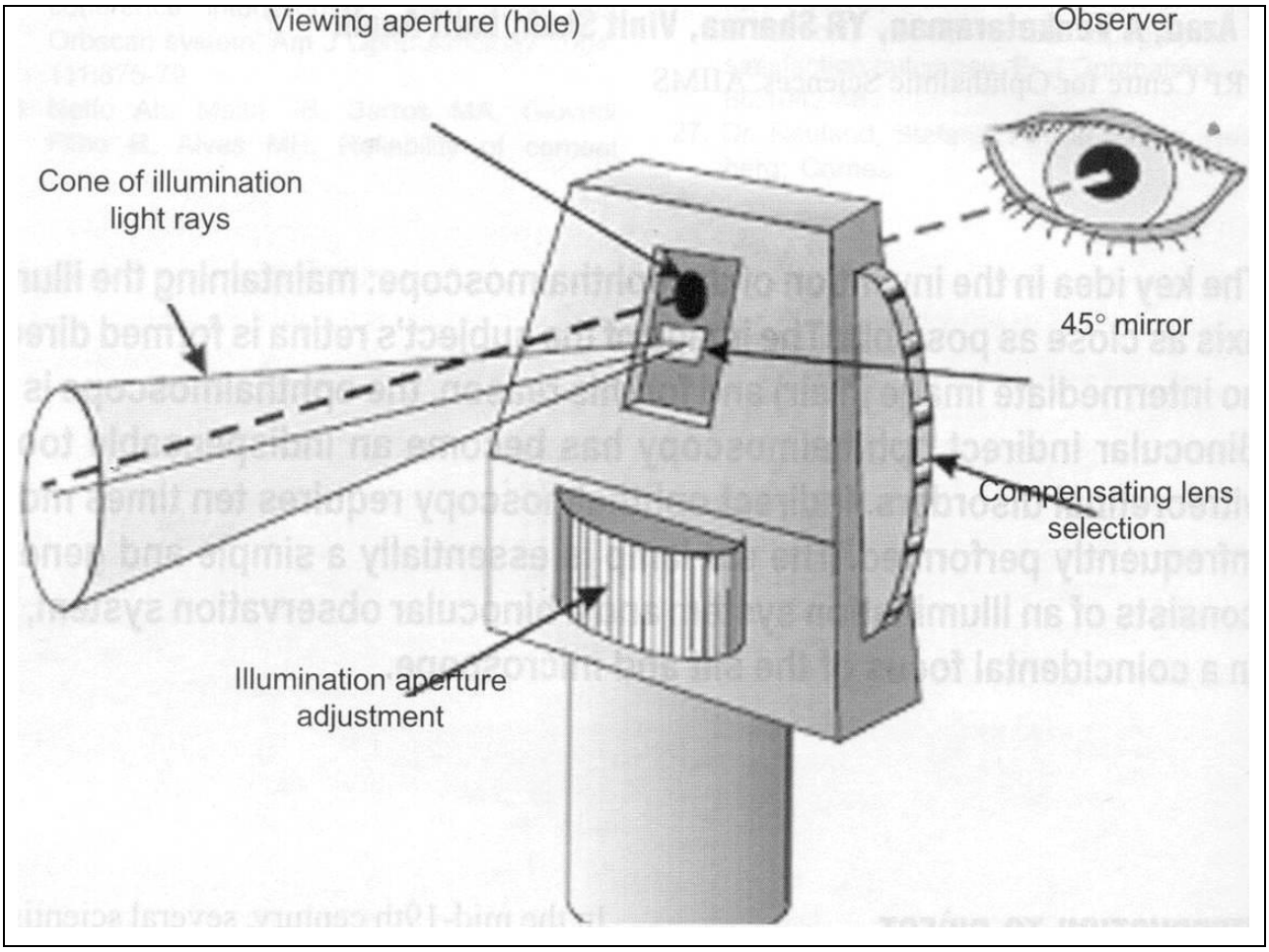

Figure 2: Showing detailed structures of a direct hand held ophthalmoscope

Further, details of hand held direct ophthalmoscope can be understood,in depth, in the **Fig 3**. Light from the light source in the ophthalmoscope is condensed into parallel pipe with the help of condensing lenses in the ophthalmoscope and then passes through desired aperture in the ophthalmoscope. This condensed light is then directed to tilted transparent mirror to reflect it into subject's eye. The reflected light is then passes through transparent tilted mirror, to be directed back into the observer's eye. This reflected light is then focused into the observer's eye using the appropriate compensatory lenses (situated in the ophthalmoscope).If an **observe**r is myopic, i.e. having minus power lenses in his spectacles, should use minus lenses (marked by red colour in compensating lens section in the ophthalmoscope)while viewing subject's fundus. Similarly if an **observer** is hypermetropic i.e. having plus power lenses in his spectacles, should

use plus lenses(marked by black or green colour in compensating lens section in the ophthalmoscope).

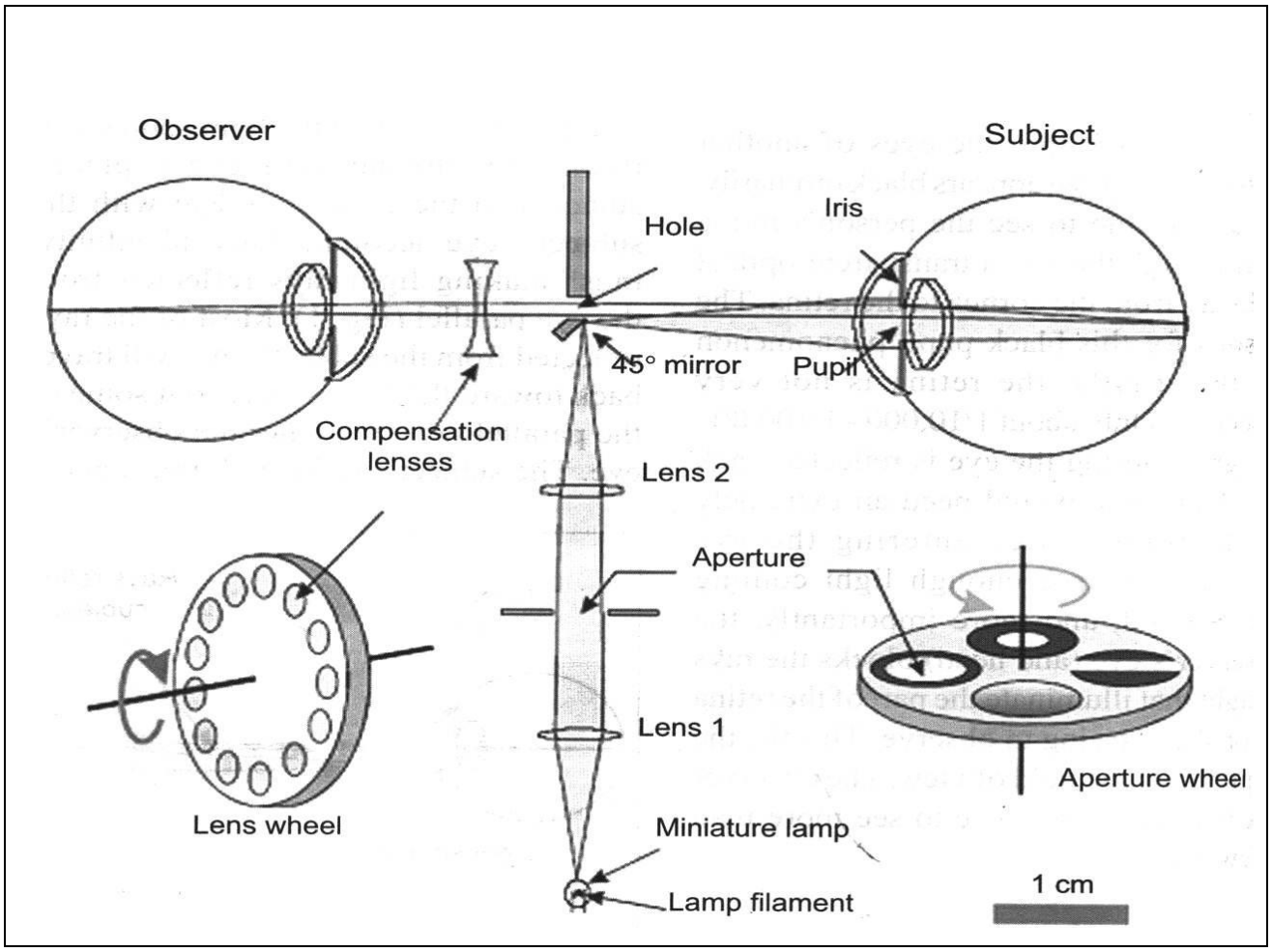

Fig : 3 Showing how an observer peep's through ophthalmoscope into subject's eye. A light source in the ophthalmoscope directs the light through tilted transparent mirror into the subject's eye and reflected light from subject's eye is directed into observer's eye using compensatory lenses.

By direct Ophthalmoscopy with the help of an eclectic ophthalmoscope the fundus details are 15 times magnified and the image formed is a virtual, erect image of the fundus.

During fundus examination the following points are to be noted:

1. ***Optic disc*** **-** It is a round or oval structure, pale pink in colour, situated at the posterior part of the fundus. It is also known as the optic nerve head as the optic nerve starts from this point. There is an excavation, at the central part known as the physiological cup**(fig 4)**, whose depth and extent varies in different subjects. The margins of the disc are normally sharp and distinct. So during examination of the fundus the colour, shape, margins of the disc, the physiological cup, presence of any abnormal vessels and or any other abnormality on the disc have to be noted.

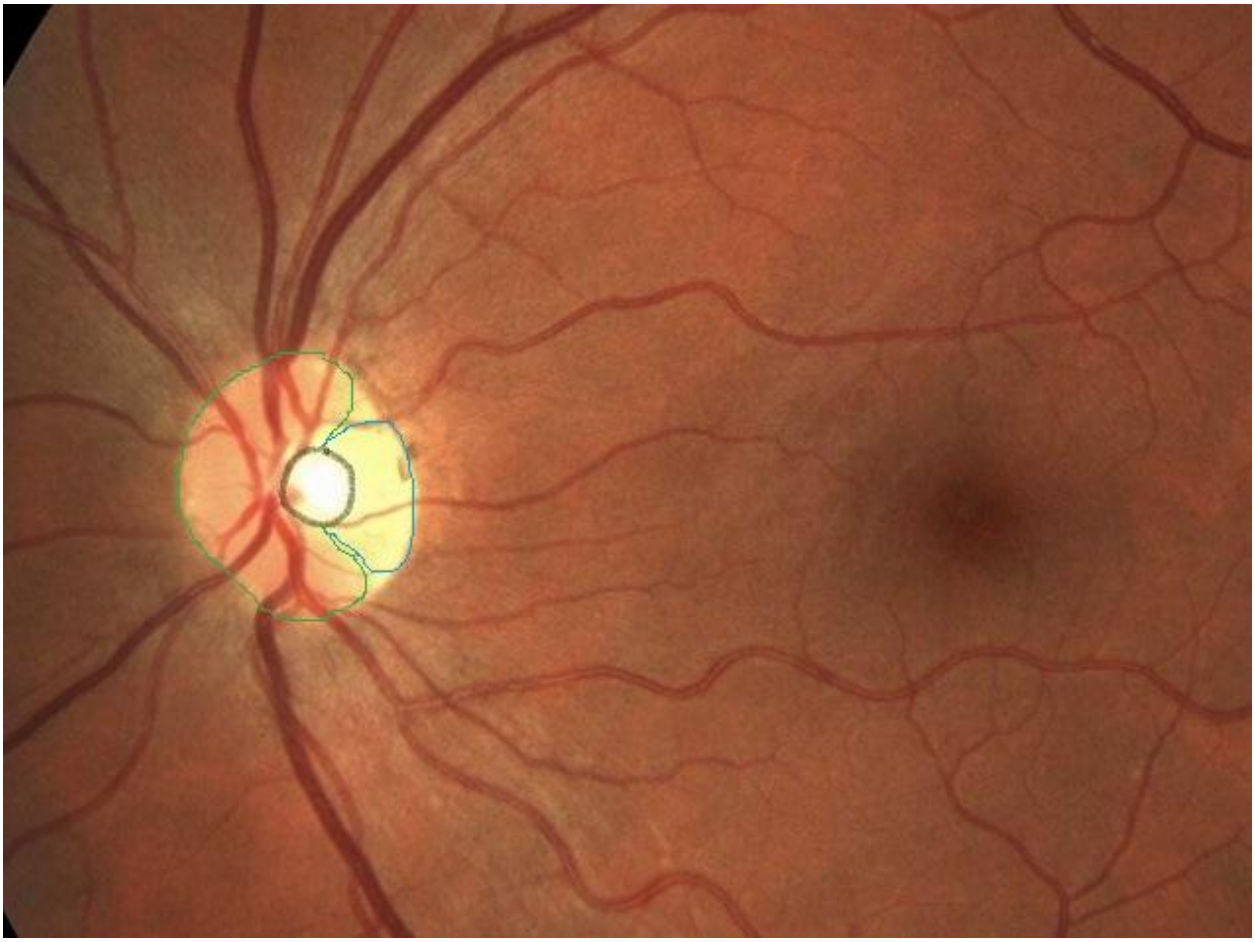

Fig:4 Physiological cup (Black circle),Area circled in green denotes pinkish hue of Neuroretinal rim and area circled in blue shows normal temporal pallor of the Disc. In Optic atrophy the pinkish NR rim area also turns pale in colour

The other important structural entity is the Neuro retinal rim(NRR) in optic disc, which is the area between the disc margin all around & the margins of the physiological cup of the optic disc.(**as shown in fig 4**).Again its(NRR) thickness, colour, shape and volume

matters for the ophthalmologist, but for physician and surgeons mainly its colour has importance. The normal Neuro retinal rim has pinkish hue due to the capillaries present on the disc surface, but if it is having pallor developing or set in, then definitely it indicates the setting of optic atrophy.

2. ***Macular area*** **-** It is a small circular area, between superior and inferior temporal arcade, with a red colour which is darker than that of the surrounding fundus and center of which is situated at about 2 to 2 1/2 disc diameter in the temporal side of the optic disc, at a level slightly below the center of the disc**(Fig 5)**. The glistening center point of the macular area is known as the fovea centralis. It is slightly depressed and this depression is known as the foveal pit. During ophthalmoscopy, due to reflection of light, this pit appears as a bright reflex which is known as foveal reflex.

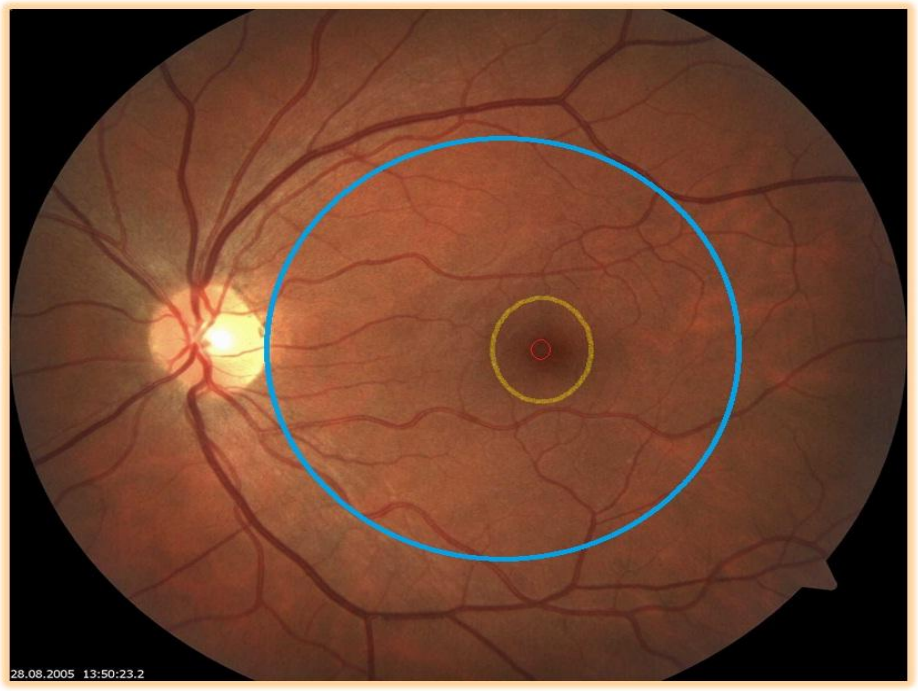

*Fig:5 Area between temporal arcade vessels-**Macula**(marked blue),**Fovea** is marked yellow, **Foveola (fovea pit)** is marked red*

3. ***Retinal blood vessels –*** These consist of arteries which are the branches of the central artery of the retina and the veins which are the branches of the central vein of the retina.

These vessels radiate from the optic disc and as they spread over the fundus & they divide dichotomously into many branches. The arteries, which are in fact arterioles, at a short distance from the disc are narrower than the veins, the ratio of the caliber of an artery to a vein being 2:3. The arteries which are end arteries are bright red in colour where as the veins are purplish red (**Fig 6)**. Normally the walls of the blood vessels are transparent; so during ophthalmoscopy the light gets reflected from the blood column within the vessel. This reflection appears as a light steak on the wall of the vessels which is more marked in case of the arteries. During examination of the fundus, any narrowing, tortuosity or dilation or sheathing of the vessels, any alteration in the light reflex from the vessel wall and any venous compression at the arterio-venous crossings are to be noted. All the blood vessels should be traced up to the periphery.

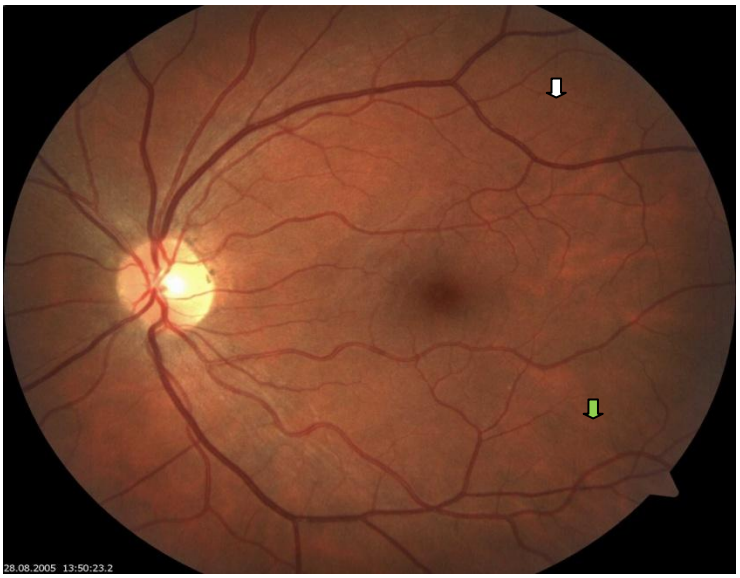

Fig:6 Retinal artery (green arrow), Retinal Vein (white arrow)

4. ***General appearance of the fundus*** **–** Normally the fundus has a uniform red appearance. But the colour depends on the degree of pigment in the retinal pigment epithelium. Frequently shimmering reflexes are seen on the surface of the fundus during examination with an ophthalmoscope and these are most evident in hypermetropic eyes. The fundus should be examined all round for any abnormality.
5. ***Choroidal vessels –*** normally many Choroidal vessels are not visible. But if pigmentation of the retinal pigment epithelium is deficient they are seen in the areas between the retinal

vessels. They appear as pinkish flat ribbons **(Fig 7)** which do not show any light reflex. They also anastomose freely. Sclerosed Choroidal vessels are whitish in colour and narrow. If the pigment content of the choroid is more than that of the retinal pigment epithelium, deeply pigmented polygonal areas may be seen in between the Choroidal vessels. This appearance is known as tessellated or fibroid fundus.

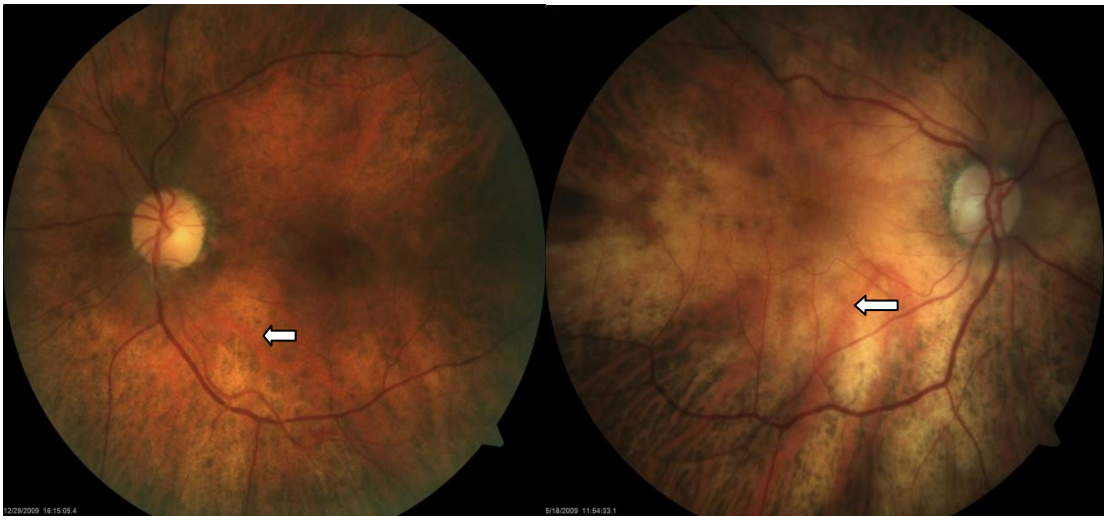

Fig 7: Showing ribbon shaped Choroidal vessels as (arrows) and Tessellated fundus showing pigmented polygonal areas beneath retinal vessls

6. ***Examination of the fundus with the red–free light*** **–** The ordinary white light used for Ophthalmoscopy can penetrate the layers of the retina i.e. the retina is transparent to white light. But a light with shorter wave length, like blue light(called red free light for ophthalmoscopy), does not possess this penetrating power and so this type of red free light gets reflected from the retinal surface. The nerve fiber layer gets accentuated superior and inferior to the disc with this light, the striations of the nerve fiber layer can be seen superior and inferior to the disc(as seen in **Fig 7a**). Due to this property of the red free light minute pathological changes in the retinal nerve fibers or in the macular area are easily detected when red free light is used for Ophthalmoscopy.

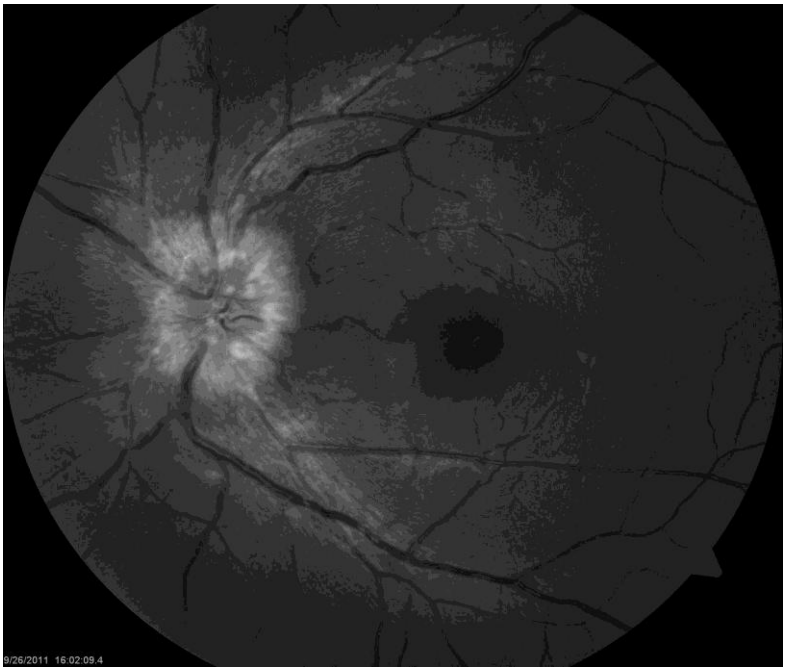

Fig 7a: Red free light photograph, striations of the nerve fiber layer can be seen superior and inferior to the disc

7. ***Linear measurement*** – Any lesion seen in fundus can be measured in terms of size and its location can be depicted for future follow up.This can be done in two ways in terms of diameters of the optic disc, e.g. it is situated so many disc diameters on the temporal or nasal side form the margin of the disc, & it is so many diameters of the optic disc size, in length and breadth .

8. ***Direct Ophthalmoscope Technique***

- The examination should be done in a dimly lighted room.
- The subject should be seated comfortably on a chair .
- Ask the subject to see at some far object so that his accommodation is relaxed.
- Ideally the pupil should be fully dilated to get better details and view of whole fundus.
- Right eye of the subject should be examined with the right eye by the examiner and vice versa for the left eye.
- Approach the subject's eye from the side on and not directly from the front.

- Start from low illumination of the ophthalmoscope then gradually increase the illumination with the rheostat provided in the ophthalmoscope, so that subject can better acclimatize to the light intensity.
- Do distant direct ophthalmoscopy first i.e. throw the light ,first ,from a distance of appx. 35 cms on to the both pupils and see for a uniform glow in the pupils. And if uniform glow is not found in both pupils then there may be media opacities such as cataract or some cornea or vitreous pathology. In such scenario you may need to opt for Indirect ophthalmoscope or slit lamp biomicroscopy to get a better view of the fundus.
- After doing distance direct ophthalmoscopy now approach the subject's eye side on i.e. right side of the right eye and left side of the left eye. Now first thing you will focus, while you approach near to eye ,is optic disc and if you move slight nasally to eye in question you will be able to see macula and temporal retina, similarly if you move up and down while seeing the fundus(if patient is keeping his eyes straight),then you will be able to see superior and inferior retina respectively.
- But if dilation of pupil is contraindicated as in some neurological or neurosurgical condition then dim light in the room helps to get some dilation and a better view of the fundus. In such cases, while viewing the fundus, intensity of the illumination light of the ophthalmoscope should be kept on lower side.

9. ***With the help of a direct ophthalmoscope*** – with the help of the lenses incorporated in the ophthalmoscope the top of an elevated area or the floor of an excavated area is focused. During the examination, the ophthalmoscope used should be held at the anterior focal plane of the patient's eye i.e. at 15.7 mm in front of his cornea, in the normal case, but at 23.27 mm in front of the cornea if the eye is aphakic(when the lens is removed) and the observer must keep his accommodation completely relaxed. In case of a depression in retina, such as in coloboma of retina choroid (***fig 8)***, concave lenses or minus lenses in the ophthalmoscope have to be used. The power of the plus or minus lens required to focus either the top surface orthe floor of a lesion is noted. The nanotech convenient object in the fundus, as for example a blood vessel is focused and the

difference in the power of lenses required to focus an object in the fundus and the top or floor of the lesion is noted. A difference of focusing of three diopters between two points on the fundus indicates a difference of level of approximately 1 mm. This is particularly useful in seeing the details in papilloedema when the disc surface is elevated. Observer has to use plus lenses in the ophthalmoscope to see the finer details on the surface of the disc.

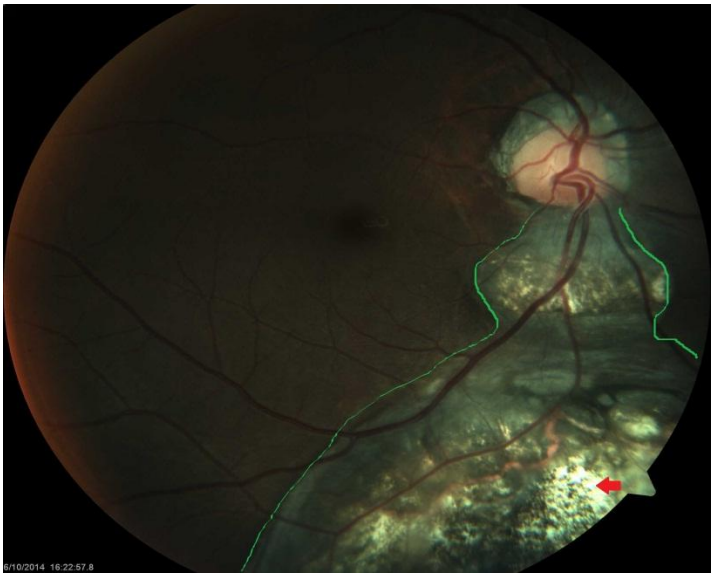

*Fig 8: Coloboma seen **(outlined by green)**, whitish sclera**(orange arrow)**is seen through coloboma*

A hand held direct ophthalmoscope may be used to examine the anterior segment, but is more useful for the posterior segment. The field of view for fundus examination is approximately 8° and, is limited by the most oblique light ray leaving the subjects pupil which enters the observer pupil. It is therefore maximized by using a larger illumination aperture and by dilating the pupils of both the subject and observer and bringing them closer.The image is virtual, erect, and magnified. The image of an emmetropic fundus is magnified 15X, whereas in a -10 diopters myope, it increases to 19X and in a +10 diopters hypermetrope it reduces to 13X. In eccentric gaze, the pupil becomes effectively elliptical, therefore, when viewing the superior or inferior fundus periphery, the ophthalmoscope should be rotated.

10. ***Many direct ophthalmoscopes have additional features*:** the contour of structures in the anterior segment and fundus and anterior chamber depth may be shown if illuminated obliquely by a slit beam. Projection of a star, cross, or reticule on to the retinal may establish eccentric fixation if it does not fall on the Foveola, when the subject looks directly at it. A green (red free filter) accentuates vascular structures, retinal nerve fiber changes and retinal holes. Cobalt blue and yellow filters respectively, highlight fluorescein and eliminate retinotoxic blue and ultraviolet wavelengths. The density of opacities in the ocular media can be assessed against the red reflex and in such eyes; illumination may be insufficiently bright to see fundus details clearly.

Other methods of Fundus Examination

Indirect Ophthalmoscope:

This method uses head mounted binocular ophthalmoscope to visualize the fundus with the help of hand held non contact lenses such as 20 D, 14D, which provides a large field as well as stereoscopic view of the fundus field. **(Fig 9)**

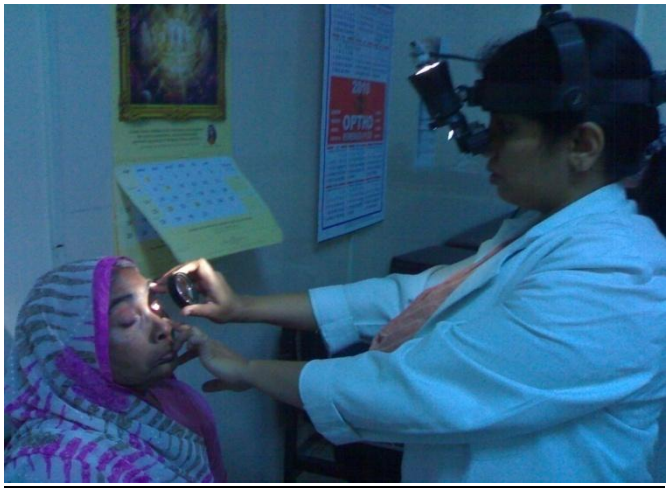

Fig:9 Showing Indirect ophthalmoscopy being done with a hand held 20 D lens

Slit Lamp Biomicroscopy:

This method with 60.78 and 90 diopters lenses allow another noncontact fundus examination. Held before the eye in combination with a slit lamp, these lenses allow quick, noncontact examination at various magnifications. (Fig 10)

These convenient optical methods allow direct clinical examination of the various tissues of the fundus in vivo. These tissues include the nerve tissue of the optic nerve and neural retina, the neurosensory layer of the retina, the pigment epithelium of the retina, the retinal vascular system, the choroid the vitreous body, and occasionally the connective tissue of the sclera.

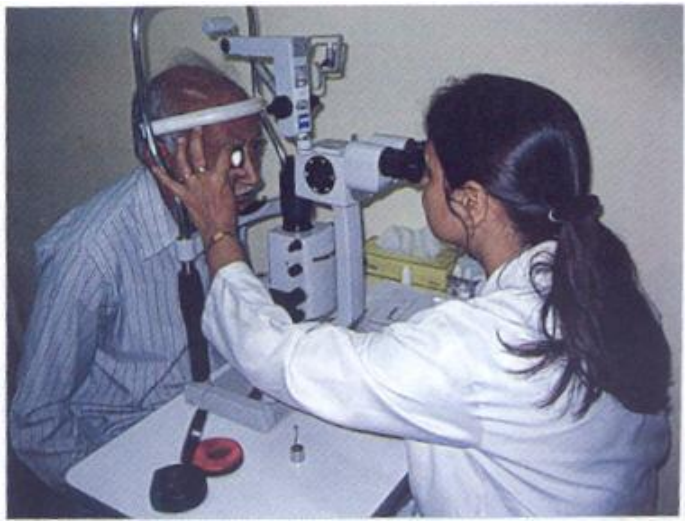

Fig.10 Slit lamp non contact biomicroscopy being done with a hand held 90D lens

Papilloedema

About 1.2 million axons converge at the optic disc to form the optic nerve. The optic nerve follows a 50mm course as it extends from the back of the eye, travels through the orbit, passes through optic canal, runs intracranial, and partially decussate along with contralateral optic nerve to form the optic chiasm. Each axon must maintain active axonal transport in both the orthograde (eye to brain) and retrograde direction. The subarachnoid space of the brain is continuous with the optic nerve sheath .A wide variety of insults may lead to dysfunction or compression of the optic nerve, potentially resulting in partial arrest of axoplasmic transport, manifested as optic disc oedema. If the compression is caused by raised intracranial pressure, the condition is termed papilloedema, thus carrying neurological and neurosurgical connotations. If the cause of the disc oedema is not increased intracranial pressure then the term optic disc oedema should be used instead of papilloedema. Long standing or severe papilloedema, in addition to reflecting

intracranial pathology, also may result in bilateral optic nerve dysfunction because of compromised axonal integrity at the lamina cribrosa.

Fundus changes in Disc Oedema

Ten clinical signs of Optic disc Oedema can be seen by direct ophthalmoscopy. It is better performed with both standard (white light) and red free light(to better visualize the nerve fiber layer). Although indirect ophthalmoscopy with a 20 D lens may reveal most of the changes, a 60, 78, or 90 D Lens used in Slit lamp biomicroscopy provides the best noncontact stereoscopic view of the optic nerve head. It is useful to divide 10 fundoscopic signs of disc oedema into 5 mechanical and 5 vascular signs.

The five mechanical signs:

Blurring of optic disc margins

Filling in off the optic disc cup

Anterior extension of the optic nerve head

Oedema of the nerve fiber layer

Retinal or choroidal folds, or both

The 5 vascular signs are –

Venous congestion of arcuate and peripapillary vessels

Papillary and retinal peripapillary hemorrhages

Nerve fiber layer infarct (cotton wool spots)

Hyperemia of the optic nerve head

Hard exudates of optic disc

Optic disc oedema can be characterized by as early, fully developed, chronic or late (atrophic).

Early Disc oedema **(Fig11)**:

Disc swelling

Blurring of optic disc margins

Blurring of nerve fiber layer

Optic disc hyperemia

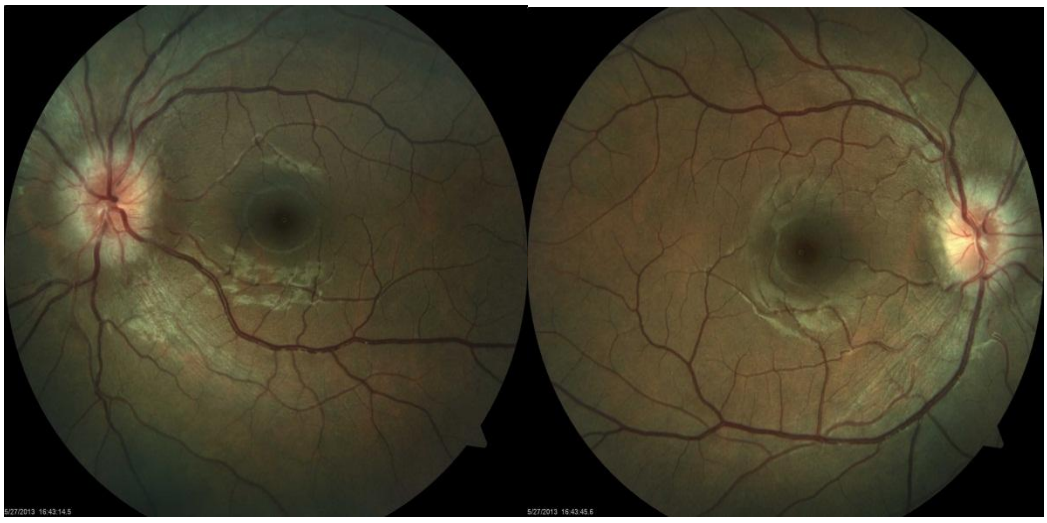

Fig 11: Bilateral Early Papilloedema oedema showing all the features as described

The earliest change may be flattening of the internal limiting membrane leading to loss of superficial light reflexes, as seen by red free light direct ophthalmoscopy.**(Fig 12).**

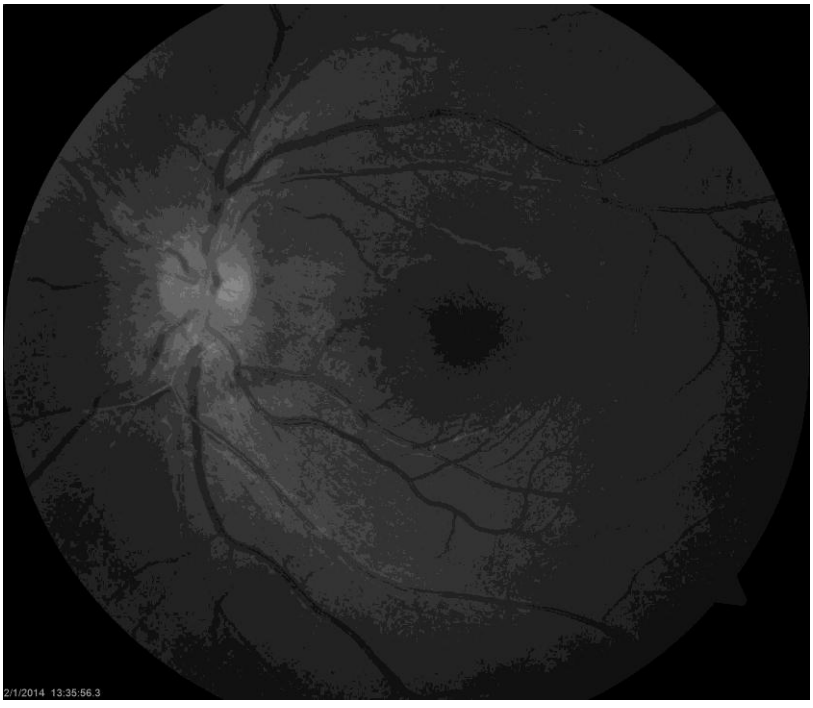

Fig 12: red free light fundus photograph showing loss of superficial light reflexes

Fully developed papilloedema : (Fig 13& 14)

Optic nerve head is markedly elevated

Circumferential retinal folds(striae or paton's line) as well as parallel choroidal folds

Engorged & dusky veins

Peripapillary splinter haemorrhages

Hard exudates & haemorrhages may be noted around the disc or in the macula

Retinal haemorrhages may dissect anteriorly to subhyaloid or vitreous spaces**(Fig 13)**.

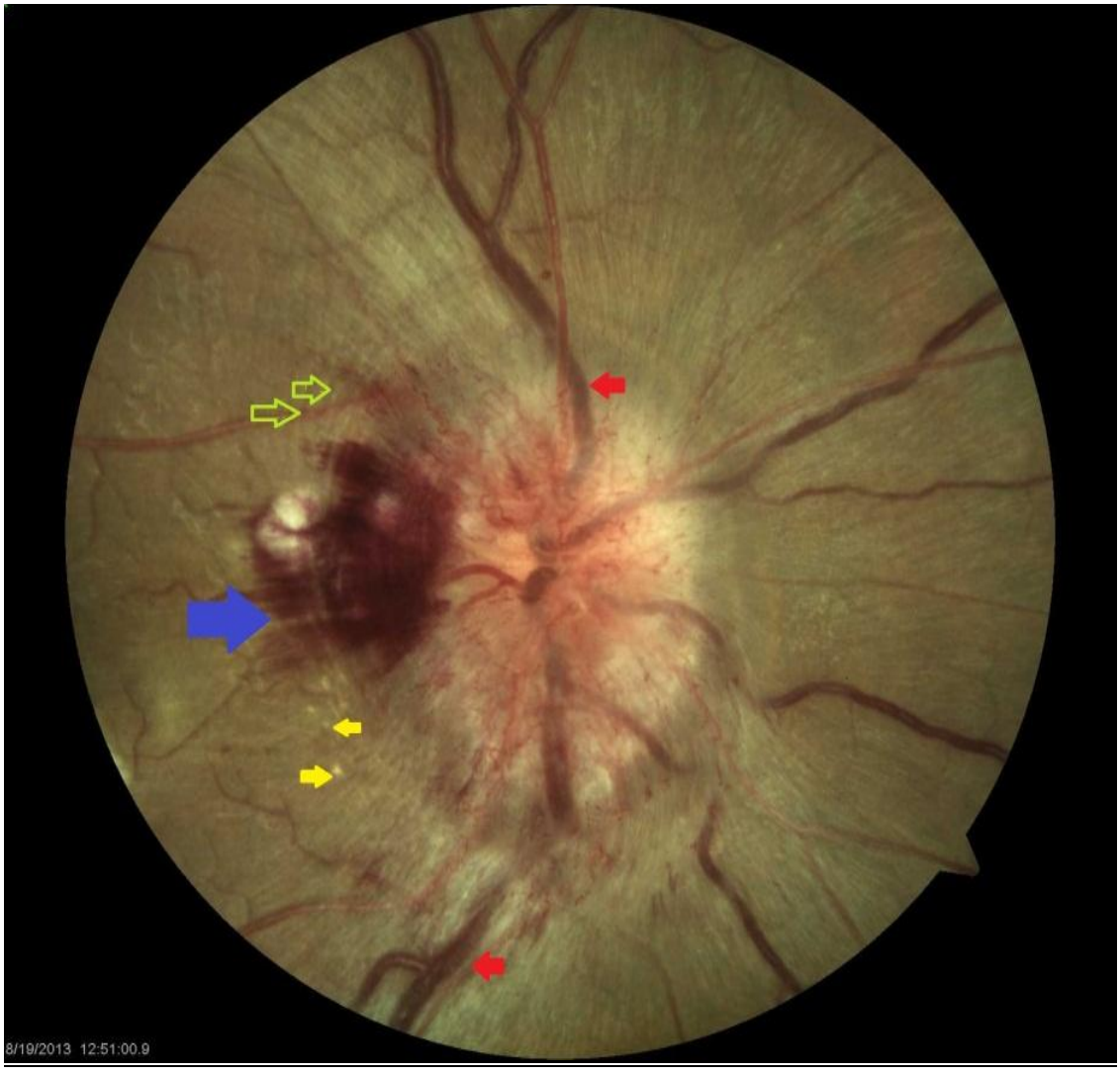

*Fig:13 Fully developed Papilloedema, Engorged dusky veins(**Red arrow),** Circumferential retinal folds (paton's lines)**(green arrow heads)**, Hard exudates around disc**(yellow arrow),** Subhyaloid hgh(blue arrow)*

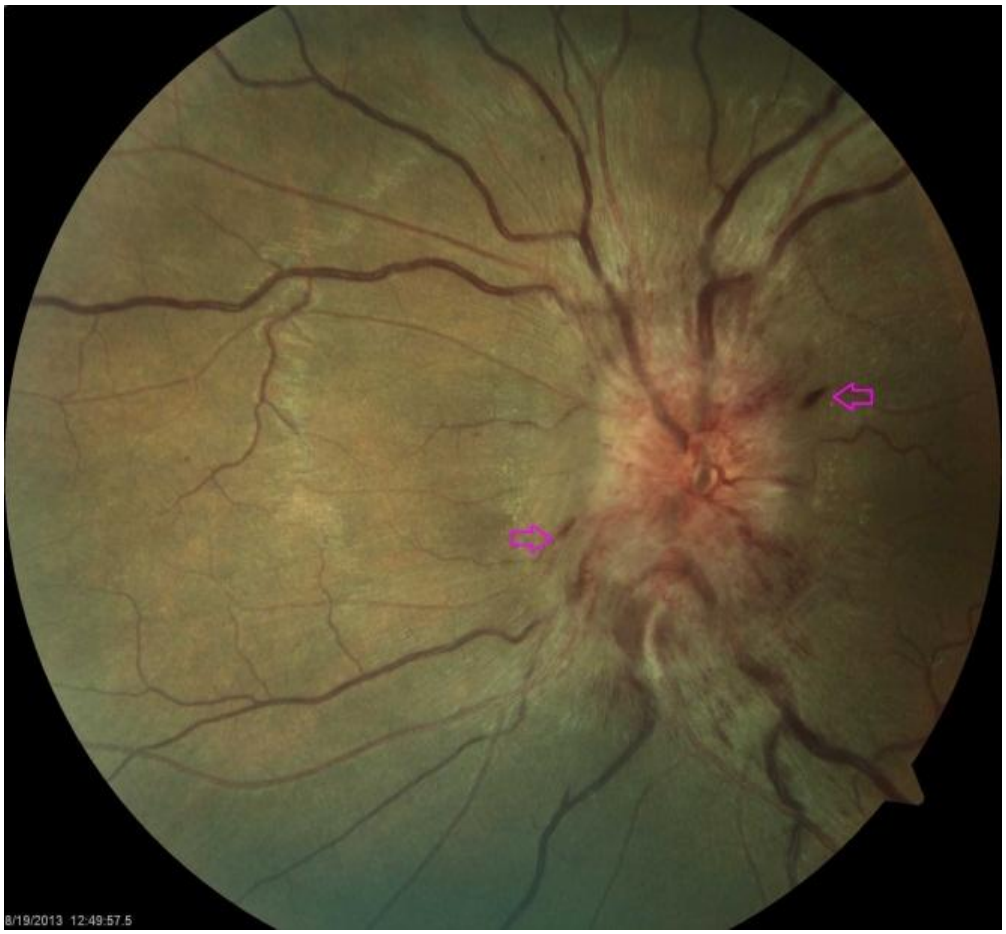

*Fig:14 Fully developed Papilloedema, showing splinter haemorrhages**(pink arrow)***

Chronic Papilloedema : (Fig 15)

The optic disc cup is completely obliterated

Less disc hyperemia

Fewer haemorrhages

More prominent hard exudates observed within the optic nerve head

Chronic papilloedema may persist for months or even years without a change occurring in appearance or severe visual impairment.

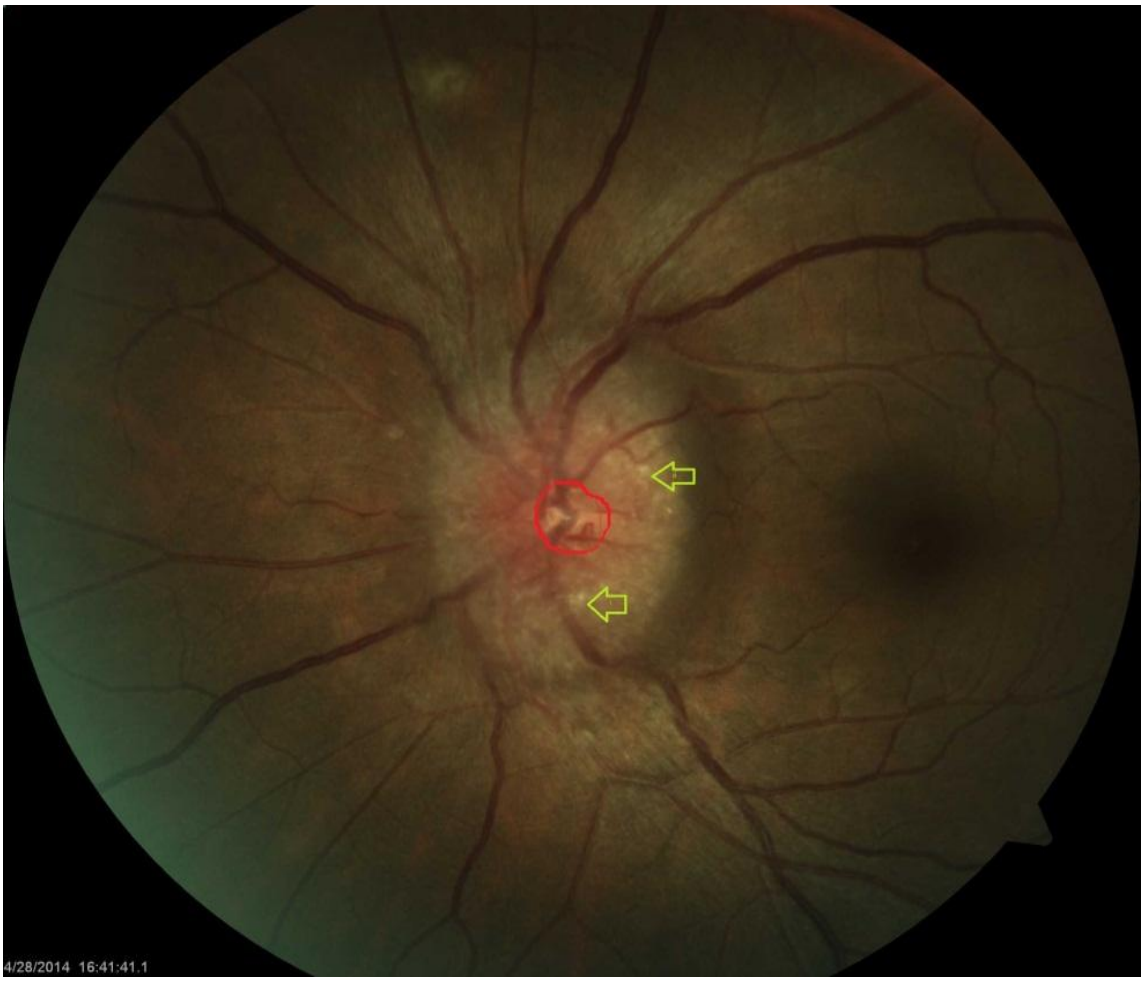

Fig: 15 Chronic papilloedema , less disc hyperemia, Optic disc cup obliterated ***(red circle),*** *Hard exudates* ***(Green arrow)****with in optic nerve substance.*

Late Optic disc oedema: **(Fig 16):**

Secondary optic atrophy occurs

Disc swelling subsides

Retinal arterioles are narrowed or sheathed

Optic disc appears dirty grey and blurred due to gliosis

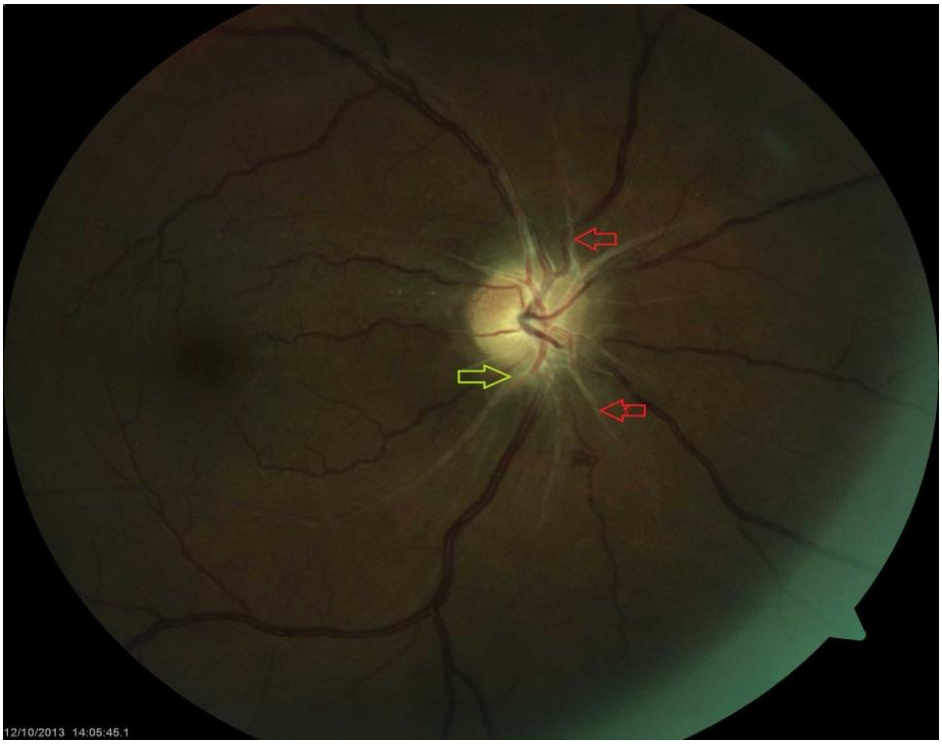

Fig: 16 Late Papilloedema, Disc swelling subsided, Sheathed retinal arteriole ***(red arrow),*** *blurred dirty margins* ***(green arrow).***

Optic atrophy

Optic atrophy is a misnomer. Although the term atrophy generally refers to physiologic involution or reduction, optic atrophy refers to cell death. In particular, optic atrophy represents the permanent loss of retinal ganglion cell axons in conjunction with retinal ganglion cell death. Optic atrophy should not be considered a diagnosis; it is a pathologic end point that is clinically discernible but does not imply cause. Optic atrophy may be due to injury of the optic nerve head. However, because of antero- grade and retrograde degeneration, it may reflect upstream injury of the posterior optic nerve, optic chiasm, or optic tract. Differentiating primary from secondary optic atrophy is clinically useful.

Primary optic atrophy

The term primary optic atrophy may be confusing because primary optic atrophy is caused by injury of the retinal ganglion cell or its axon, as a result of myriad inflammatory, vascular, or compressive causes. A less confusing alternative term would be simple optic atrophy. The essence of primary or simple optic atrophy is a loss of optic nerve fibers with otherwise minimal

disturbance of the optic nerve head microanatomy. In particular primary optic atrophy is attended by only minimal gliosis of the optic nerve head.

Primary optic atrophy can be a consequence of any injury of the retinal ganglion cell or its axon at the level of the nerve fiber layer; the optic nerve head; the orbital, the intracanalicular, or intracranial optic nerve; the optic chiasm; the optic tract; or the lateral geniculate nucleus. Common causes of primary optic atrophy include anterior ischemic optic neuropathy, optic neuritis, and compressive lesions of the optic nerve, including orbital (e.g. optic nerve sheath Meningioma), intracranial (e.g. olfactory groove Meningioma) or optic chiasmal (e.g. pituitary adenoma) tumors. Other ischemic, inflammatory, and compressive lesions can also produce primary optic atrophy.

Primary optic atrophy, in its severe from, is characterized fuduscopically by a pale optic disc with clearly delineated borders **(Fig 17)**. In some cases, the surface of the disc may appear waxy. The shaggy or gray fuzzy gliotic reaction characteristic of secondary optic atrophy is not seen overlying the disc or its margins. The absence of this gliotic reaction defines primary or simple optic atrophy and is the basis for its clinical differentiation.

The pink or rose colour of the disc is absent in primary optic atrophy **(Fig 17)** because of decreased blood perfusion, a direct manifestation of capillary drop out.

Optic atrophy may also be noted by a reduction in the number of ophthalmoscopically visible small blood vessels crossing the disc margin (Kestenbaum's capillary number test), from about 10 in normal nerves down to 7 or even fewer in atrophic nerves.

Optic disc atrophy may vary widely in appearance. The reduction of blood supply, disruption of axon columns, and formation of glial bridges may account for changes from slight temporal pallor of disc to a chalk-white optic nerve head. However, changes in optic disc colour may be difficult to describe or quantitate. Factors such as type of illumination, changes in the ocular media (e.g. cataract), and even weak ophthalmoscope batteries may influence the perception of optic disc coloration.

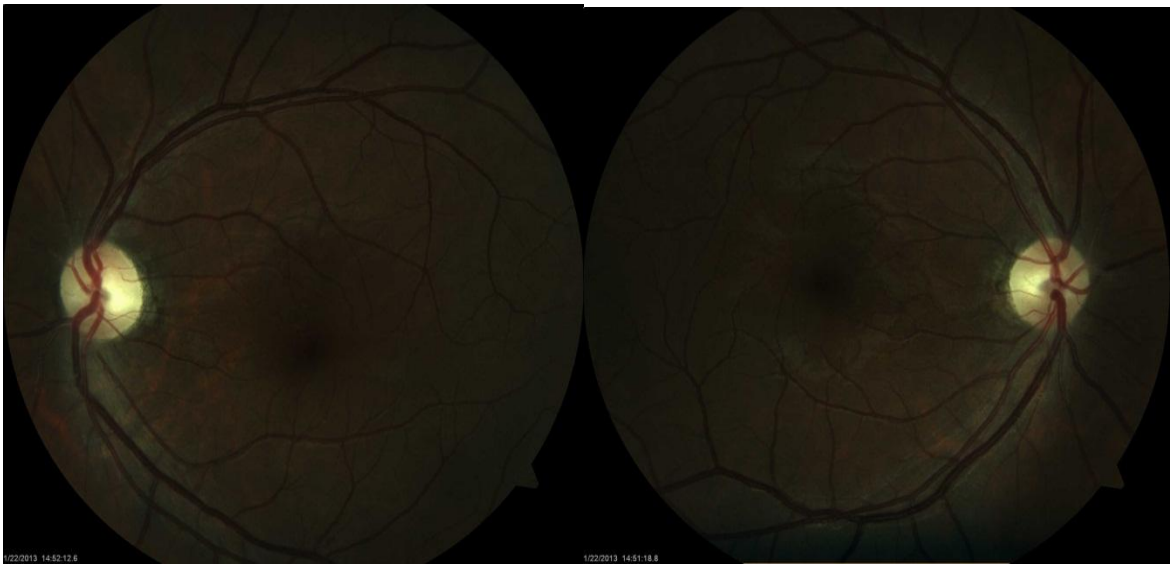

Fig:17 Primary optic atrophy both eyes, a case of chiasmal compression with bitemporal hemianopia

Secondary optic atrophy

The distinction between primary and secondary optic atrophy is important because the later is usually a consequence of severe disc edema. Secondary optic atrophy reflects the disorganized appearance of the surface of the optic disc seen if axonal injury occurs in association with severe edema or inflammation at the optic nerve head.

Although secondary optic atrophy most commonly occurs in cases of severe papilloedema, it may also develop in cases of long standing severe orbital inflammation. More often the observation of bilateral secondary optic atrophy provides an important clinical clue that suggests past episode of prolonged severe papilloedema. This finding is common in patients who have had undiagnosed or untreated pseudotumor cerebri(**Fig 19).**.

The fundus has a characteristic appearance in secondary optic atrophy. The optic disc is greyish instead of pink or white, and the margins appear fuzzy and blurred.**(Fig 18)** & (**Fig 19).** The glial proliferation may be sufficient to give a raised appearance to the optic disc and its margins. Other funduscopic features of secondary optic atrophy are similar to those in primary optic atrophy. These include the absence of the pink color of perfusion, a reduction in Kestenbaum's number, and a constriction of the vasculature**(fig 19 red arrows)**. However, because secondary optic atrophy is not characterized by a stark white optic disc, it may be misinterpreted as less severe that it really is.

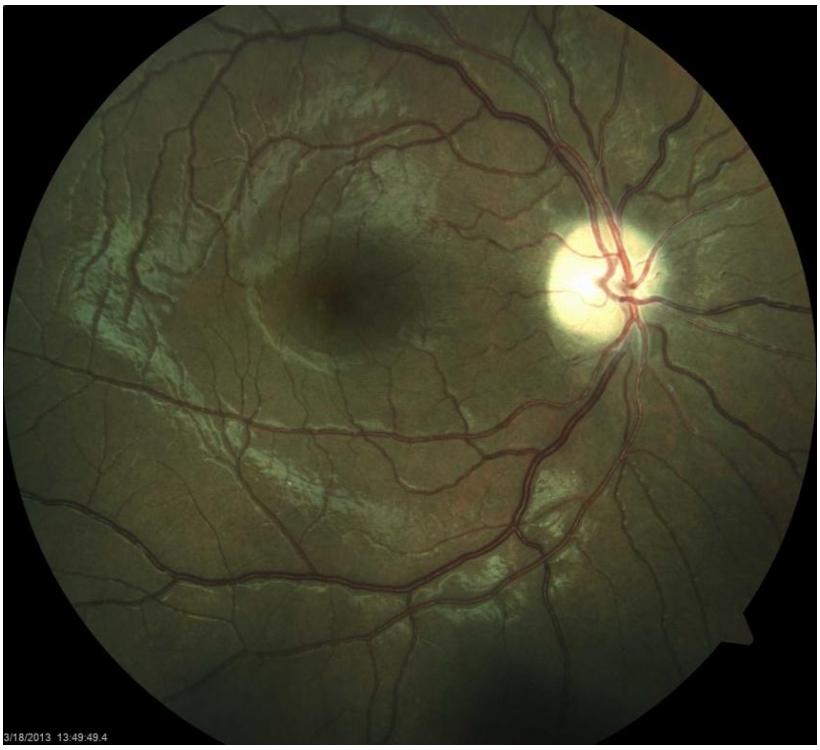

Fig 18 Secondary optic atrophy , disc margins blurred

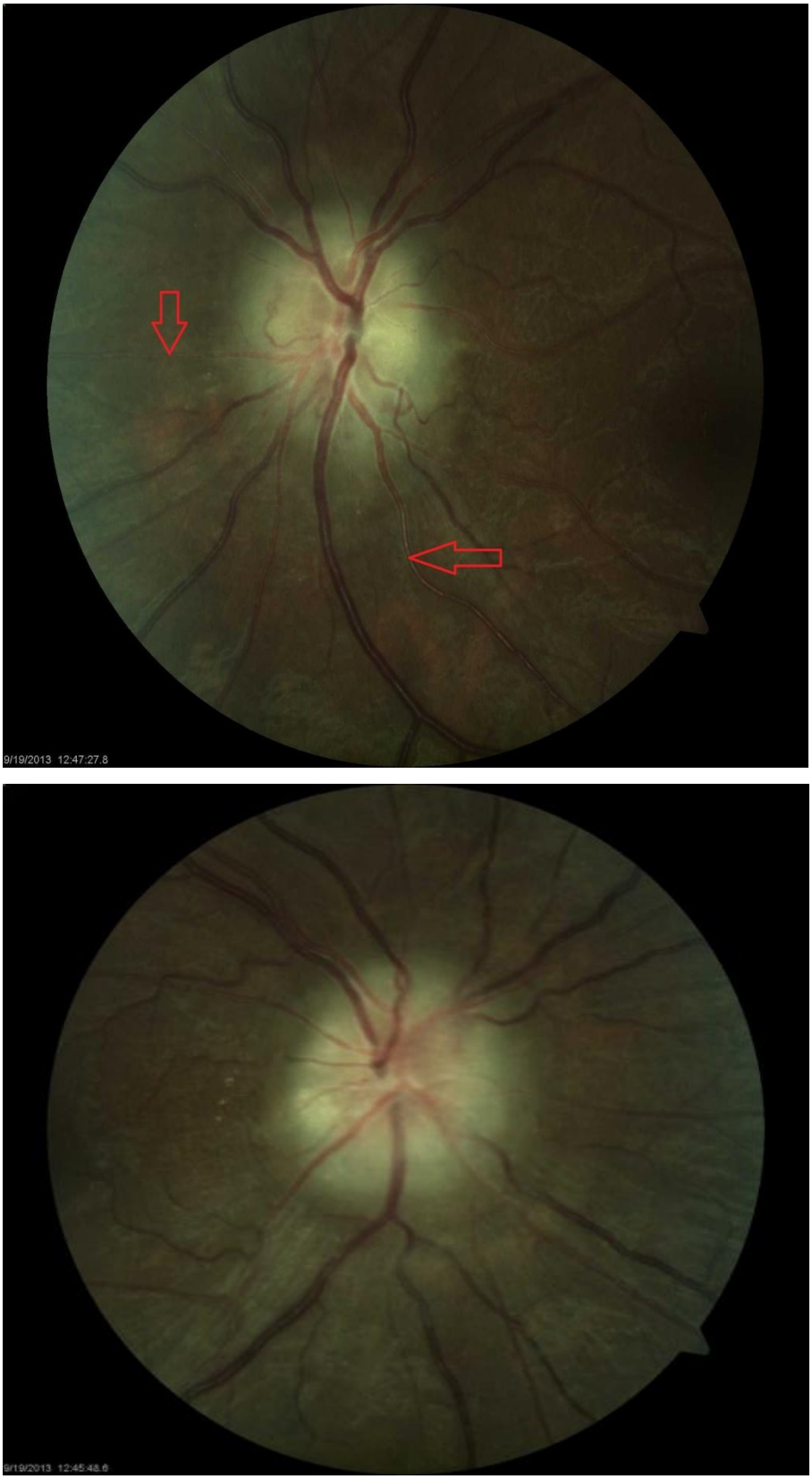

Fig 19 : Bilateral secondary optic atrophy

OCULAR MOVEMENT DEFICITS

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Eye movement examination evaluates integrity of the six levels of the ocular motor system (**Fig. 1**):

1. Upper motor neurons in the cortex, responsible for the voluntary saccadic eye movements;
2. Premotor neurons in the brainstem, which generate all saccadic eye movements.
3. Lower motor neurons, which activate of the extraocular muscles.
4. Cerebellar circuits, responsible for the calibration of smooth pursuit, saccadic, and vestibular-evoked eye movements and for appropriate visual-vestibular interaction.
5. Neurons of the vestibuloocular reflex arc, which are responsible for the rapid involuntary activation of the lower motor neurons in response to head movement.
6. Extraocular muscles themselves, are responsible for moving the eyes.

How to approach the eye movements

Eye movements are of two main types: gaze holding and gaze shifting. The term gaze refers to the direction of the line of sight in an earth fixed (not a head-fixed) frame of reference; thus gaze may remain constant if the eyes and head rotate in opposite directions by the same amount. Certain defects of eye movements, such as those made to remembered locations by patients with frontal lobe disorders, require laboratory testing. However, most disorders can be appreciated at the bedside, provided the examiner understands what properties are being tested.

Neurobiological basis for eye movements

Near their insertion, the extraocular muscles are surrounded by fibromuscular pulleys that guide their pulling directions and appear to dictate the geometric properties of eye rotations (Listing's law). The abducens nucleus is the horizontal conjugate gaze centre; it contains motoneurons that innervate the lateral rectus muscle and internuclear neurons that project across the midline, via the medial longitudinal fasciculus(MLF), to the contralateral medial

rectus motoneurons (Figure 1). Interruption of this pathway causes internuclear ophthalmoplegia (INO), with slowing of the adducting eye during horizontal saccades; this is an important sign in multiple sclerosis. The VOR for horizontal head rotations depends on vestibular afferents from the lateral semicircular canals, which relay their signal to the contralateral abducens nucleus via the medial vestibular nucleus (Figure 1). Wernicke's encephalopathy involves the vestibular nuclei and impairs the horizontal VOR. Command signals for horizontal saccades project to the abducens nucleus from the adjacent paramedian pontine reticular formation (PPRF); lesions here cause slow or absent horizontal saccades. Smooth-pursuit commands reach the abducens nucleus from the vestibulecerebellum; lesions of the flocculus and paraflocculus impair pursuit. The nucleus prepositus hypoglossi (NPH), medial vestibular nucleus (MVN) and the cerebellum play an important role in holding the eyes in an eccentric position (e.g., far right gaze) against the elastic pull of the orbital tissues; lesions of these structures cause the eyes to drift back to centre, leading to gaze-evoked nystagmus.

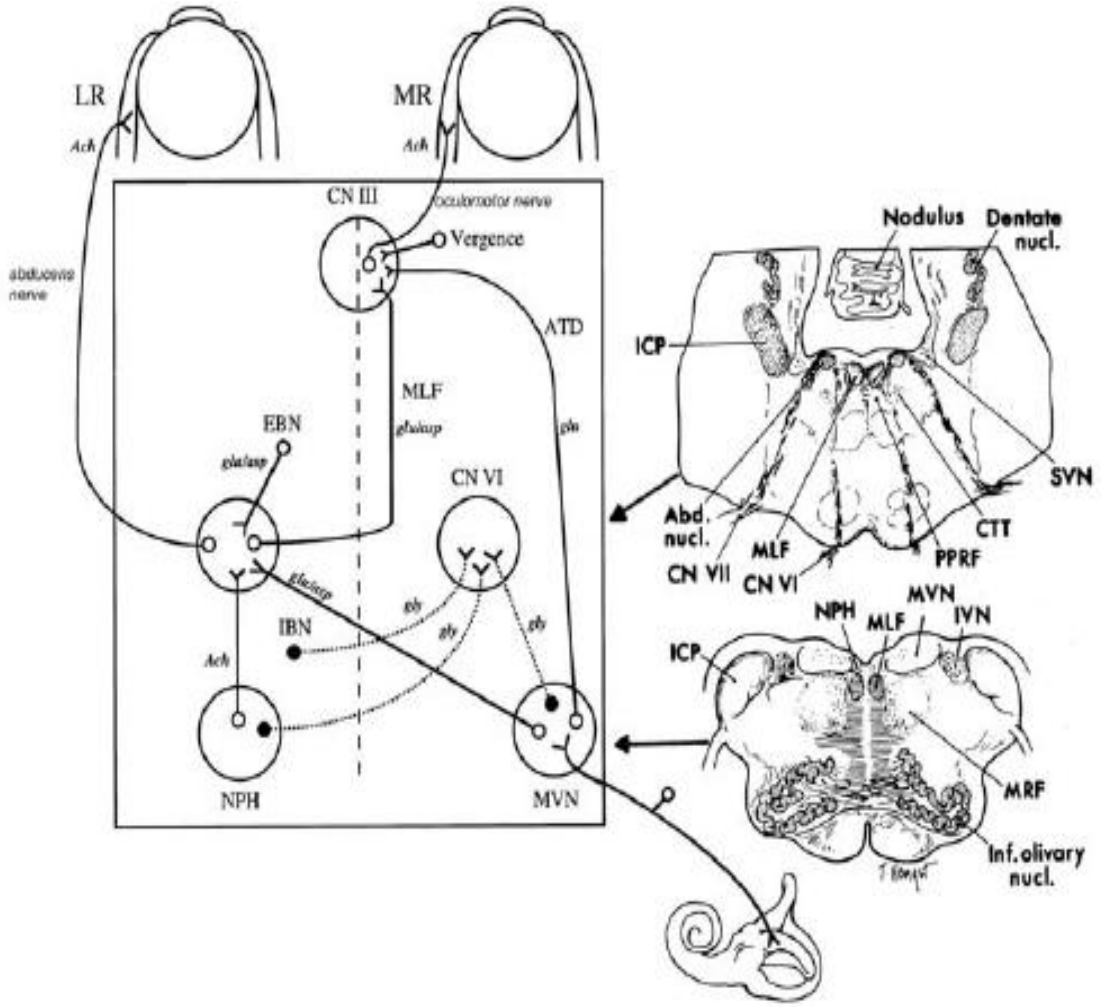


Figure 1. Anatomic scheme for the synthesis of signals for horizontal eye movements. The abducens nucleus (CN VI) contains abducens motoneurons that innervate the ipsilateral lateral rectus muscle (LR), and abducens internuclear neurons that send an ascending projection in the contralateral medial longitudinal fasciculus (MLF) to contact medial rectus (MR) motoneurons in the contralateral third nerve nucleus (CN III). From the horizontal semicircular canal, primary afferents on the vestibular nerve project mainly to the medial vestibular nucleus (MVN), where they synapse and then send an excitatory connection to the contralateral abducens nucleus and an inhibitory projection to the ipsilateral abducens nucleus. Saccadic inputs reach the abducens nucleus from ipsilateral excitatory burst neurons (EBN) and contralateral inhibitory burst neurons (IBN). Eye position information (the output of the neural integrator) reaches the abducens nucleus from neurons within the nucleus prepositus hypoglossi (NPH) and adjacent MVN. The medial rectus motoneurons in CN III also receive a command for vergence eye movements. Putative neurotransmitters for each pathway are shown: Ach: acetylcholine; asp: aspartate; glu: glutamate; gly: glycine. The anatomic sections on the right correspond to the level of the arrow heads on the schematic on the left. Abd. nucl.: abducens nucleus; CN VI: abducens nerve; CN VII: facial nerve; CTT: central tegmental tract; ICP: inferior cerebellar peduncle; IVN: inferior vestibular nucleus; Inf. olivary nucl.: inferior olivary nucleus; MVN: medial vestibular nucleus; MRF: medullary reticular formation; SVN: superior vestibular nucleus. (Reproduced, with permission from Leigh and Zee, 2006).[1]

The oculomotor and trochlear nuclei (**Fig 2**) house the motoneurons that innervate extraocular muscles that mainly rotate the eyes vertically (superior and inferior recti) or torsionally (around the line of sight – superior and inferior oblique muscles). These

motoneurons receive their saccadic input from burst neurons in the rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF), which lies in the pre-rubral fields of the rostral midbrain. Lesions involving the riMLF cause

slow or absent vertical saccades (such as in progressive supranuclear palsy, PSP). The signals for vertical vestibular and pursuit eye movements ascend from the medulla and pons to the midbrain in the MLF and other pathways. The interstitial nucleus of Cajal plays an important role in holding steady vertical eccentric gaze (eg: far upward gaze). The superior colliculus is a midbrain tectal structure that is

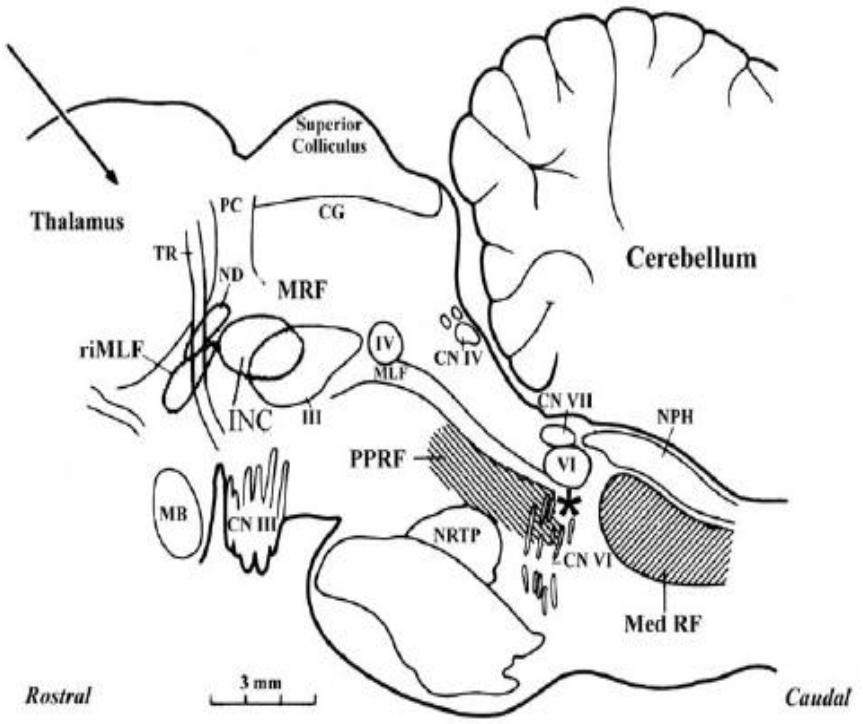


Figure 2: A sagittal section of the monkey brain stem showing the locations of premotor burst neurons: excitatory burst neurons for horizontal saccades lie in the paramedian pontine reticular formation (PPRF) and, for vertical and torsional saccades lie in the rostral interstitial nucleus of the medial longitudinal fasciculus (rostral iMLF). Burst neurons project to ocular motoneurons lying in the abducens nucleus (VI), the trochlear nucleus (IV) and the oculomotor nucleus (III). Omnipause neurons (indicated by an asterisk) lie in the midline raphe of the pons between the rootlets of the abducens nerve (CN VI) and gate the activity of burst neurons. CG: central gray; MB: mammillary body; MT: mammillothalamic tract; N III: rootlets of the oculomotor nerve; N IV: trochlear nerve; ND: nucleus of Darkschewitsch; NRTP: nucleus reticularis tegmenti pontis; PC: posterior commissure; NPH: nucleus prepositus hypoglossi; TR: tractus retroflexus; T: thalamus; Med RF: medullary reticular formation. The arrow refers to the Horsley-Clarke plane of section. (Figure adapted courtesy of Dr Jean Büttner-Ennever).

important for triggering both horizontal and vertical saccades; it receives inputs from frontal and parietal cortex. Two regions of the cerebellum contribute to the control of eye movements. The vestibulocerebellum (flocculus, paraflocculus, nodulus) are important for normal smooth pursuit (eye alone or eye-head tracking), eccentric gaze holding, and adjustment of the VOR so that it is optimised to guarantee clear vision. These latter functions are all impaired in patients with vestibulocerebellar lesions such as Chiari malformation; downbeat nystagmus is also often present. Lesions of the nodulus and adjacent ventral uvula cause periodic alternating nystagmus, a form of horizontal nystagmus that reverses direction every 2 minutes; it is suppressed with baclofen. The second cerebellar region, comprising the dorsal vermis and the fastigial nucleus to which it projects, is important for saccades to be

accurate. Thus, dorsal vermis lesions cause saccadic hypometria (undershoots), and fastigial nucleus lesions cause hypermetria (overshoots).

The cerebral cortex contains several areas that are important for eye movements (Figure 3). Primary visual cortex (V1) is the "royal gateway" for vision; without it, visually guided eye movements cannot be made (at least in humans). Secondary visual areas, such as the middle temporal visual area (MT, or V5), and the medial superior visual temporal area (MST) are essential or extracting information on the speed and direction of moving targets and subsequent programming of pursuit movements. The parietal eye field contributes to saccades in the context of shifts of the direction of attention. The frontal eye field is important for voluntary saccades and suppression of saccades during steady fixation.

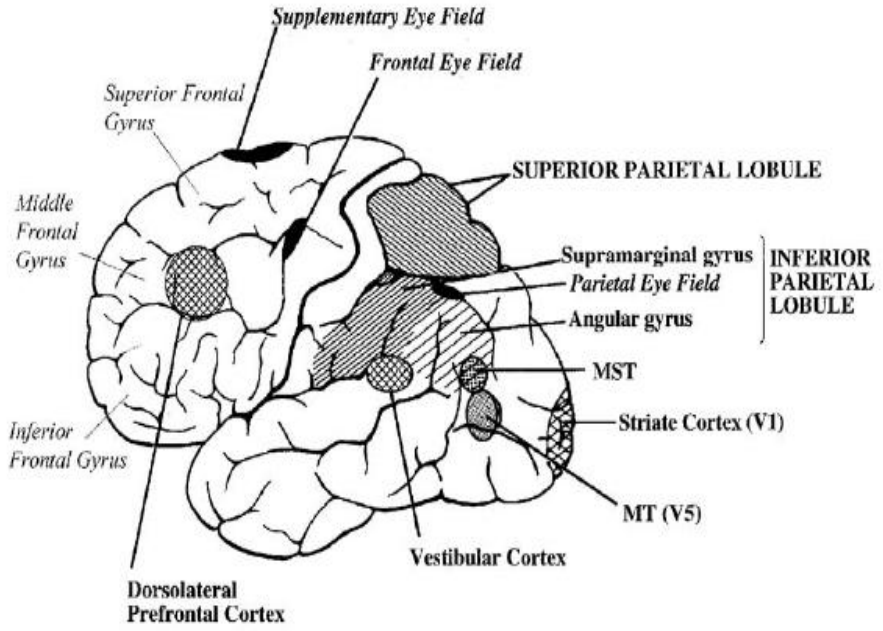


Figure 3: Probable location of cortical areas important for eye movements in human brain. MST: medial superior temporal visual area; MT: middle temporal visual area; these areas may form a contiguous cortical area. (Reproduced, with permission from Leigh and Zee, 2006).[1]

The supplementary eye fields, and adjacent pre-supplementary motor cortex, guide saccades during complex tasks, such as sequences of movements and responses when the instructional set changes. The dorsolateral prefrontal cortex is important for memory- guided saccades and programming saccades in the opposite direction (mirror image) to a visual stimulus (antisaccade). These cortical areas project to the superior colliculus and, via pontine nuclei to the cerebellum; direct projections to the PPRF or RIMLF are sparse, and there are no projections to the ocular motoneurons. The descending pathways to the superior colliculus

are both direct and also via the basal ganglia (caudate, substantia nigra pars reticulata, and subthalamic nucleus). Disease affecting the basal ganglia has subtle effects on eye movements, but seems concerned with behaviours that are rewarded.

EXAMINATION OF OCCULO MOTOR SYSTEM

The examination of the ocular motor system generally consists of the assessment of (a) fixation and gaze-holding ability, (*b*) range of monocular and binocular eye movements, (*c*) ocular alignment, and (*d*) performance of versions (saccades, pursuit). In addition, depending upon the findings of the basic examination, it may be appropriate to test the vestibulo-ocular and optokinetic reflexes and to attempt mechanically to move the eyes using forced duction testing.

FIXATION AND GAZE-HOLDING ABILITY

Principles

In an awake individual, the eyes are never absolutely still. Fixation is interrupted by three distinctive types of miniature eye movements: (a) microsaccades, with an average amplitude of about 6 minutes of arc and a mean frequency of about 2 per second; (*b*) continuous microdrift at rates of less than 20 minutes of arc/second; and (*c*) microtremor, consisting of high frequency (40–60 Hz) oscillations of 5–30 seconds of arc. Square wave jerks—spontaneous, horizontal saccades of about 0.5, followed about 200 msec later by a corrective saccade and occurring at a rate of less than 9 per minute—can also be observed during fixation in normal individuals.

When no efforts are being made toward ocular fixation or accommodation, the eyes are said to be in a “physiologic” position of rest. With total ophthalmoplegia, there is usually a slight divergence of the visual axes and this position usually also occurs during sleep, deep anesthesia, and death.

Technique

In patients complaining of intermittent diplopia, visual confusion, or strabismus, tests of sensory fusion (e.g., stereoacuity) and fixation should be performed before the eyes are dissociated by tests of monocular visual function (e.g., visual acuity, color vision, visual fields). The initial part of the ocular motor examination should consist of a careful study of fixation.

The patient should be instructed to focus on a distant target, and the eyes should be observed carefully. Asking the patient to describe the target can control attention. If strabismus is present, any

preference for fixation with one eye should be noted. Constant or intermittent monocular and binocular eye movements, whether conjugate or dissociated, should be noted. Subtle degrees of abnormal fixation can often be easily detected during the ophthalmoscopic examination.

RANGE OF EYE MOVEMENTS

Principles

The primary position of the eyes has been arbitrarily designated as that position from which all other ocular movements are initiated or measured. It was once assumed that all ocular motions occurred

around a fixed point in the orbit called the center of rotation. It has been shown, however, that there is no fixed center of rotation that does not move when the globe rotates and that the globe translates during every eye movement.

Thus, horizontal movements rotate the center of the globe in a semicircle in the plane of eye rotation, called the *space centroid*. All movements of the globe around the hypothetical center of rotation can be analyzed in terms of a coordinate system with three axes perpendicular to each other and intersecting at the center of rotation **(Fig. 18.1)**. These three axes, described by Fick in 1854, are called the *x*, *y*, and *z* axes of Fick. The *y* axis is equivalent to the visual axis; the *z* axis is vertical (around which the eye rotates horizontally); and the *x* axis is horizontal (around which the eye rotates vertically). These axes are stable with respect to a frontal plane, fixed in the skull, that corresponds roughly with the equatorial plane of the eye when it is directed straight ahead (Listing's plane).

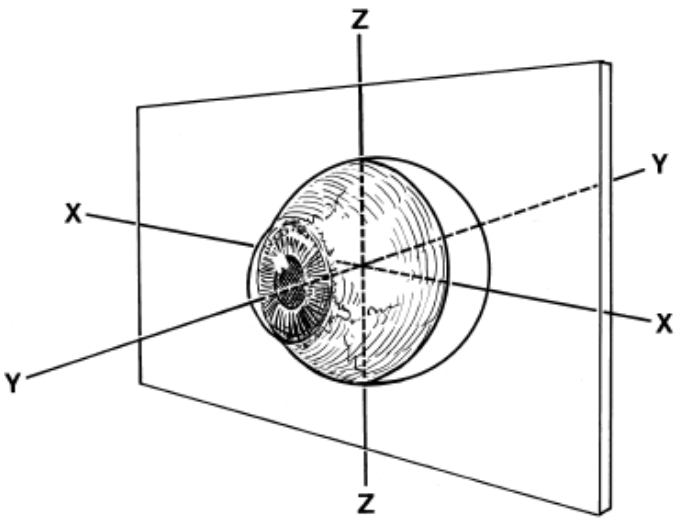


Figure 18.1. The axes of rotation of the eye. The *y* axis corresponds to the line of sight when the eye is in the primary position, looking straight ahead.

Rotations of both eyes (relative to one another) in the same direction are called *versions*. Rotations in opposite directions are called *vergence*. Only *convergence* (movement of the

eyes toward one another) in the horizontal plane is volitionally significant. *Divergence* amplitudes (movement of the eyes away from one another) are small in normal individuals. Rotations of either eye alone without attention to the movements of the other eye are called *ductions.* Horizontal rotation (rotation around the *z* axis of Fick) is termed *adduction* if the anterior pole of the eye is rotated nasally (i.e., inward, medially) and *abduction* if the anterior pole of the eye is rotated temporally (i.e., outward, laterally). Vertical rotation (around the *x* axis) is called *elevation* (or sursumduction) if the anterior pole of the eye rotates upward and *depression* (or deorsumduction) if it rotates downward.

Rotation during ductions or versions around either the horizontal or vertical axis places the eye in a secondary position of gaze. In achieving this position, there is no rotation of the globe around the *y* axis (i.e., there is no torsion).

Techniques

When testing the range of ocular movement, the examiner should ask the patient to follow a target through the full range of movement, including the cardinal (or diagnostic) positions of gaze. The eyes are tested individually with one eye covered (ductions) and together with both eyes open (versions). The normal range of movements is fairly stable throughout life for all directions except upgaze. Normal abduction is usually 50 degrees; adduction, 50 degrees; and depression, 45 degrees. Upward gaze decreases somewhat with advancing age. Thus, limitation of upward gaze in an older individual may simply be age-related and not necessarily a new, pathologic process. When the range of motion is limited, it is necessary to determine whether the limitation is mechanical and, if not,

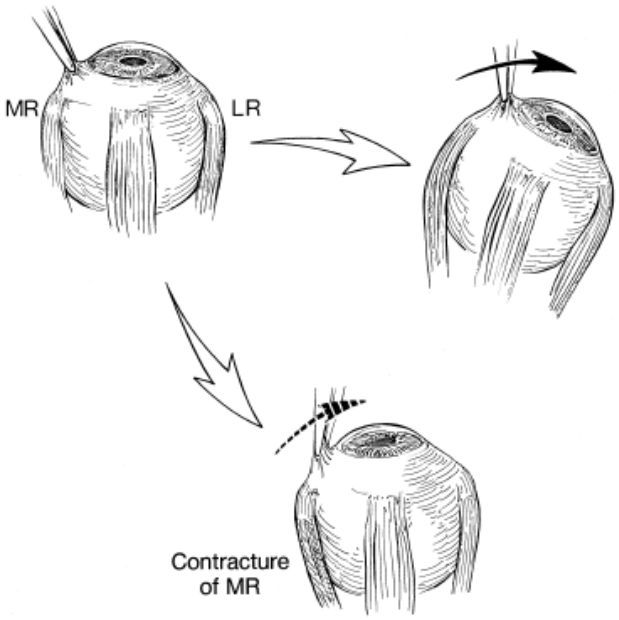


Figure 18.2. Forced duction testing. After the eye has been anesthetized with topical proparacaine and cocaine, the conjunctiva just posterior to the limbus is grasped with a fine-toothed forceps at a point opposite the direction of limitation. An attempt is then made to rotate the eye in the direction of limitation. If no mechanical limitation is present, the eye can be moved fully into the direction of limitation (*solid black arrow*). If mechanical limitation is present, the eye will resist attempts to rotate it into the field of limitation (*dashed black arrow*).

whether the disturbance is supranuclear or peripheral. Several tests may be used to determine whether a mechanical restriction of ocular motion is present. Mechanical limitation of motion (such as that seen in patients with thyroid ophthalmopathy or orbital floor fracture with entrapment) can be inferred if intraocular pressure increases substantially when the patient attempts to look in the direction of gaze limitation. The intraocular pressure measurements are most easily performed using a Tonopen or a pneumatic tonometer.

Mechanical limitation of motion can more reliably be detected with forced duction (or traction) testing. In such tests, an attempt is made to move the eye forcibly in the direction(s) of gaze limitation (Fig. 18.2).

In addition to the forced duction test, mechanical determination of muscle force can be used to assess the function of apparently paretic muscles with contracture of their antagonists. An estimate of active muscle force present in patients with limitation of ocular motility can be made by stabilizing

the anesthetized eye with a toothed forceps in a position near the limbus on the side of the limitation while the eye attempts to look into the field of the limitation (**Fig. 18.4**).

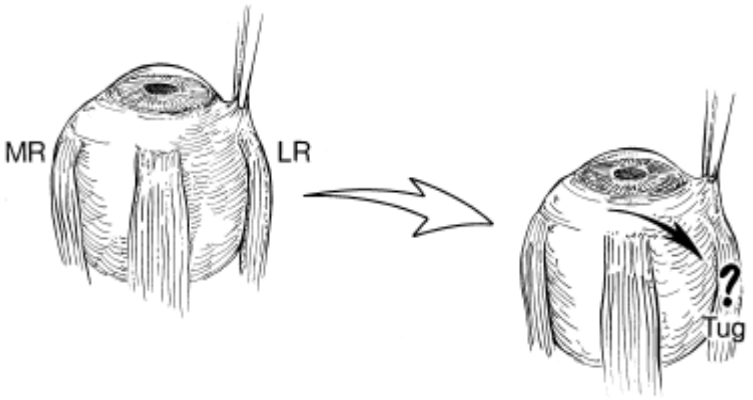


Figure 18.4. Estimation of active, generated muscle force. After the eye has been anesthetized with topical proparacaine and cocaine, the conjunctiva just posterior to the limbus is grasped with a fine-toothed forceps on the side of the limitation. The examiner then holds the eye while the patient attempts to look in the direction of limitation. If there is an intact nerve supply to the muscle that could move the eye into the field of limitation, the examiner will feel a tug on the forceps.

The presence of a tug on the forceps indicates that a contraction of the suspected paralytic muscle has occurred. The results of this ‘‘forced generation test’’ can even be quantified.

Nonrestrictive limitation of eye movements may occur from disease of supranuclear or infranuclear structures. Since the workup and management of the patient will vary considerably depending on the location of the lesion, supranuclear disorders must be distinguished from infranuclear disorders. From a practical standpoint, supranuclear disorders that cause abnormalities in the range of eye movements usually result from lesions of the cerebral hemispheres or the brain stem premotor structures.

In the oculocephalic test, the awake patient is asked to fixate a target straight ahead while the head (or the entire body) is rotated from side to side and up and down. A normal response consists of a conjugate eye deviation in the direction away from head or body rotation such that the eyes remain stable with respect to space despite the head movement. Asking the patient to read a Snellen chart during head or body rotation can demonstrate the remarkable integrity of this VOR. In patients with intact vestibular systems, there is no degradation of visual acuity, even with rotations of up to 40/sec. Patients with vestibular disease have a rapid decline in this dynamic visual acuity with head rotation. To perform the oculocephalic test in comatose patients, the eyelids are simply held open and the rotational head movements are performed. In unconscious patients, oculocephalic testing may be the most useful method of assessing eye movements. The VOR is often intact in such patients, whereas saccadic and pursuit eye movements are absent. Thus, rapid horizontal rotation of the head results in deviation of the eyes away from the direction of the head turn. The eyes then make an exponential drift back to primary position if the head rotation is maintained. Though not saccadic, this recentration of the eyes may be quite rapid, occurring with a time constant of less than 0.5 seconds in the most severe vegetative states. Normal responses during oculocephalic testing indicate that the nuclear and infranuclear ocular motor structures are intact and capable of being stimulated by an intact vestibular system. This test can also be used in patients with functional (nonorganic) limitation of gaze to show that a full range of eye movement can be elicited despite apparent gaze restriction during testing of voluntary eye movements.

In some patients with paresis of upward gaze, Bell's phenomenon may be helpful in differentiating an infranuclear from a supranuclear lesion. Bell's phenomenon consists of outward and upward rolling of the eyes when forcible efforts are made to close the eyelids against resistance. It does not occur with blinks, and it is observed in only 50% of individuals during voluntary unrestrained lid closure. The presence of this movement in individuals who cannot voluntarily elevate their eyes usually indicates that brain stem pathways between the facial nerve nucleus and that portion of the oculomotor nucleus responsible for ocular elevation are intact, and thus that an upward gaze paresis is supranuclear in origin. However, an intact Bell's phenomenon may occur in patients with Guillain-Barre´ syndrome and was also demonstrated in a patient with complete ophthalmoplegia caused by myasthenia gravis. Absence of a Bell's phenomenon has less diagnostic usefulness, since about 10% of normal subjects do not have this fascio-ocular movement. A downward Bell's response is present in up to 8% of individuals.

OCULAR ALIGNMENT

Principles

When the eyes are not aligned on the same object, *strabismus* is present. The strabismus may be congenital or acquired and may be caused by central or peripheral dysfunction. In some individuals, particularly those with isolated congenital strabismus, the amount of ocular misalignment is unchanged regardless of the direction of gaze or of which eye is fixating the target. This type of strabismus is termed **comitant or concomitant**. On the other hand, when the amount of an ocular deviation changes in various directions of gaze, with either eye fixing, or both, the strabismus is said to be **incomitant or noncomitant**. Congenital comitant strabismus is occasionally associated with other neurologic dysfunction, and acquired comitant strabismus may rarely occur as a sign of intracranial disease Most cases of acquired comitant strabismus appear in otherwise normal children and adults, as well as in persons with neurologic or systemic disease, from decompensation of a pre-existing phoria, or as a result of latent hypermetropia. Thus, most instances of neuropathic or myopathic strabismus are of the incomitant variety.

Primary and Secondary Deviations

Any patient with a manifest deviation of one eye (heterotropia) will fixate a target with only one eye at a time. During viewing with one eye, the visual axis of the opposite (nonfixing) eye will be deviated a certain amount away from the target. Patients with a comitant strabismus have the same amount of deviation of the non-fixing eye regardless of the eye that is fixing or the field of gaze. Most patients with in comitant (and especially paralytic) strabismus tend to fix with the non-paretic eye if visual acuity is equal in the two eyes. In these patients, the deviation of the non-fixing eye is called the primary deviation. When such patients are forced to fix the same target with the paretic eye, the deviation that results, the secondary deviation, is always greater than the primary deviation. The explanation for this phenomenon is related to the position of the eyes within the orbits. When a single muscle is paretic, the deviation between the two eyes is proportional to the difference between the forces generated by the paretic muscle and its normal yoke muscle. Furthermore, the amount of force contributed by each muscle toward holding the eye in a specific orbital position increases as the eye is moved into the direction of action of that muscle. Under normal circumstances, this force, thus obeying Hering's law, is equal for yoke pairs of muscles. However, as the eyes move into the direction of action of the paretic muscle, the difference in

forces generated by the normal and paretic yoke muscles increases, thus increasing the deviation between the two eyes. When this change in deviation is tested as a function of orbital position, it is actually found to be independent of which eye is fixing. Thus, when the paretic eye is fixating a target, it is held in an orbital position further in the direction of action of the paretic muscle than when the non-paretic eye is fixating the same target. This results in a secondary deviation that is greater than the primary deviation simply because of the change in eye position toward the direction of action of the paretic muscle when the paretic eye is forced to take up fixation.

Past-pointing and Disturbances of Egocentric Localization

Von Graefe first described anomalies of spatial localization that he referred to as *past-pointing* or *false orientation* in patients with paralytic strabismus. If a patient is asked to point at an object in the field of action of a paretic muscle while the paretic eye is fixating, the patient's finger will point beyond the object *toward* the field of action of the paretic muscle (**Fig. 18.6**). During this test, it is important that the hand be covered or that the patient point rapidly toward the object so as to avoid visual correction of the error of localization while the hand is still moving toward the object. Patients with accommodative esotropia exhibit a similar phenomenon while they have a manifest esotropia, pointing in the direction that the nonfixating eye is looking.

The explanation of past-pointing is controversial. One explanation is that the image of the

Figure 18.6. Past-pointing in a patient with a right sixth nerve paresis. *A,* In primary position, there is only a slight amount of past-pointing. *B,* In right gaze, however, the amount of past-pointing increases. The *white arrows* indicate the amount of past-pointing (the difference between the actual target location and the area in space to which the patient points).

target lies in an abnormal location relative to the fovea so that the patient incorrectly localizes the object into that field. This explanation, however, does not account for the past-pointing that is observed when the image of the target lies on the fovea of the paretic eye. Clearly non-visual information about eye position is involved in spatial localization, and an argument has raged over the relative role of efferent command to the eye muscles versus proprioceptive afferent information from the eye muscles. In support of the efferent command theory, Helmholtz argued that past-pointing depends on the ‘‘intensity of the effort of will’’ that is sent to the paretic muscle.

Head Turns and Tilts

Patients with strabismus commonly turn or tilt the head to minimize diplopia. Head turns are frequently associated with paresis of the horizontal extraocular muscles. Similarly, patients with vertical extraocular muscle paresis may carry their head flexed or extended. Most patients with such head turns adopt the particular posture to minimize or eliminate diplopia by moving the eyes away from the field of action of the paretic muscle; however, some patients adopt a head posture that actually increases the distance between the two images, allowing one of the images to be more easily ignored. Head turns also occur in patients with congenital nystagmus. In such patients, keeping the eyes in an eccentric (null) position in the orbit by means of the head turn may result in reduction in the amplitude or frequency of the nystagmus. Head tilts are most commonly observed with paresis of the oblique muscles, particularly the superior oblique. With an acquired superior oblique palsy, for example, the face is usually turned away from the paretic eye, the chin is down slightly, and the head is tilted toward the side opposite the paretic muscle. This permits fusion of images. Patients with congenital superior oblique palsy may adopt a similar head tilt or one in the opposite direction (i.e., toward the side of the paretic muscle) to more widely separate the images. Head tilts that occur from ocular causes often must be differentiated from nonocular torticollis. Finally, some patients develop head turns that seem to be caused by central visual field defects and not by ocular misalignment. Such patients turn their heads toward the hemianopic field under both monocular and binocular conditions. The explanation for the head turn in these patients is unclear.

Techniques

Ocular alignment may be tested subjectively or objectively, depending on the circumstances under which the examination is performed and the physical and mental state of the patient.

Subjective Testing

When a patient is cooperative, subjective testing of diplopia reliably indicates the disparity between retinal images. The simplest subjective tests of ocular alignment use colored filters to dissociate the deviation and to emphasize and differentiate the images so that the patient and the observer can interpret them. A fixation light is used to provide the image. A red filter held over one eye suffices in most cases. However, the addition of a green filter over the opposite eye gives better results in children, in patients with a tendency to suppress or ignore one of the images, and in patients with a slight paresis and good fusion ability who can overcome their deviation. The use of complementary colored filters, one over each eye, produces maximum dissociation of images, since there is no part of the visible spectrum

common to both eyes. The red filter is always placed over the patient's right eye, and all questions posed to the patient relate to the relationship of the red image with respect to the white (or green) image. The patient is first asked if he or she sees one or two lights. If the patient sees two lights, he or she is then asked what color they are. After the appropriate answer, the patient is asked if the red light is to the right or left of the other light and if it is above or below the other light. The information thus received will be that the patient sees two lights, one red and one white or green, that they are crossed (each image is perceived on the side opposite the eye that is seeing it) or uncrossed (each image is perceived on the same side as the eye that is seeing it), and that the image relating to the right eye (red image) is higher or lower than the other image. The image of an object is always displaced in the *opposite* direction to the position of the eye (Fig. 18.7). Thus, if an eye is exotropic, the patient will have *crossed* diplopia, and (with a red filter over the right eye) the patient will see the red image to the *left* of the other image. Similarly, if the patient has an esotropia, the red image will be seen to the *right* of the other image (*uncrossed* diplopia). If the patient has a vertical deviation of the eyes, the eye that is higher will see the image of an object *below* that of the opposite eye.

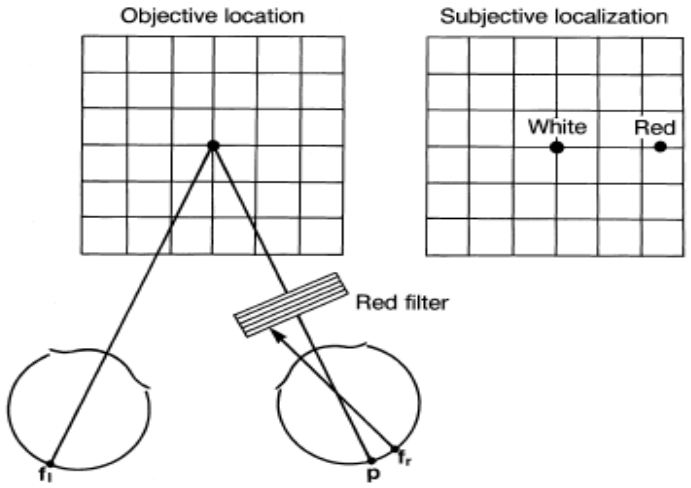


Figure 18.7. The principle of diplopia tests. A red filter is placed in front of the right eye, and the patient fixes a single light in the distance. If the eyes are misaligned, the light is imaged on the fovea of one eye (fl) and the nonfoveal retina (p) of the opposite eye. The patient thus sees two images, white and red, in different locations in space.

Once the patient indicates that there is a clear separation of images when he or she is fixing on a light held straight ahead, the examiner can determine the area of maximum vertical separation, horizontal separation, or both, by having the patient look at a light held in the eight other cardinal positions of gaze (right, upper right, up, upper left, left, lower left, down, lower right). In addition to the use of filters placed over one or both eyes, one can place a red Maddox rod over one eye and have the patient fixate on a white light. The ''rod'' is, in fact, a set of small half-cylinders aligned side by side in a frame in such a way that when the eye views a light through the cylinders, the image seen is that of a line perpendicular to the cylinder axis. Thus, if one views a white light with one eye covered by a red Maddox rod, the

images will be those of a red line and a white light. The Maddox rod can be placed in such a

manner as to produce a vertical, horizontal, or oblique line. Individuals who are orthophoric

will see the line pass through the light. When the rod is oriented to produce the image of a

vertical line, patients with a horizontal strabismus will see the line to the left or right of the

light. When the rod is oriented so that a horizontal line image is produced, patients with a vertical strabismus will see the line above or below the light. Torsional misalignment of the eyes (e.g., superior oblique palsy) can be tested with two Maddox rods, one over each eye. This is best performed using a trial lens frame. If both rods are oriented so as to produce a horizontal line image, an eye with torsional dysfunction will see the line as oblique rather than horizontal. The patient is then asked to rotate the rod until the line is perceived as horizontal. By this method, the amount of torsion can be measured and followed. Traditionally, one clear (or white) Maddox rod and one red Maddox rod are used for this test. However, Simons et al. showed that the subjective torsion more reliably localizes to the paretic eye if two red Maddox rods are used and if the room is dark during testing. In addition to the tests described above, other subjective techniques may be used in which two test objects rather than one are presented to the patient in such a way that each object is viewed with only one eye. The patient is then required to place the two objects in such a fashion that they appear to be superimposed. The objects appear superimposed only when their images fall on the fovea of each eye. Misalignment of the foveas results in the patient placing the objects in different locations in space. The eyes are differentiated and

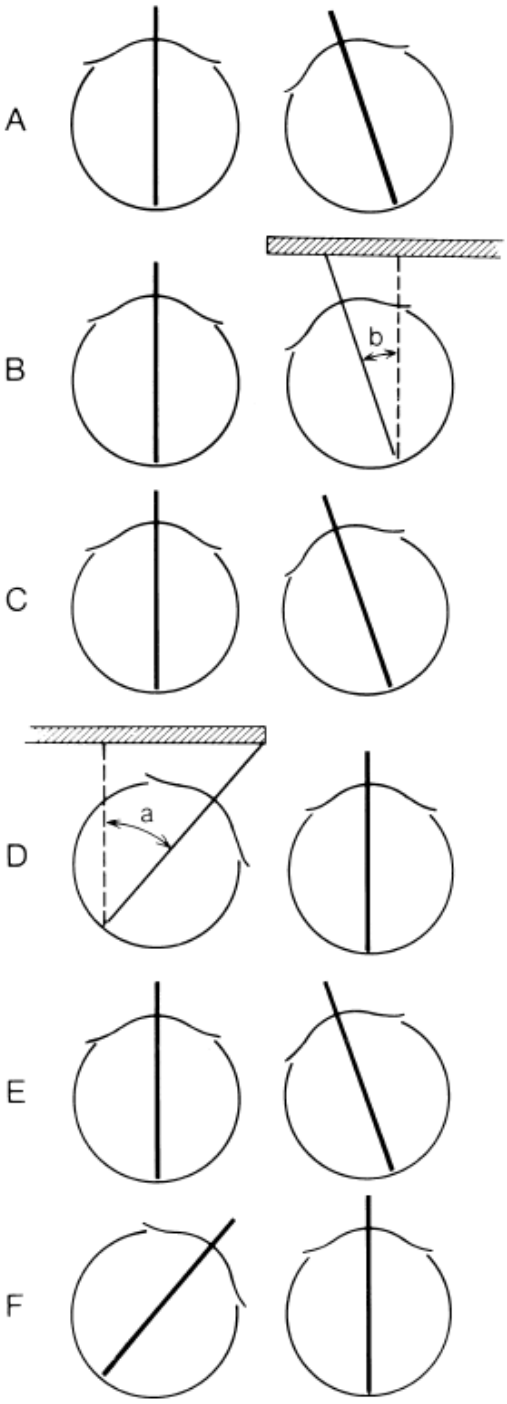


Figure 18.10. The single cover and cover/uncover tests. In both tests, one eye is covered at a time; however, in the single cover test, one eye is covered and the opposite eye is observed. In the cover/uncover test, one eye is covered and the behavior of that eye is observed when the cover is removed. *A*, Initially, with both eyes viewing, the left eye is fixating the target and the right eye is esotropic. When the right eye is covered (*B*), no movement of the left (uncovered) eye is observed, nor is any movement of the right eye observed when the cover is removed (*C*). *D*, When the left eye is covered, the right eye moves outward to take up fixation. The deviation of the normal eye under cover (the secondary deviation, a) is greater than that of the paretic eye under cover (primary deviation, b). When the cover is removed, either the left eye again takes up fixation (E) or the paretic right eye continues to fixate (F).

dissociated in various ways. Each eye may be presented with a different target, or complementary colors may be placed into the visual field, either directly or by projection, with each eye being provided with a corresponding colored filter. These haploscopic tests include the use of a major amblyoscope and the Hess screen and Lancasterred-green tests.

Objective Testing

The simplest objective method of determining ocular alignment is the use of a hand light to cast a reflection on the corneal surfaces of both eyes in the cardinal positions of gaze. If the images from the two corneas appear centered, then the visual axes are often correctly aligned. If the light reflexes are not centered, one can either estimate the amount of misalignment based on the apparent amount of decentration of the light reflex (with the fixation light held 33 cm from the patient, 1 mm of decentration equals 7 degrees of ocular deviation), or prisms can be placed over either of the eyes until the light reflexes appear centered.

Other ocular abnormalities that produce decentration of the corneal light reflex include eccentric fixation and ectopic macula (e.g., in patients with retinopathy of prematurity or other retinal disease with macular traction).

The most precise objective methods of measuring ocular alignment are the cover tests. Although these tests require that the patient be able to fixate a target with either eye, they generally require less cooperation than do the subjective tests described above. The three types of cover tests used by most clinicians are the single cover test, the cover/uncover test, and the alternate cover (cross-cover) test. In the single cover test, the patient fixates an accommodative target at 33 cm (near target) or 6 meters (distant target). An opaque occluder is placed in front of one eye, and the examiner observes the opposite eye to see whether it moves to take up fixation of the target (Fig. 18.10). If movement is observed, its direction and speed should be noted. The test is then repeated on the opposite eye. If the patient has a manifest ocular deviation (heterotropia), the previously nonfixing eye will be observed to change position to take up fixation when the fixing eye is covered. On the other hand, when the nonfixing eye is covered, no movement of the fixing eye will be observed (since it is already fixing on the target). This test is usually performed with the patient fixing in primary position and with the eyes in the other cardinal positions of gaze. In our experience, movements of as little as 1 degree can be easily observed.

In the cover/uncover test, the patient fixates on an accommodative target, and one eye is occluded. The behavior of that eye is then observed as the cover is removed (Fig. 18.10). The direction of any deviation and the speed and rate of recovery to binocular fixation are noted. If no movement of the uncovered eye is observed when the cover/uncover test is performed, an alternate cover test may be used to detect a latent deviation of the eyes (heterophoria). Instead of occluding one eye and then taking the occluder away, first one eye and then the other are alternately occluded. The cover should remain in front of each eye long enough to allow the patient to take up fixation with the uncovered eye. This test prevents fusion and dissociates the visual axes. Any movement of either uncovered eye suggests that although the eyes are straight during binocular viewing, loss of fusion (i.e., by the alternate occlusion of the two eyes) results in a deviation of which ever eye is covered (**Fig. 18.11**). The importance of distinguishing between heterophorias and heterotropias cannot be overemphasized, since patients with heterophorias have binocular central fusion, whereas patients with heterotropias do not. If either the cover/uncover or alternate cover test detects evidence of ocular misalignment, prisms can be used to neutralize the movement and thereby measure the deviation, whether it is a heterotropia or heterophoria. When there is a vertical ocular deviation, it is often helpful to perform cover tests with the head tilted first toward one side and then toward the other. The patient is instructed to maintain fixation on a distant target, and the position of the eyes relative to each other is measured. The patient is then instructed to tilt the head to one side while maintaining fixation on the target, and the eyes are again examined to see whether one eye has moved higher than

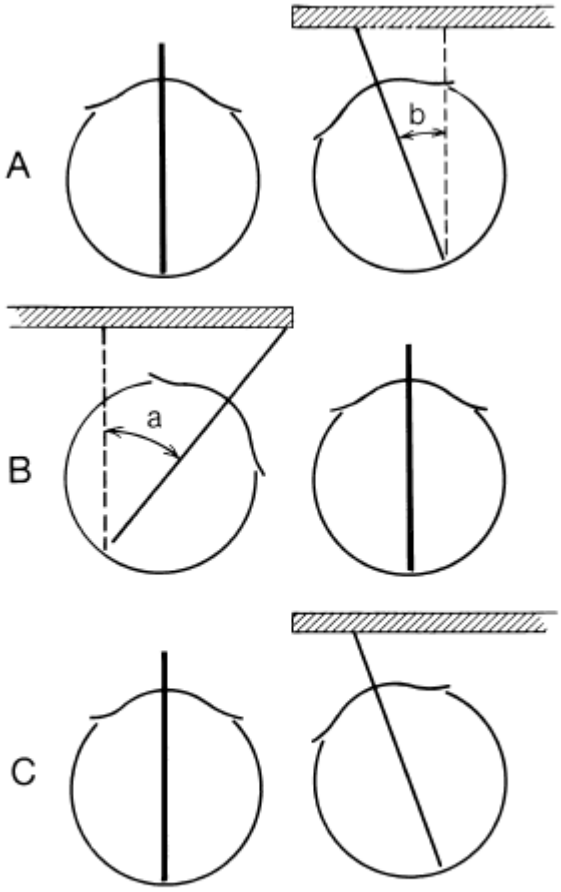


Figure 18.11. The alternate cover test. This test prevents fusional vergence and thus tests both phorias and tropias but does not differentiate between them. In this test, the cover is quickly moved from one eye to the other, and any movement of either eye is noted. In this example, there is an esodeviation.

the other. Measurements are made with the head tilted to each side. This test is useful primarily in the diagnosis of superior oblique palsy, since patients with this disorder consistently show a hypertropia of the damaged eye when the head is tilted toward the affected side. When a patient with a trochlear nerve palsy tilts the head toward the affected side, intorsion of that eye should occur to keep the vertical meridian perpendicular to the horizon. As noted above, this intorsion is usually about 10 degrees and is produced by the otolith-ocular reflex, resulting in synergistic contractions of the superior rectus and superior oblique muscles. If, however, the superior oblique muscle is paretic, its secondary actions, one of which is depression, are also impaired. The superior rectus muscle is therefore the only means by which the eye is intorted, and its main action, elevation of the eye, is unopposed.

PERFORMANCE OF VERSIONS

Versions may be tested by examining the saccadic, pursuit, vestibular, and optokinetic systems.

Clinical Examination of Saccades

Saccadic eye movements are examined clinically by instructing the patient to alternately fixate upon two targets, usually the examiner's finger and nose. Saccades in each direction can be examined in each field of gaze in both the horizontal and vertical planes.

Saccadic latencies can be appreciated by noting the time it takes the patient to initiate the saccade. Abnormal voluntary saccadic velocities may be accentuated by using a drum or hand-held tape with repetitive patterns. Rotating the drum or passing the tape horizontally and vertically across the patient's visual field stimulates the optokinetic system to produce nystagmus.

Abnormal saccadic velocities, particularly slowing, thus become more evident when the patient is forced to make multiple, repetitive saccades to refixate the passing targets. Altered saccadic velocities that occur in only one plane of movement can also be appreciated by using obliquely placed targets to stimulate oblique saccades. In such patients, the normal-velocity component of the saccade (e.g., the horizontal) will be completed before the other component (e.g., the vertical), so that the trajectory appears L-shaped. Disorders of saccadic accuracy (e.g., saccadic dysmetria) can be inferred from the direction and size of corrective saccades that the patient must make to ultimately acquire the fixation target. When a saccadic

abnormality is detected during the clinical examination, the examiner must attempt to localize the disturbance within the hierarchical organization of the saccadic eye movement system. The first step in localization is to establish whether the disease process affects reflexive saccades. Next, an attempt should be made to determine whether saccades can be performed without visual targets or in response to auditory targets, by asking the patient to refixate under closed lids. The eye movements thus generated can be observed, palpated. During the evaluation of saccadic eye movements, it is often helpful to observe gaze changes when the patient makes a combined eye/head movement to see whether an accompanying head movement can facilitate the production of a saccade. This strategy is used by some patients with ocular motor apraxia. Finally, asking the patient to repetitively refixate between two targets may reveal fatigue of saccadic eye movements.

Clinical Examination of Pursuit

Patients with isolated deficiency of smooth pursuit do not usually complain of visual symptoms, since they can track moving objects with a series of saccades. Only very demanding tracking tasks (e.g., playing tennis, handball, or baseball) may cause patients with impaired pursuit to report difficulties. The vision of normal subjects deteriorates during tracking of targets moving at high frequencies, however, so that even complaints of inability to track fast-moving objects may not signify a disorder of smooth pursuit. To test the pursuit system, the patient should be asked to track a small target, such as a pencil tip held a meter or more from the eyes, with the head held still. The target should initially be moved at a low, uniform velocity. Pursuit movements that do not match the target velocity result in corrective saccades. If these are ‘‘catch-up’’ saccades, then the pursuit gain is low. If pursuit gain is too high, then ‘‘backup’’ saccades are observed. Small children, uncooperative patients, or individuals thought to have nonorganic blindness may be tested with a slowly rotating large mirror held before their eyes. The VOR generates eye movements that compensate for angular displacement of the head and maintain the visual axes ‘‘on target.’’ If one observes a slowly moving target moving the head, so that the target remains stationary relative to the head, the eye movements generated by the VOR are inappropriate and must be suppressed. The ability of a patient to suppress (or cancel) the VOR can be evaluated by using a central fixation target that moves in the same direction and at the same velocity as the head. Patients often do this best by fixating on their thumbnail with their arm outstretched while being rotated in the examination chair. Those who have limb muscle weakness can be rotated in a

wheelchair while fixating a target that rotates with the chair. When suppression of the VOR and pursuit are compared in normal human subjects, similar frequency response curves are obtained, leading to the hypothesis that suppression of the VOR depends directly on information derived from the smooth pursuit system. This hypothesis is supported by the clinical observation that patients with impaired smooth pursuit also have abnormal suppression of the VOR.

The evaluation of VOR suppression is thus another way to test the integrity of the pursuit system. Deficits in VOR suppression, however, are nonlocalizing, since they may occur with either cerebral or cerebellar disease. In some patients, it is difficult to test smooth pursuit because of spontaneous nystagmus; however, in some of these patients, the nystagmus is less prominent in a specific position of gaze. In these patients, cancellation of the VOR during head rotation can be tested with the eyes fixing on a target in this position. As with pursuit, the head rotation should be gentle at first. In patients with inadequate cancellation, the eyes are continually taken off target by the intact VOR, and corrective saccades therefore occur. An asymmetric deficit may imply a pursuit imbalance provided that the VOR is intact and symmetric: deficient cancellation of the VOR on rotation to one side corresponds to a low pursuit gain to that side. Furthermore, when there is a discrepancy between the performance of smooth pursuit and cancellation of the VOR (e.g., poor pursuit but good cancellation), one should suspect an inadequate or asymmetric VOR.

QUANTITATIVE ANALYSIS OF EYE MOVEMENTS

Voluntary Eye Movements

Most disturbances of ocular motility and alignment can be detected during a standard clinical examination; however, performing a quantitative analysis of eye movements may more accurately reveal subtle abnormalities of the pursuit, saccadic, optokinetic, and vestibulo-ocular systems. The most common methods used to record eye movements are electro-oculography and infrared oculography.

These techniques may be used to distinguish myopathic (restrictive) from neuropathic conditions that affect ocular motility and to determine the presence or absence of improvement of ocular motor function. The value of vertical saccadic velocity or amplitude determinations in adduction versus abduction to identify and monitor superior oblique muscle dysfunction remains controversial.

Although electro-oculography can yield reasonable recordings of horizontal eye movements, vertical measurements with this technique are affected by eyelid artifacts and nonlinearities. Changes in illumination and skin resistance also affect the readings with this method.

Vestibulo-Ocular Reflex

Rotation tests give more accurate and reproducible results, although the mental state of the patient while in darkness may influence the results. The gain of the VOR may be obtained by measuring the peak eye velocity in response to a velocity step (e.g., sudden sustained rotation at 60 degrees/sec) in darkness. This is usually done in vestibular laboratories equipped with servo-controlled chairs and eye monitoring equipment, although portable systems for this purpose are available.

Optokinetic System

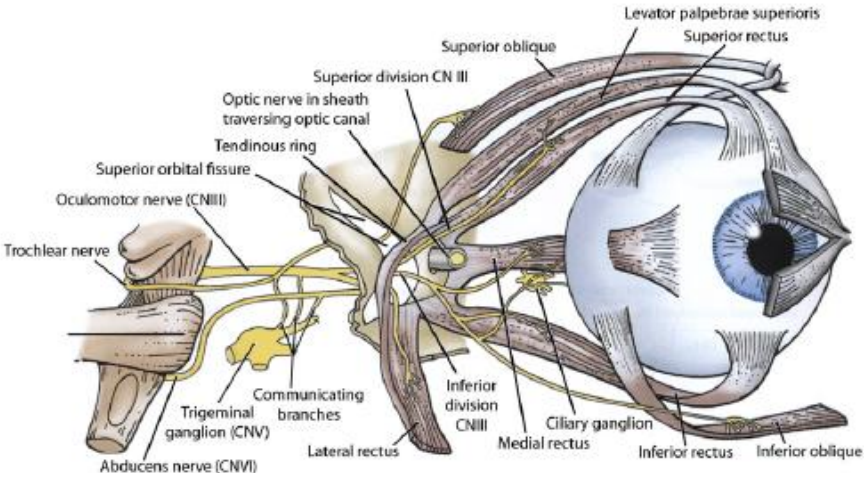
The hand-held ‘‘optokinetic’’ drums or tapes that are used to elicit smooth movements primarily test the pursuit system. True optokinetic testing requires a stimulus that fills the field of vision. A common technique is to have the patient sit inside a large, patterned optokinetic drum that is rotated around the patient. A true optokinetic stimulus induces a sensation of self-rotation. Another method of eliciting a true optokinetic response is rotation of an individual at a constant velocity in the light for over 1 minute. The sustained nystagmus that results is caused by purely visual stimuli.

ASSESSMENT OF THIRD, FOURTH & SIXTH CRANIAL NERVES

Anatomy

The ocular motor nerves (cranial nerves 3, 4, and 6), innervate the 6 extraocular muscles of each eye and cause movements of the eye (Fig. 1). The oculomotor (third) nerve innervates the medial rectus, inferior rectus, superior rectus, and inferior oblique muscles, as well as the levator palpebrae. The trochlear (fourth) nerve innervates the superior oblique muscle, and the abducens (sixth) nerve innervates the lateral rectus muscle.

The nuclear complex of the third nerve lies in the dorsal midbrain, anterior to the cerebral aqueduct. It consists of multiple subnuclei that give rise to distinct sets of fibers destined for the muscles targeted by the third nerve. In general, the axons arising from these subnuclei travel in the ipsilateral nerve, except axons arising from the superior rectus subnucleus that travel through the contralateral third nerve complex to join the third nerve on that side.



In addition, a single central caudate nucleus issues fibers that join both third nerves to innervate the levator palpebrae muscles bilaterally. The preganglionic cholinergic fibers that innervate the papillary constrictor arise from the paired Edinger-Westphal nuclei. The third nerve fascicles travel then ventrally, traversing the red nucleus and the cerebral peduncles, before exiting the midbrain into the interpeduncular fossa. The proximal portion of the nerve passes between the superior cerebellar and posterior cerebral arteries. The axons are topographically arranged, with fibers for the inferior rectus, medial rectus, superior rectus, and inferior oblique arranged along the medial-to-lateral axis. Pupillary fibers are generally located superficially, in the superior and medial portion of the nerve.

The trochlear (fourth) nucleus is situated in the pontomesencephalic junction, ventral to the cerebral aqueduct. The axons exit the brainstem dorsally, then decussate within the anterior medullary velum and innervate the contralateral superior oblique muscle. The abducens (sixth) nucleus lies in the dorsal pons. The sixth nerve fascicle travels ventrally, through the corticospinal tracts, before exiting anterolaterally at the pontomedullary junction. Although the third and fourth nerves are situated along the lateral wall of the cavernous sinus, the abducens nerve has a more medial position, just lateral to the internal carotid artery. All 3 ocular motor nerves exit the cavernous sinus via the superior orbital fissure, and then pass through the orbital apex to reach their target muscles. The blood supply to the third, fourth, and sixth nerves have multiple sources that feed a vasa nervorum capillary network.

Third Nerve Palsy

Clinical Features

Complete, isolated third nerve palsy causes ipsilateral weakness of elevation, depression, and adduction of the globe, in combination with ptosis and mydriasis. Depending on the specific cause, complete third nerve palsy may involve the pupil (causing mydriasis) or spare the pupil (Figs**. 3 and 4**). In partial third nerve palsy, different patterns of impaired motility may occur with or without pupillary involvement. The motility deficit may be subtle, and a reduced duction may not be easily observed. In this case, more detailed assessment of alignment, with alternate cover or Maddox rod testing, willshow an incomitant pattern of ocular misalignment supporting the diagnosis of partial third nerve palsy. A characteristic feature is that the affected eye is hypotropic in upgaze but hypertropic in downgaze, because of the combined weakness of the superior and inferior rectus muscles.

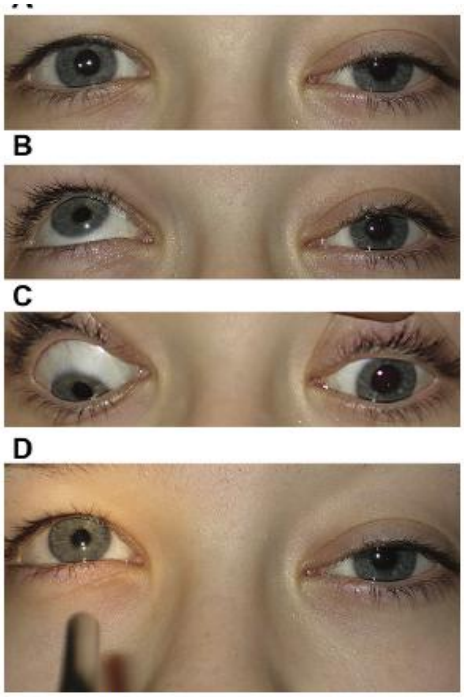


Fig. 3. A 32-year-old woman with traumatic complete left third nerve palsy, showing right hypertropia in upgaze that becomes left hypertropia in downgaze. (*A*) Left ptosis, mydriasis, exotropia, and right hypertropia in primary gaze. (*B*) Absent left elevation. (*C*) Reduced left depression. (*D*) The left pupil shows minimal consensual response to light, with greater anisocoria.

As opposed to lesions of the third nerve fascicle or nerve, a lesion of the third nerve nucleus will cause bilateral abnormalities. Specifically, there is bilateral ptosis (because the central caudal nucleus supplies both levator palpebrae muscles) and a bilateral elevation deficit (because the superior rectus subnucleus sends fibers through the contralateral third nerve nucleus to join the opposite nerve). Therefore, the classic clinical picture of unilateral nuclear third nerve palsy is ipsilateral mydriasis; ipsilateral weakness of the medial rectus, inferior rectus, and inferior oblique muscles; bilateral ptosis; and bilateral superior rectus weakness.

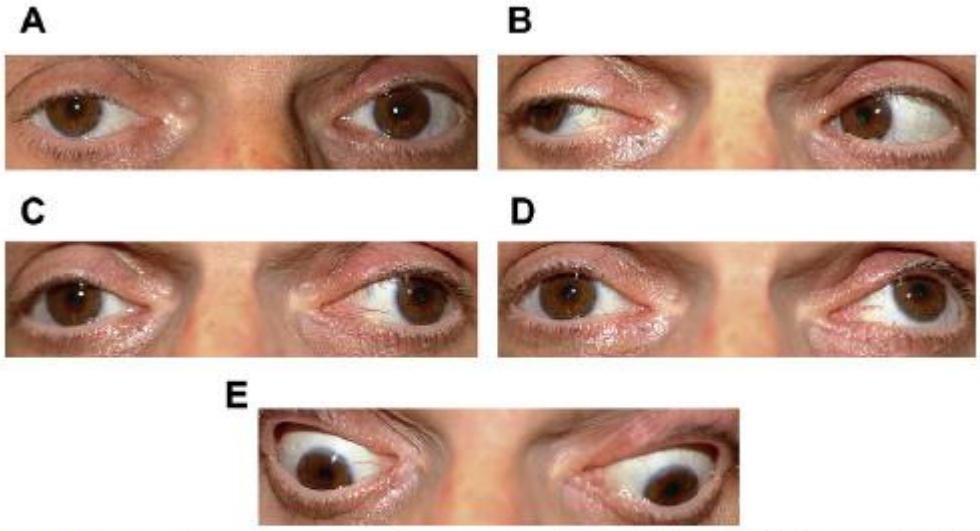


Fig. 4. A 55-year-old woman with a microvasculopathic partial right third nerve palsy due to diabetes and hypertension. Magnetic resonance imaging and magnetic resonance angiography were normal. (*A*) Right ptosis without mydriasis in primary position. Mild physiologic anisocoria was present. (*B*) Normal right gaze. (*C*) Decreased right adduction on left gaze. (*D*) Decreased right elevation on upgaze. (*E*) Decreased right depression on downgaze. The motility became normal within 8 weeks.

The appropriate workup for a patient with third nerve palsy depends on the patient's age and pupil function. In adults with acquired, isolated, complete, or partial third nerve palsy that involves the pupil, there is no controversy to the workup: these patients need urgent imaging to exclude a PComm aneurysm or other mass. Computed tomography angiography (CTA) and magnetic resonance angiography are useful, but the exact sensitivity and vailability of these tests vary across institutions. Nevertheless, if these tests are negative, a catheter angiogram often remains necessary in these patients because small aneurysms are potentially missed on non invasive imaging studies.

Fourth Nerve Palsy

Clinical Features

Fourth nerve palsy presents with vertical diplopia and is commonly accompanied by compensatory contralateral head tilt. Identification of a fourth nerve palsy in a patient with vertical diplopia involves application of the Parks-Bielchowsky three-step test. First, hypertropia suggests weakness of the ipsilateral superior oblique, ipsilateral inferior rectus, contralateral inferior oblique, or contralateral superior rectus muscle.Second, increased hypertropia in contralateral gaze narrows the possibilities to the weakness of the ipsilateral superior oblique or contralateral superior rectus muscles. Third, increased hypertropia on

ipsilateral head tilt further reduces the possibilities, ultimately identifying ipsilateral superior oblique weakness.

Although the abnormal ductions may be detected by direct observation, in many cases, patients with vertical misalignment to have no visible impairment in ocular motility (Fig. 10). Therefore, assessment of alignment using alternate cover or Maddox rod testing can be particularly useful to show the characteristic pattern of impaired motility. The reason that hypertropia is exacerbated in contralateral gaze is that superior oblique palsy causes weakness of depression in adduction (in long-standing cases, the hypertropia in adduction is further enhanced by overaction of the ipsilateral inferior oblique). The reason hypertropia is worse with ipsilateral head tilt is that the ocular

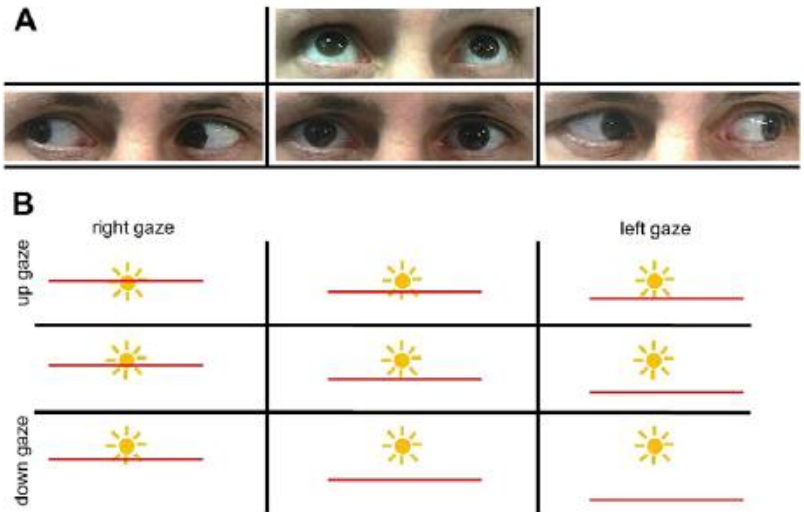


Fig. 10. A 36-year-old man with right fourth nerve palsy following resection of cerebellar hemangioblastoma. (*A*) Essentially normal ductions, with small right hypertropia in primary gaze and upgaze, increased in left gaze. (*B*) Simulation of patient's view through Maddox rod in each direction of gaze. Note greatest vertical separation in down-and-left gaze. (*C*) Pre- and

counterroll reflex stimulates ipsilateral intorters (superior oblique and superior rectus) and contralateral extorters (inferior oblique and inferior rectus); when the superior oblique is weak, this reflex causes a compensatory increase in ipsilateral superior rectus action, resulting in additional hypertropia (because the superior rectus is an elevator).

Torsional diplopia, which results from ocular cyclotorsion, often accompanies vertical diplopia in acquired fourth nerve palsy. This condition can be quickly assessed by having the

patient view a horizontal straight line, such as the edge of a door. A patient with cyclotorsion from unilateral fourth nerve palsy will see a horizontal line and a second tilted line above or below it, intersecting on the side of the affected eye. Cyclotorsion can also be evaluated with the double Maddox rod, which refracts a light source into one red line (seen by the right eye) and one white line (seen by the left eye). The degree of relative cyclotorsion is measured by rotating the filters until the subject reports that the lines are parallel.

Bilateral fourth nerve palsy, which most commonly results from trauma, is characterized by a unique constellation of findings. Primary position vertical alignment may be fairly good because of the canceling effect from bilateral palsies. Esotropia may be present, making the initial diagnosis difficult by potentially suggesting sixth nerve palsies. However, with careful examination, bilateral fourth nerve palsies are readily identified. First, hyperdeviation alternates such that it is contralateral to the direction of gaze and ipsilateral to the side of head tilt. Second, there is esotropia greatest in downgaze (so-called V-pattern esotropia, with >15 prism diopters difference between upgaze and downgaze) because of weakened abduction in depression (the superior oblique acts as an abductor). Third, there is often a large angle of excyclotorsion (>10 degrees), accompanied by prominent torsional diplopia. Rarely, bilateral congenital fourth nerve palsy may occur (Fig. 11).

Identifying fourth nerve palsy in the setting of concomitant third nerve palsy can be difficult, because the failure of adduction prevents complete testing of superior oblique function. In this setting, the superior oblique can be evaluated by

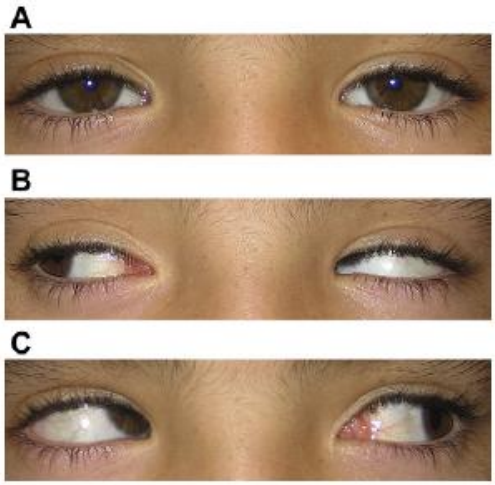


Fig. 11. A 7-year-old girl with bilateral congenital fourth nerve palsy. Brain MRI was normal. (*A*) Normal alignment in primary gaze. (*B*) Left hypertropia in right gaze, with left inferior oblique overaction. (*C*) Right hypertropia in left gaze, with right inferior oblique overaction.

assessing its secondary function: intorsion of the abducted eye on attempted downgaze. The torsional movement that indicates intact superior oblique function is best appreciated by observing a conjunctival vessel.

Sixth Nerve Palsy

Clinical Features

Weakness of the lateral rectus due to sixth nerve palsy leads to horizontal diplopia, worse to the affected side and at distance. Often, the abnormal duction is easily observed, but in subtle cases, an incomitant esotropia must be shown by testing binocular alignment. Many patients with acute sixth nerve palsy require MRI to exclude structural, inflammatory, or neoplastic causes. All children and young adults with sixth nerve palsy should undergo imaging because of a higher prevalence of tumors and demyelinating lesions. However, in a patient in whom a microvasculopathic cause is strongly considered, observation for spontaneous improvement over several weeks may prevent the need for imaging. Additional workup in selected patients with sixth nerve palsy may include serologies and CSF analysis.

Combined Third, Fourth, and Sixth Nerve Palsy

Diseases of the subarachnoid space that may affect multiple cranial nerves include infectious, inflammatory, and neoplastic processes. Infectious processes include viral, fungal, or bacterial infection (including tuberculosis, syphilis, and Lyme disease). Inflammatory diseases include sarcoidosis and idiopathic pachymeningitis.

Neoplastic processes include carcinomatous and lymphomatous meningitis. Peripheral demyelinating disorders including Guillain-Barre syndrome, the Miller Fisher variant, chronic inflammatory demyelinating polyneuropathy, and idiopathic cranial neuropathies (Fig. 18) are other considerations when multiple ocular motor nerves are involved. Patients with myasthenia gravis may present with complex patterns of limited eye movements that closely mimic cranial nerve dysfunction, but these patients are often distinguished by the presence of normal pupillary responses.

Expanding masses at the base of the skull can cause compression of multiple ocular motor nerves. One consideration is meningioma of the sphenoid wing (causing ophthalmoplegia, proptosis, and hyperostosis of the temporal bone) or clivus. Other rare possibilities are chordoma, which may arise in the region of the clivus from remnants of the embryologic notochord, or chondrosarcoma, which arises from cartilage in bone.

A process involving the cavernous sinus may affect any combination of the third, fourth, or sixth nerves and cause dysfunction of the first and second divisions of the trigeminal nerve. The differential diagnosis of a superior orbital fissure process and a cavernous sinus process is similar, with the main clinical distinction that the second division of the trigeminal nerve is spared in the former. The differential diagnosis for these syndromes includes neoplastic, infectious, inflammatory, and vascular diseases.

Neoplastic considerations include meningiomas, lymphoma, pituitary adenoma, metastases,trigeminalneuromas, chordomas, chondrosarcomas, and nasopharyngeal carcinomas. Although the slow growth of apituitary adenoma makes it less likely to involve the ocular motor nerves, the rapid onset of headache and ophthalmoplegia strongly suggests pituitary apoplexy. The third nerve is the most commonly affected by apoplexy, followed by the sixth nerve and lastly the fourth nerve.

Infectious considerations include herpes zoster ophthalmicus, may lead to ophthalmoplegia on the basis of secondary vasculitis or direct inflammation of the ocular motor nerves. Vascular lesions of the cavernous sinus include aneurysms and carotid cavernous fistulas CCF). Carotid aneurysms may present with pain and diplopia due to involvement of any of the ocular motor nerves, most frequently the sixth nerve. CCFs are characterized as being high flow (direct) or low flow (indirect) based on the source of the feeder vessel and the rate of flow.

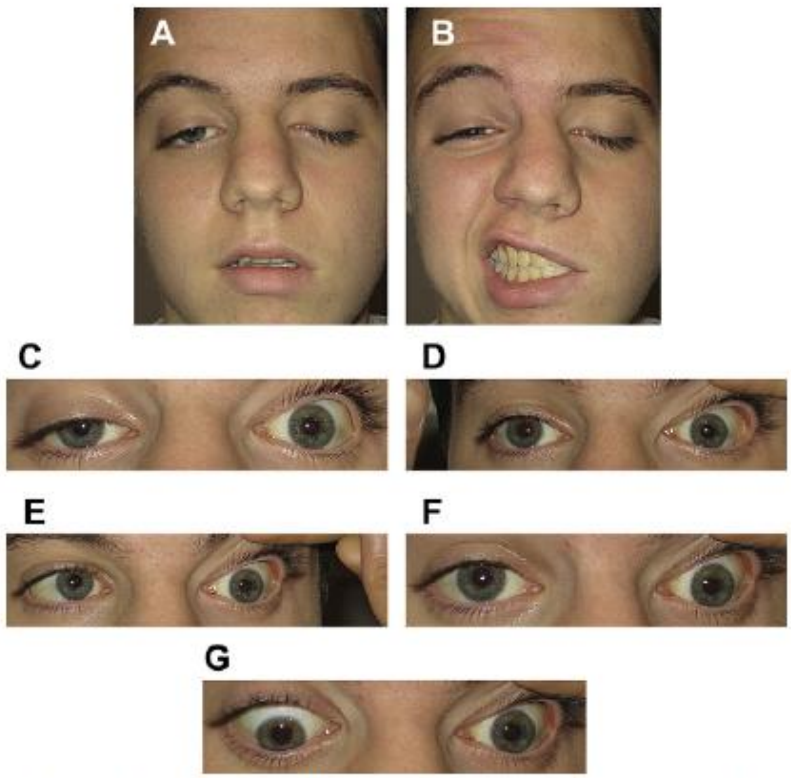


Fig. 18. A 15-year-old boy with multiple idiopathic cranial neuropathies who presented with 3 days of diplopia and facial weakness. There was hyporeflexia and mild ataxia in the arms. Brain MRI was normal and spinal fluid revealed 1 leukocyte and normal protein

Sections of the content are adapted from:

1. Principles and Techniques of the Examination of Ocular Motility and Alignment. *Mark S. Borchert.* Text Book*:* Clinical Neuro-Ophthalmology.
2. Paralytic Strabismus: Third, Fourth, and Sixth Nerve Palsy. Sashank Prasad, Nicholas J. Volpe.
3. Neuroscience of Eye Movements. Visual Neuroscience. ACNR, Vol 5 No 6 • Jan/Feb 2006

Supranuclear control of eye movements

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Supranuclear movement

- Saccades
- Pursuit
- Vergence
- Vestibulo-ocular movement
- Optokinetic Nystagmus

Saccades

- Are conjugate rapid eye movements that quickly redirect the eyes so that the image of the object falls on the fovea.
- These are random eye movements, eye movements on command, refixation eye movement, fast phase of OKN (optokinetic Nystagmus) & vestibular Nystagmus.
- They are present at birth, velocity of saccades varies from 30 – 700°/sec.
- Latency of saccades is 200 – 250 m sec (interval between appearance of target and the onset of a saccade).
- Not affected by drugs.
- They are examined clinically (a) by asking patient to look right left, up and down – saccade on command (b) ask patient to look at the examiner's nose and then to the fingers held at 30°.
- Observe latency and accuracy of the saccades.
- Latency time taken for generation of the eye movements.
- Accuracy of saccades
    - When eye movements fall short of the target hypometric saccade.
    - When eye movements overshoot the target hypermetric saccade.

Pathway for saccades

- Saccades are generated at FEF (Frontal eye field) and superior colliculus.
- FEF (area 8) is the posterior end of the middle frontal gyrus.
- FEF receives input from the peristriate, parietal, superior temporal cortex and thalamus prefrontal cortex and supplementary eye field.
- Fibres from FEF descend through internal capsule, reticular formation adjacent to cerebral peduncles and end in the PPRF (paramedian pontine reticular formation) and riMLF (rostral interstitial nucleus of medial longitudinal fasciculus). Decussation of these fibres occurs in midbrain.
- At cerebral level, the saccade control is contraversive i.e. right FEF stimulation causes saccadic eye movement to left side and vice versa.
- Deeper layers of superior colliculus also control saccade on the contralateral side.
- Superior colliculus receives visual input from retina and visual cortex, from frontal and parietal cortex, substantia nigra (via thalamus).
- Superior colliculus sends output to thalamus, PPRF and riMLF.

Parietal lobes

- Contains neurons that discharge in relation to saccades performed to pay visual attention to a peripheral target.

Cerebellum

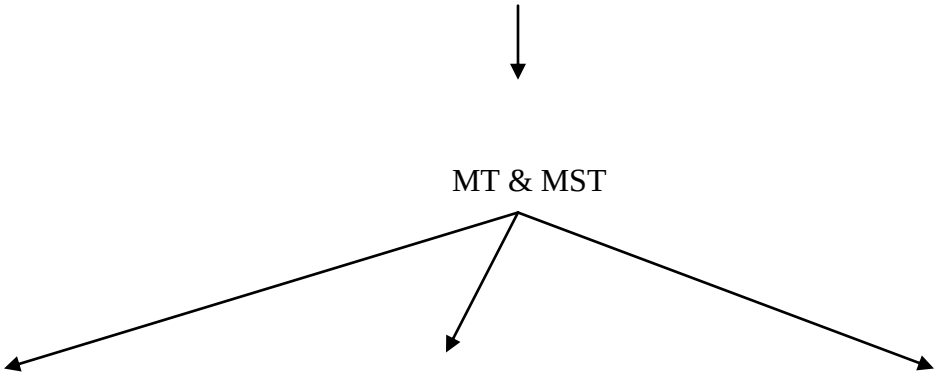
- Is also involved in the control of saccadic eye movement, Vermis, fastigial nucleus, floccules are important for various aspect of saccadic eye movement control like amplitude and gaze holding.

- Dorsal vermis and fastigial nuclei are important for saccadic amplitude while floccules, paraflocculles play role in gaze holding mechanism or stabilize the image on retina.

Pursuit eye movements

- Are slow, continuous, conjugate eye movements that occur when a fixated object is moved slowly.
- Pursuit is accurate and smooth up to 30°/sec.
- If the object is moved more rapidly, smooth pursuit breaks down and is replaced by multiple saccades of small amplitude (saccadic pursuit).
- The pursuit eye movements are not present at birth and appear around 6 weeks of age.
- They have latency of 120 msec.
- They are sensitive to drugs.
- Primary areas of pursuit generation are MT and MST (middle temporal and medial superior temporal).

Visual input from areas 18 & 19 (Para and peristriate areas)



FEF Posterior parietal cortex Brain stem reticular formation

Connections from MT and MST

in internal sagittal striatum

Corona radiata

Thalamus

PPRF and RiMLF (Brain stem)

- Fibres connecting to PPRF and riMLF undergo double decussation, hence right hemisphere controls pursuit to right side and left hemisphere controls pursuit to left side.

Cerebellum

- Flocculus play role in controlling smooth pursuit movement.
- Floccules have connections with vestibular nuclei, perihypoglossal nuclei, PPRF.

Vergence eye movements

- Are dysconjugate eye movements (convergence, divergence).
- Slow, 20°/sec, latency of 160msc.
- The hemisphere control center for Vergence is located in striate and perisriate cortex and parietal cortex.
- Fibres from above cortical areas terminate in the pretectal area which is ventral in the tegmentum of midbrain.
- There are two brainstem centers for Vergence movement.
- Convergence and divergence cells are found intermixed in the mesencephalic reticular formation with 1-2 mm of the oculomotor nucleus.
- Cerebellum also functions in the production of vergence movements.

Vestibular eye movements

- Stabilize gaze during head rotation so that the image can be held steady on the retina.
- The sensory organs for the vestibular eye movements are the semicircular canals, utricle and saccule.
- For the horizontal eye movements, the right horizontal semicircular canal drives the eyes to the left, while left semicircular canal drives the eyes to the right.
- VOR have short latency period, attain speed of up to 300/sec.

- They can be tested clinically by caloric stimulation or by actively moving the head – Doll's head maneuver.
- Stimulation of anterior semicircular canal moves eyes upward, while stimulation of posterior semicircular canal moves the eye downward.
- The pathway for VOR is from the vestibular end organs via the vestibular nerve to vestibular nuclei.
- From vestibular nuclei to PPRF and 3,4,6 nuclei

Optokinetic Nystagmus (OKN)

- Precise pathway not known in human.
- OKN is produced by rotating an image of the environment around the patient.
- There is a slow phase in the direction of stimulus movement followed by a fast corrective phase to re-fixate the next stimulus.
- OKN response to a pocket tape is a useful beside test.
- Cortical area (parieto occipital temporal junction) may share pathways carrying smooth pursuit command.

Brainstem gaze center

- There are two main brain stem centres (premotor centre for ocular eye movement (a) Para median pontine reticular formation for horizontal gaze (b) Rostral interstitial nucleus of medial longitudinal fascicle for vertical gaze.
- Controls horizontal eye movement is a part of the reticular formation that extends from the level of trochlear nucleus to abducens nuclei.
- Electrical stimulation of this region results in ipsilateral horizontal movement of eyes.
- PPRF get afferent connections from vestibular nuclei, cerebellum, superior colliculus, FEF, pretectal nuclei, inferior colliculus, perihypoglossal nuclei, spinal cord.
- It sends efferent connections to vestibular nuclei, spinal cord and cerebellum. The two main afferent connections of PPRF which are important for ocular motility are
  a) Projections to ipsilateral abducens nucleus
  b) Projections to riMLF (vertical gaze centre) via extra MLF pathway.

- From abducens nucleus the premotor commands for gaze (saccade, pursuit, VOR, OKN) are sent to the ipsilateral lateral rectus and to the contra lateral medial rectus sub nuclei (in midbrain) via internuclear neuron ascending in the contra lateral MLF.
- In PPRF are burst neurons that discharge at high frequencies just prior to and during horizontal saccade. These cells project to the abducens nucleus to generate horizontal saccades. Similar burst neurons are found in riMLF which project to vertical ocular motor neuron to generate vertical saccade.
- The activity of these burst neurons (both horizontal and vertical) is controlled by pause neurons that lie in the midline of the caudal pons and mid brain. Pause neurons cease their activity prior to and during every saccade. The pause neurons exert an inhibitory control over saccadic burst neurons to prevent extraneous saccades during attempted fixation.
- There are inhibitory burst neurons in the PPRF which discharge and project to the neurons of the ocular motor nuclei supplying the antagonistic muscles during saccade movement.

Rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF)

- Is a small horizontally oval group of cells located on either side of the midline in the rostral mesencephalon.
- They receive afferent and send efferent to PPRF.
- riMLF sends projection to motor neurons in the 3 and 4^{th} nuclei for vertical rectus and oblique muscles.
- The gaze centre for upward gaze is located dorsal and rostral to that for downward gaze.
- For vertical saccades, commands from the riMLF for upward saccades first pass through the posterior commissure while commands for downward saccades pass directly caudal to ocular motor nuclei.

Medial longitudinal fascicle

- Is a group of fibres located in the midline of the brain stem ventral to the ventricular and aqueductal systems. These fibres extend from the cervical cord to the rostral midbrain.

- The bulk of fibers in the MLF come from vestibular nuclei to various ocular nuclei. It contains ascending and descending fibres. The most important fibres are located in the medial MLF and connect the contra lateral abducens nucleus with ipsilateral medial rectus sub nucleus.

- Some ascending fibres of MLF synapse in the midbrain within the nucleus of Darkshevich, the interstitial nucleus of Cajal, riMLF.

- Descending fibres of MLF are from interstitial nucleus of Cajal to vestibular nucleus or in the spinal cord.

Abnormalities of supranuclear movement

1. **Gaze palsy**

   Horizontal gaze palsy

   - Patient is unable to move both eyes in a specific horizontal direction.
   - Lesions of the cerebral hemisphere cause gaze paresis or palsy in the opposite direction, the gaze palsy improves with a week (transient).
   - While gaze palsy or paresis due to PPRF lesions (pontine lesions) is ipsilateral to lesions palsy takes longer time to recover weeks to months.
   - Cause – Ischemia, hemorrhage, tumor.

2. **Internuclear ophthalmoplegia**

   - Seen in the lesion of MLF
   - Causes - Demyelination (younger).
     - Vasculopathy (older).
   - There is total or partial failure of adduction on the side of the lesion, while contralateral abducting eye shows nystagmus.
   - Cogan divided INO into-

- Anterior group (Convergence impaired).
- Posterior (Convergence intact).

- Impaired convergence in the anterior group of INO is due to involvement of fibres passing from or to the oculomotor sub nucleus that control medial rectus muscle.

3. One and one half syndrome

- Also called paralytic pontine exotropia.
- There is INO and gaze palsy on the side of lesion.
- The lesion involves PPRF and MLF on the same side.
- The patient is unable to move both eyes towards the side of lesion (Gaze palsy) and the eye on the side of lesion does not adduct. The eye on the side of lesion is straight but the opposite eye shows exotropia.

4. Vertical gaze palsy

Parinaud's syndrome

- Upgaze palsy which is one of the features of this syndrome, is due compression of posterior commissure by dilated suprapineal recess
- Later on down ward gaze also shows paresis/palsy

  Cause

  1. Pinealoma
  2. Hydrocephalus with dilated suprapineal recess

5. Skew deviation

- Is a disconjugate vertical deviation of the eyes due to disturbance of the supranuclear pathways for vertical gaze
- It is associated with other neurological signs and symptoms of brainstem dysfunction
- It does not have localizing value

Other supranuclear ocular motor disorders

1. **Nystagmus**

2. **Non Nystagmus ocular oscillations**

Nystagmus

- Is an involuntary biphasic rhythmic ocular oscillation in which one or both phases are slow.
- The slow phase is responsible for initiation and generation of the Nystagmus, while fast phase (saccadic) is a corrective movement bringing the fovea back on the target.

Types

- Pendular Nystagmus
- Jerky
- Horizontal
- Vertical
- Rotational

Jerky Nystagmus direction is labeled by the direction of the fast phase.

Mechanism of Nystagmus

Caused by disorders of

- Vestibular end organ
- Vestibular nerve
- Brain stem
- Cerebellum
- Cerebral centers for pursuit movement

Pendular Nystagmus is central in origin; cause may be in the brainstem or the cerebellum.

Jerk Nystagmus may be central or peripheral in origin.

Clinical Features:

- Onset – since birth or acquired.
- Family history.
- Headache, impaired vision, oscillopsia, vertigo or other neurological abnormalities.
- Ophthalmic examination.
- Ophthalmoscopy to detect subtle Nystagmus.

Look for

- Nystagmus is in primary position or on eccentric gaze.
- Binocular and conjugate or dissociate Nystagmus.
- Pendular/Jerk – direction of the fast phase.
- Purely horizontal or purely vertical or mixed (has tortional component also).
- Observe Nystagmus for some time to see whether direction changes as PAN and rebound Nystagmus.
- Null point is present or not (direction of gaze when nystagmus is absent or minimal)
- Nystagmus decreases on convergence or not.
- Nystagmus changes by head position or posture.
- Associated rhythmic movement palate, head (spasmus nutans).

1. Congenital Nystagmus (CN) -

- Present since birth, noticed early in life.
- Usually associated with retinal or optic nerve disorders.
- Usually horizontal and Pendular and became jerky on lateral gaze.
- May have normal vision unless there is associated retinal or optic nerve pathology is present.
- Convergence damps the congenital Nystagmus.
- A null point is usually present associated; head turn is seen so eyes move in direction of null point.
- Wave form of CN on recording is sinusoidal or exponentially increasing velocity of slow phase.

Less common pattern of CN-

- PAN, upbeat, downbeat.
- See saw Nystagmus.

2. Gaze Evoked Nystagmus

- Most common type.
- Absent in primary position and seen on extreme gaze on either side.
- It is symmetric, up beating on up gaze and down beating on down gaze.
- It is jerky Nystagmus with fast phase in the direction of gaze.

Causes

- Dysfunction of neural integrator (cerebellum).
- Alcohol intoxication.
- Drug intoxication (anticonvulsants and tranquilizers).
- On recording – it has exponentially decreasing velocity of slow phase.

3. Upbeat Nystagmus:

- Is a spontaneous jerk Nystagmus with the fast phase upward while the eyes in primary position.
- Probable cause is interruption of the anterior semicircular canal projections which are responsible for upward VOR causing downward drift of eyes with corrective upward saccades.
- The amplitude of the oscillation increases on looking up.

Cause

- Structural disease of brainstem usually pontomedullary or pontomesencephalic junction.
- Vermian lesions.
- Congenital rarely seen.
- Wernicks encephalopathy.
- Intoxication with anti convulsions, organophosphates, lithium or nicotine.

Treatment – Gabapentin

4. Downbeat Nystagmus:-

- Is a spontaneous jerk Nystagmus in primary position with fast phase down ward.
- Is due to interruption of the posterior semicircular canal projection (which are responsible for the downward VOR), causing upward drift of the eyes with corrective downward saccades.
- The amplitude of oscillation increases on looking down and laterally (Daroff's sign).

Cause:

- Seen in structural lesion craniocervical junction
    - Chiari malformation
    - Basilar invagination
    - Foramen magnum tumor
- MRI of the foramen magnum in the sagittal plane is the investigation of choice
- Cerebellar degeneration (alcohol, SCA-6 paraneoplastic syndrome)
- anticonvulsant
- Wernicke's encephalopathy
- Brain stem encephalitis
- Rarely congenital

Treatment: Baclopen, clonazepam

5. Periodic alternating Nystagmus

- Is horizontal jerk Nystagmus in which fast phase beats in one direction and then damps or stop for a few seconds before changing direction to the opposite side
- During this transition period Nystagmus may beat vertically
- Complete cycle takes 3 minutes

Cause:

- Craniocervical junction disorders
- Cerebellar disorders
- Congenital
- Rx –
    - Baclopen
    - Treatment of cause

6. Rebound Nystagmus:

- Is a horizontal gaze evoked Nystagmus in which the direction of the fast phase reverses with sustained lateral gaze or beats transiently in opposite direction when eyes return to primary position

Cause:

- Cerebellar disorders
- Dysfunction of the per hypoglossal nuclei in medulla

7. Vestibular Nystagmus

- Result from damage to labyrinth , vestibular nerve (peripheral type)
- or from damage to vestibular nuclei or their connections in the brainstem (central type)

Peripheral vestibular Nystagmus	**Central vestibular Nystagmus**
Is associated with nausea, vomiting, Perspiration, diarrhea, or may be Associated with hearing loss and tinnitus	severe but there are other associated neurological features like headache, dysconjugate gaze, pyramidal tract sign

Seesaw Nystagmus (SSN)

- In this one eye rises and intort, while other eye falls and extorts
- Causes –
    - Lesions at mesodiencephalic junction

- Chiasmal lesion
- Congenital (achiasmatic or septo-optic dysplasia)

Acquired SSN may be accompanied by bitemporal hemianopia

Tortional (Rotatary Nystagmus)

- Eyes oscillate in a pure rotatory plane.
- It may be present in primary position or with head positioning or gaze deviation
- Cause – lesion of central vestibular pathway, mixed Nystagmus horizontal tortional Nystagmus occurs with peripheral vestibular disease.

Brun's Nystagmus

- Seen in large CP angle tumor.
- Nystagmus is bilateral, asymmetric.
- On gaze towards the side of lesion large amplitude low frequency Nystagmus is seen due to compression of brain stem neural integrator (ipsilateral medical vestibular nucleus).
- On gazing away from the side of lesion small amplitude, high frequency Nystagmus is seen due to ipsilateral vestibular dysfunction.

Non Nystagmus ocular oscillations

- Ocular dymetria
- Ocular flutter
- Opsoclonus

These disturbances in ocular movements are seen in cerebellar disorders.

Ocular dysmetria

- Is the over shoot of gaze during change of the fixation
- Left cerebellar lesion will cause overshoot of both eyes. When the gaze is shifted from the left to primary position, while right cerebellar lesion will cause overshoot of both eyes when they are moved from right to central position

Ocular flutter

- Is a rapid small amplitude, horizontal conjugate back to back saccades that occur spontaneously during fixation or after fixation
- They occur in intermittent bursts

Opsoclonus

- Is a spontaneous, chaotic multi directions saccadic eye movements.
- Opsoclonus is usually continuous and persists during sleep.
- It is aggravated by attempts at fixation and may be associated with myoclonic jerks of the limbs and cerebellar ataxia (dancing eyes dancing feet syndrome).
- Caused by the dysfunction of the pause cells in the pons.
- Seen in infections encephalitis
  - Occult neuroblastoma in children.
  - Paraneoplastic disorders.

Ocular bobbing

- Rapid downward movement of both eyes followed by slow drift to primary position.
- Seen in usually comatose patient.
- Reverse bobbing
- Inverse bobbing

Ocular myoclonus is a vertical pendular oscillation, usually associated with similar oscillations of the soft palate and sometimes other muscles of branchial origin.

- Seen after brain stem infarction

Treatment: Carbamezepine, clonazepam, valproic acid or chronically patching one eye.

EXAMINATION OF NYSTAGMUS

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Objective:

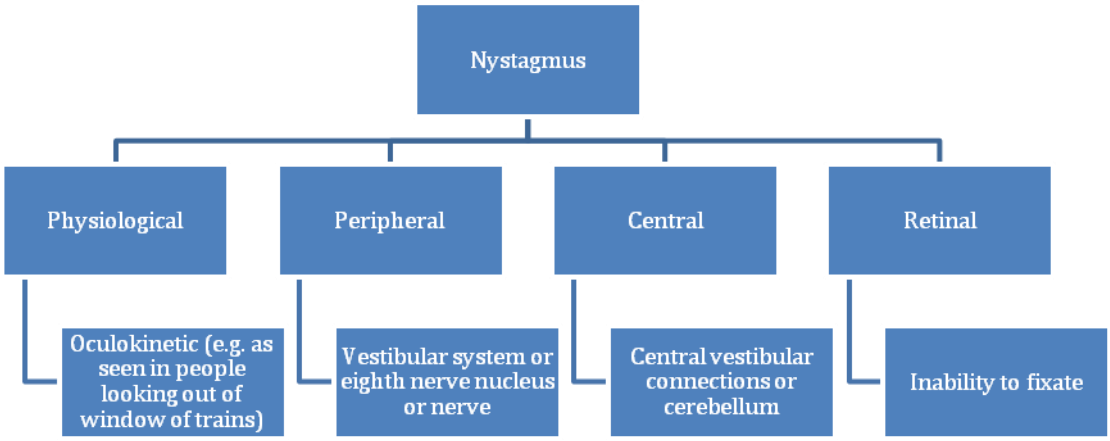
After reading this chapter, you should be able to

- Define nystagmus
- Identify the correct sequence to evaluate nystagmus
- Differentiate various nystagmus varieties and their causes

Definition:

Nystagmus is defined as to and fro, involuntary rhythmic, repetitive oscillations of the eyeballs, either pendular or with slow and fast components. Conventionally, nystagmus is defined in the direction of the fast component.

Types of Nystagmus:



How to Examine:

During examination, sit straight in front of the patient and ask the patient to follow your finger with both eyes. Move the finger in turn up, down and to each side. Hold

the fingers briefly in each position, so that finger can be seen easily by both eyes (i.e. binocular vision). Now watch for the following:

- Is there any abnormal to-fro movement of the eyeballs?
- Is it a nystagmus or a nystagmoid movement?
- Is the movement symmetrical, moving at the same speed in both directions *(pendular nystagmus)* or if there is a fast component in one eye with a slow component *(jerk nystagmus)*
- What is the direction of the fast component; is it in horizontal plane, vertical plane or is it rotatory?
- What is the position of the eye when nystagmus occurs, and when is it most marked?
- What is the degree of nystagmus; primary, secondary or tertiary?
- Which eye is most affected; abducting eye or adducting eye?
- Does it occur in one direction only or in many directions?
- Whether it occurs in direction of gaze in more than one direction (multi directional gaze evoked nystagmus)?

Nystagmus or nystagmoid?

- Nystagmoid movements are present in many people at extremes of gaze (physiologic nystagmus).
- Nystagmus present in the eyes with deviation less than 30 degrees from the midline is abnormal.
- To remove this error - at the extremes of the lateral gaze one or two nystagmoid jerks can be seen normally; ensure that the target remains within binocular vision
- If nystagmoid jerks are present, repeat the test. If there is true nystagmus, it will be apparent at less than extreme gazes.

Pendular or jerk nystagmus?

Pendular nystagmus: smooth sinusoidal oscillations

Jerk nystagmus: alternation of slow drift and corrective quick phase

Degree of nystagmus?

First degree - Nystagmus is present only with the eyes deviated to one side only

Second degree - Nystagmus is present with eyes deviated to one side and in midline also

Third degree - Nystagmus is present in all directions of gaze

Conjugate or dysconjugate and/or dissociative nystagmus?

Conjugate nystagmus: the direction of the oscillations in each eye is the same

Dysconjugate nystagmus: the direction of the oscillations in each eye is different

Dissociative nystagmus: the size of the oscillation differs in each eye

Physiological or pathological nystagmus?

Physiological nystagmus:

- Rotational acceleration produces nystagmus in the plane of the rotation e.g. optokinetic nystagmus
- Caloric testing sets up convection currents in the lateral semicircular canals producing a horizontal nystagmus
- Nystagmus in extreme lateral or vertical gaze (end-point nystagmus). End point nystagmus is a gaze evoked nystagmus, may be pathologic if they persist beyond few beats in end gaze.

Pathological nystagmus: Dysfunction of labyrinth, vestibular nerve or central nervous system

Central versus peripheral nystagmus?

	Central	Peripheral
Sustained	+	-
Fatigue	-	+
Associated with symptoms of vertigo	-	+
Reduced by fixation	-	+

- Peripheral nystagmus is not associated with any other eye movement abnormalities and usually has a rotatory component.

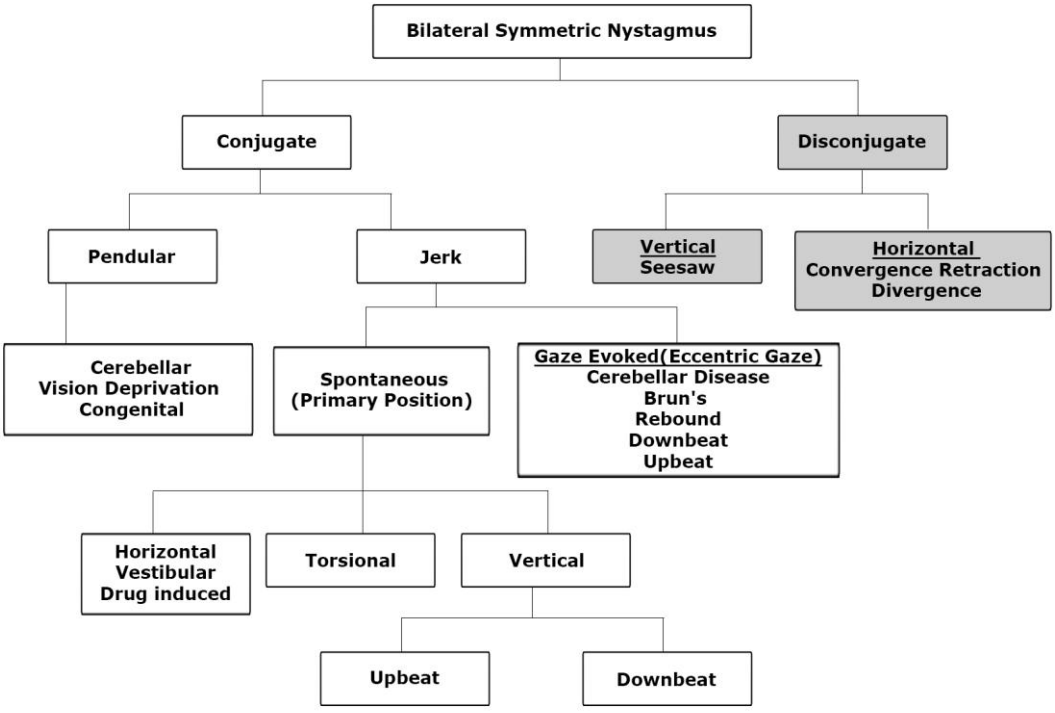


Figure: Approach to a patient with bilateral symmetric nystagmus

Physiology of eye movements:

Vestibular component of eye movement:

The utricle and saccule respond to linear acceleration, whereas the semicircular canals respond to angular acceleration. These responses are mediated by hair cells and transmitted to the vestibular ganglion and subsequently to the vestibular nuclei. Under normal circumstances, the neural activity in the labyrinths is equal on both sides. It is convenient to visualize the action of each vestibular system as pushing the ipsilateral eye toward the opposite side. When the two labyrinths push equally, the system is in balance and function is normal. When **one labyrinth is underactive**, the opposite labyrinth pushes the eyes, extremities, and body toward the side of under-activity. Nystagmus results from a corrective saccade initiated by the frontal eye fields in response to the deviation of gaze toward the side of the less active labyrinth. The fast component of the nystagmus is therefore in the opposite direction from the hypoactive labyrinth.

When both labyrinths are diseased or malfunctioning, there is no vestibular imbalance and hence no nystagmus.

Optokinetic nystagmus (OKN):

Optokinetic nystagmus is a normal physiologic phenomenon. Presence of nystagmus indicates an intact visual pathway. It means conjugate nystagmus induced by a succession of moving visual stimuli. With normal vision, a nystagmus develops in both adults and infants.

Physiology of OKN:

The nystagmus consists of initial slow phases in the direction of the stimulus (smooth pursuits), followed by fast, corrective phases (saccade). The ipsilateral parieto-occipito-temporal junction mediates pursuit movements while ipsilateral frontal lobe generates corrective saccadic movement.

Eliciting optokinetic nystagmus:

- Rotate the optokinetic drum in front of the patient. Ask the patient to look at the drum as you rotate it slowly.

- Move a strip of paper with alternating 2-inch black and white strips across the patient's visual field. Pass it in front of the patient's eye at reading distance while instructing the patient to look at it as it rapidly moves by.
- Hold a mirror in front of the patient and slowly rotate the mirror to either side of the patient.

OKN can be used as a crude assessment of the visual system, particularly in infants. When factitious blindness or malingering is suspected, check for optokinetic nystagmus to determine whether there is an intact visual pathway.

Jerk nystagmus:

It is slow phase in one direction followed by saccadic fast phase in the opposite direction. The direction of the nystagmus is the direction of the fast phases despite the fact that it is the slow phases, which are responsible for generating the nystagmus.

Gaze-evoked nystagmus:

It is the most common form of nystagmus. It is a jerk nystagmus elicited by attempted maintenance of eccentric eye position; no nystagmus is present in primary position.

Gaze paretic nystagmus:

The patient is unable to sustain eccentric gaze and requires repeated saccades to gaze laterally. With a lesion of one cerebellar hemisphere the eyes at rest may be deviated 10 degrees to 30 degrees toward the unaffected side. When the patient attempts to gaze elsewhere, the eyes saccade toward the point of fixation with slow return movements to the resting point. The movements are more marked and of greater amplitude when the patient looks toward the affected side.

Bruns' nystagmus:

This is a horizontal jerk nystagmus (ipsilateral coarse and contralateral fine nystagmus), whose direction is dependent upon gaze and whether or not fixation is suppressed. It is characteristic of cerebellopontine angle tumors.

With gaze directed toward the side of the lesion, a large amplitude, gaze-evoked nystagmus is seen. The fast phases are in the direction of gaze and the slow phases are decreasing-velocity exponentials. With gaze directed to the side opposite the lesion, a small-amplitude, linear slow phase nystagmus is elicited with the fast

phases in the direction of gaze. When the eyes are closed, a nystagmus beating in the direction opposite the side of the lesion predominates.

The nystagmus that occurs on gaze toward the side of the lesion is gaze evoked nystagmus caused by defective gaze holding due to brainstem compression and cerebellar dysfunction, whereas the nystagmus that occurs during gaze toward the side opposite the lesion is caused by vestibular imbalance.

Rebound nystagmus:

The fast component is in the direction of lateral gaze, but transiently reverses direction when the eyes come back to primary position. This type of nystagmus may be unique to cerebellar disease.

Nystagmus retractorius:

Results from co-contraction of all extraocular muscles. It may accompany convergence spasm. Lesion in upper midbrain tegmentum (usually vascular disease or tumor especially pinealoma)

Downbeat nystagmus:

A vertical jerk nystagmus present in primary position with linear upward slow phases and fast phases beating in the downward direction. It is highly suggestive of a disorder of the cranial-cervical junction such as Chiari malformations. It is best seen when the eyes are deviated laterally and slightly below the horizontal. A defect in downward pursuit has been suggested as the cause of this form of pursuit-defect nystagmus.

See-saw nystagmus:

A conjugate, pendular, torsional oscillation with a superimposed disjunctive vertical vector. The intorting eye rises while the opposite, extorting eye, falls. Torsional movements predominate in all fields of gaze, but the see-saw vertical feature may be restricted to the primary position or to downward gaze.

It is probably due to diencephalic (thalamic) dysfunction involving a pathway from the zona inserta to the interstitial nucleus of Cajal. Seen in parasellar tumors

expanding within the third ventricle, upper brainstem vascular disease and severe head trauma. Look for bitemporal hemianopia in parasellar masses.

Internuclear ophthalmoparesis (INO):

The most commonly recognized syndrome that results from MLF damage is INO and is characterized by slowing or limitation of adduction (on the sameside as the MLF lesion) during horizontal eye movements and the contralateral abducting eye will usually exhibit a disassociated horizontal nystagmus.

The abduction nystagmus implicates an adaptive response to overcome the weakness of the contralateral medial rectus as explained by Hering's law of equal innervation, which states that attempts to increase innervation to the weak muscle in one eye must be accompanied by a commensurate increase in innervation to the yoke muscle in the other eye.

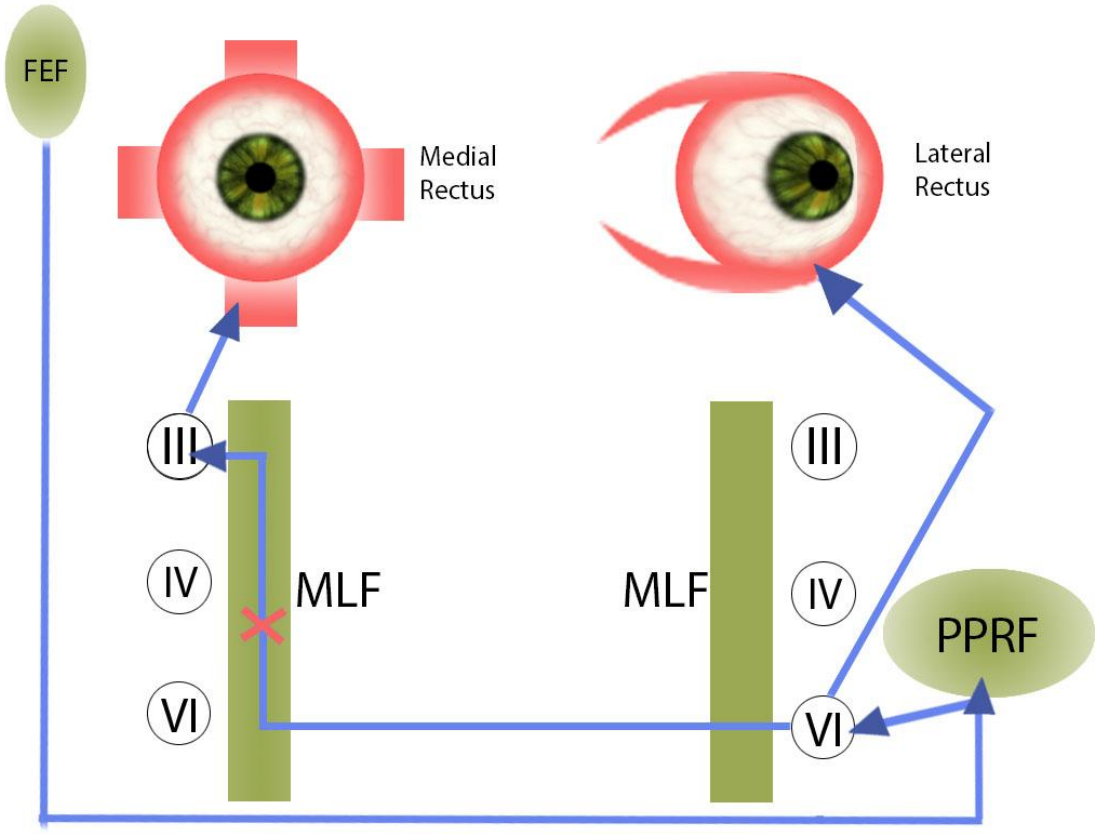


Fig: In right INO, there is RIGHT MLF dysfunction, RIGHT EYE ADDuction impairment, horizontal nystagmus in the ABDucting LEFT EYE.

One-and-a-half syndrome:

This syndrome consists of a gaze palsy in one direction with an INO when executing a saccade to the opposite side. It is produced by damage to the PPRF or abducens nucleus and the MLF on the same side within the pontine tegmentum. Convergence is generally spared as cranial nerve III is spared bilaterally.

Given the preserved abduction of the eye contralateral to the lesion, one commonly observes a primary position exotropia also known as paralytic pontine exotropia

Wall-eyed bilateral INO syndrome:

If the lesion affects the MLF within the pons or midbrain, vergence pathways and the oculomotor apparatus can be coincidentally disrupted, resulting in a variety of eye movement abnormalities that include impaired convergence. These lesions are typically bilateral and produce divergence of the eyes.

Interpretation:

- Nystagmoid jerks: Normal
- PendularNystagmus: congenital
- Rotatory nystagmus
    - Pure rotatory nystagmus is central in origin while peripheral horizontal nystagmus usually has a rotatory component.
- Vertical nystagmus: usually indicates brainstem lesions
    - Upbeat nystagmus: indicates upper brainstem. Common causes: demyelination, stroke, Wernicke's encephalopathy
    - Downbeat nystagmus: indicates medullary-cervical junction lesion. Common causes are: Arnold Chiari malformation, syringobulbia, demyelination.
- Horizontal nystagmus:
    - Ataxic nystagmus: nystagmus of abducting eye>>adducting eye. May be associated with internuclear ophthalmoplegia. Common causes are lesion in pons either multiple sclerosis, cerebrovascular disease.
    - Multidirectional gaze evoked nystagmus: Always central in origin-vestibular or cerebellar or drug induced

- Unusual eye movements:
    - Opsoclonus: rapid oscillations of the eyes the horizontal rotatory or vertical direction- indicates brainstem lesion, sometimes a paraneoplastic syndrome
    - Ocular bobbing: eyes drifting up and down in the vertical plane- associated with pontine lesions
- Nystagmus retractorius- lesion in upper midbrain tegmentum (usually vascular disease or tumor especially pinealoma)

Suggested reading:

1. De Jong's Neurological Examination. 6th edition Page 187-190
2. Localization in Clinical Neurology by Brazis Paul W. 5th Edition Page 354-356
3. Bickerstaff's Neurological Examination in Clinical Practice. 6th edition Page 66-71

Examination of the Trigeminal nerve

Awadhesh Kumar Jaiswal, Guruprasad Bettaswamy, kuntal kanti Das, Sanjay Behari

The trigeminal nerve is the largest cranial nerve and has both sensory and motor components. It provides sensory innervation to face and mucous membrane of oral and nasal cavities and motor innervation to the muscles of mastication.

Anatomy:

The motor part:

The motor nucleus of the trigeminal nerve is situated at mid pons. It receives the supranuclear control through corticobulbar fibers that originate from the lower third part of the motor cortex. The supranucelar control is bilateral with the contralateral contribution larger than the ipsilateral. The motor root (portio minor) exits from the motor nucleus, traverses the substance of pons and emerges from the anterolateral part of the pons, anteromedial to the sensory root (portio major).It enters in the Meckel's cave, passes beneath the trigeminal ganglia and exits through the foramen ovale and joins with the mandibular division of the trigeminal nerve to form mandibular nerve. It gives motor innervation to the masticatory muscles: masseter, temporalis, lateral and medial pterygoid muscles. In addition, it also gives motor supply to mylohyoid, anterior belly of digastric, tensor veli palatini and tensor tympani muscles.

The sensory part:

The sensory neurons of the trigeminal nerve are located in the trigeminal (Semilunar or Gasserian) ganglia situated near the petrous apex. From this ganglia, the central fibers enter the pons and terminate in to the main sensory nucleus of trigeminal nerve, spinal nucleus of the trigeminal nerve and the mesencephalic nucleus.

Fibers entering the main sensory nucleus of trigeminal nerve are concerned with fine touch and

tactile discrimination. Second order neurons arising from this nucleus form crossed and uncrossed dorsal trigeminal tract (dorsal trigeminal lemniscus) which terminate in the ventro-posterior-medial(VPM) nucleus of thalamus. Third order neurons arising from VPM nucleus of thalamus project via internal capsule to the inferior portion ofthe sensory cortex.

Fibers entering the spinal nucleus of the trigeminal nerveform a bundle (spinal tract of trigeminal nerve) and these fibers are concerned with pain, temperature and crude touch. The spinal nucleus of the trigeminal nerve has three parts: from cranial to caudal- pars oralis, pars interpolaria and pars caudalis. The lamination of nerve fibers in this nucleus has a distinct pattern. The fibers of the ophthalmic division of the trigeminal nerve terminate in to the most caudal part of the spinal nucleus of trigeminal nerve. The fibers of the mandibular division of the trigeminal nerve terminate in to the most rostral part of the spinal nucleus of the trigeminal nerve. Thus, the midline areas (nose and mouth) are represented rostrally in the spinal nucleus and the more lateral facial sensations are represented caudally in the spinal nucleus. This pattern of arrangement is responsible for the characteristic onion-peel pattern of facial sensory loss which has a great localizing value.

In addition, the spinal nucleus of trigeminal nerve receives general somatic afferent fibers from the facial, glossopharyngeal and vagus nerve.

The mesencephalic nucleus of the trigeminal nerve is located rostral to the main sensory nucleus to the superior colliculus of the midbrain. It receives proprioceptive impulses from the muscles of mastication and other motor cranial nerves (3^{rd}, 4^{th} and 6^{th}).

The peripheral fibers from the Gasserian ganglia give rise to three nerve trunks: the **ophthalmic, maxillary and mandibular** divisions of the trigeminal nerve.

The **ophthalmic** division divides in to frontal, lacrimal and nasociliary branches and they carry sensations from the skin of the forehead, scalp (as far back as lambdoidal suture), nose, upper eyelid, upper half of cornea and conjunctiva, temple, meninges of the anterior cranial fossa, frontal, ethmoidal and sphenoidalparanasal sinuses, lacrimal canals and upper part of the nasal cavity and nasal septum.

The **maxillary** division gives rise to palatine nerves, superior alveolar nerves, inferior palpebral

nerve, superior labial nerve and zygomaticofacial branches. It supplies the skin of the lower eyelid, upper half of cornea and conjunctiva, maxillary sinus, lower nasal cavity, hard and soft palate, teeth and gum of upper jaw and meninges of middle cranial fossa.

The **mandibular** division joins the motor root of the trigeminal nerve to form the mandibular nerve. In addition to giving motor supply to the muscles of the mastication: masseter, temporalis, medial and lateral pterygoid muscles, it also gives motor supply to the mylohyoid, anterior belly of digastric, tensor velipalatini and tensor tympani muscles. It also gives off branches: lingual, inferior alveolar and mental nerves that receives sensory inputs from the skin of the lower lip, lower jaw, chin, tympanic membrane, external auditory meatus including tragus, mucosa of the floor of mouth, anterior two-thirds of the tongue (general sensations, not taste sensation, which is carried by facial nerve), lower gums and teeth and meninges of the posterior cranial fossa.

Clinical examination of the trigeminal nerve:

The clinical examination of the trigeminal nerve is performed in three parts.

1. Motor evaluation
2. Sensory evaluation
3. Reflex evaluation

Motor evaluation

Motor evaluation of the trigeminal nerve comprises of evaluation of motor power of muscles of mastication: masseter, temporalis, medial and lateral pterygoid muscles, in the following manner.Look for any signs of wasting of masseter and temporalis muscles in form of hollowing of temporal region and flattening of the contour of jaw (area of masseter muscle). Sometimes, fasciculation may also be noted over these muscles. Ask the patient to clench the teeth and palpate for the contractions of temporalis and masseter muscles on both sides. If there is weakness or paralysis of these muscles, there will be impairment or absence of muscle contractions on the involved side.

Ask the patient to open his mouth widely and look for any deviation of the jaw. Normally, while opening the mouth, the jaw remains in midline and there is no deviation of jaw on either side. In case of weakness or paralysis of muscles of mastication especially lateral pterygoid, there will be deviation of jaw to the side of paralyzed muscle. This is because when both sides of the pterygoid muscles contract, it helps in elevation and depression of mandible and there is no deviation of mandible.When the pterygoid muscle of one side contracts, it deviates the mandible to the opposite side due to unopposed action of the lateral pterygoid muscle of the normal side. The deviation of the jaw should be appraised by noting the relation of the upper and lower incisor teeth when the jaw is opened or closed, not by the position of the lip to avoid the apparent deviation of jaw caused by facial nerve paralysis.

Ask the patient to move his jaw from side to side against resistance, in case of weakness or paralysis of the muscles of mastication, the patient will be able to move the jaw to the side of paralysis but not to the side of non-paralysis.

For examination of subtle motor weakness of muscles of mastication, the patient is asked to clench his teeth with a wooden spatula between the molar teeth of the upper and lower jaw. The depth of the tooth marks is noted and compared with other side. The tooth marks will be poor in cases of weakness of the muscles of mastication.

The weakness or paralysis of the muscles of mastication is most marked in nuclear or infranuclear lesions of the trigeminal nerve. In cases of unilateral supranuclear lesion of the trigeminal nerve, there is rarely any weakness or paralysisof the muscles of mastication is noted due to bilateral supranuclear innervation. However, in cases of bilateral supranuclear lesions, there may be marked weakness.

Clinical examination of other muscles (mylohyoid, anterior belly of digastric, tensor veli palatini and tensor tympani) supplied by the trigeminal nerve is very difficult and often not possible. However, flaccidity of the floor of mouth due to paralysis of mylohyoid and anterior belly of digastric may be noted on palpation. The palatal arch may be lower than normal side due to paralysis of tensor veli palatine. Difficulty in hearing high tone and dysacusis for high tones may be due to paralysis of tensor tympani.

Sensory evaluation:

Sensory evaluation of the trigeminal nerve comprises of evaluation of all modalities of sensations on the skin of the face and the mucus membrane in the following manner.

The patient is instructed to close his eyes and respond to:

-When touched with a wisp of cotton (light touch)

-When stuck with a pin (pain)

-When touched with test tubes filled with hot and cold water (heat and cold)

The representative areas chosen for the preliminary evaluation are **(Not in the midline):**

- The forehead and the upper part of the side of nose (ophthalmic division)

- The malar region and the upper lip (maxillary division)

- The chin and the anterior part of the tongue (mandibular division)

Any area of sensory dysfunction (anaesthetic zone) is mapped carefully, and an assessment should be made:

-Whether the aneasthetic zone corresponds to the any peripheral division or any segmental distribution (onion-peel pattern) of the trigeminal nerve

-Whether the sensory loss involves all the modalities of sensations or is in a pattern of dissociated anaesthesia (loss of pain and temperature sensations with preservation of light touch sensation)

During sensory evaluation of the trigeminal nerve, it should be remembered that:

The skin over the angle of mandible is not supplied by the trigeminal nerve but by the greater auricular nerve from the second cervical segment. The overlap between the cutaneous zones of the major divisions of the trigeminal nerve is minimal, in contrast to the rest of the body, especially the trunk.

-On the scalp, the area supplied by the trigeminal nerve and the second cervical segment meet a little posterior to the vertex, but the exact point varies.Erosion of the ala nasi and surrounding skin can occur in cases of severe sensory loss of the trigeminal nerve.

-Corneal ulceration may be due to profound sensory loss in the distribution of the ophthalmic division of the trigeminal nerve but it may also be due to facial nerve palsy leading to inadequate close of the eye causing exposure keratitis.

Reflex evaluation:

Following reflexes are examined for the evaluation of the trigeminal nerve.

1. Jaw jerk (masseter or mandibular reflex)
2. Corneal reflex

Some less important reflexes associated with the trigeminal nerve are:

1. Zygomatic reflex
2. The head retractor reflex
3. The sneeze (nasal or sternutatory reflex):

Jaw jerk (masseter or mandibular reflex):

It is a monosynaptic muscle stretch reflex: the afferent arc is through the mandibular division of the trigeminal nerve that runs through the mesencephalic nucleus and the efferent is through the mandibular nerve that originates in the motor nucleus of the trigeminal nerve. This important reflex is elicited in the following way:

The patient is asked to open the mouth partly and let the jaw muscles relax. The examiner places his forefinger or little finger below the lower lip on the chin and taps it in a downward direction with a percussion hammer. The response is in form of a reflex contraction of the masseter and the temporalis muscles with an upward jerk of the jaw.

An exaggerated jaw jerk is indicate of lesion above the level of midpons. In a patient having exaggerated deep tendon reflexes in the limb with extensor planter reflex and spasticity, a hyperactive jaw jerk is suggestive of lesion above the pons rather than a cervical cord lesion. An exaggerated jaw jerk is often found in the pseudobulbar palsy, motor neuron disease and multiple sclerosis. It should also be remembered that it may be absent in normal individuals and hence absent jaw jerk rarely has a localizing value.

Corneal reflex:

This is an extremely sensitive reflex and sometime it may be the sole manifestation of trigeminal nerve impairment for example in cases of cerebellopontine angle tumors. The afferent arc of the corneal reflex travels through the ophthalmic (upper cornea) and the maxillary (lower cornea) division of the trigeminal nerve. The efferent arc moves through the ipsilateral (direct response) and contralateral (consensual response) facial nerve to the orbicularis oculi muscle causing blinking of the eye. This reflex is elicited in the following way:

The test must first be explained to the patient, who will otherwise undoubtedly flinch if some pointed object is suddenly thrust towards his eye. The patient is asked to look at upward and opposite direction as far as possible in order to widen the palpebral fissure as much as possible. The examiner should approach from the side to avoid blinking or visual-palpebral reflex. The cornea (just lateral or below the pupil) is touched or stroked with a wisp of moistened cotton wool. It is important to touch the cornea, not the bulbar conjunctive or lid or lashes. A normal corneal reflex consists of an immediate blink response bilaterally. In the presence of trigeminal nerve impairment, the direct and consensual responses will be absent when the affected side cornea is stimulated, but will be present on stimulation of the contralateral normal side of cornea. With facial nerve palsy, blinking will occur only on the non-paralyzed side when either of the

cornea is stimulated.

It is worth to remember that some people have very insensitive cornea, especially if there is some degree of exophthalmos. If no response is obtained from the either side, it is worthwhile to ask the patient if he can feel the touch stimulus on the cornea and if it is equal on both sides.

Zygomatic reflex:

It is a modification of the jaw reflex. Both the afferent and efferent arcs are carried by the trigeminal nerve. The reflex is elicited by gentle percussion over the zygoma which causes ipsilateral deviation of the mandible. The reflex is positive only in cases of supranuclear lesions and absent in normal individuals.

The head retraction reflex:

This reflex is not of much importance. The afferent arc is formed by the trigeminal nerve and the efferent arc is by the cervical nerves to the retractor muscles of the neck. The center of the reflex is in the upper cervical cord. The reflex is elicited as follows: The patient is asked to bend the head slightly forward and the upper lip is sharply tapped just below the nose. A quick, involuntary, backward jerk of the head is indicative of positive reflex. It is present in cases of bilateral supracervical lesion of the corticospinal tract. The reflex is absent in normal persons.

The sneeze (nasal or sternutatory reflex):

The afferent fibers of this reflex are carried from the nasal mucosa through the trigeminal nerve and the efferent fibers are carried through the trigeminal, facial, glossopharyngeal and vagus nerve. To elicit this reflex, the nasal mucosa is stimulated by tickling it with a hair or some similar object which leads to sneezing by contraction of the nasopharyngeal and thoracic muscles with a violent expulsion of air from the nose and mouth.

Localization of the lesions affecting the trigeminal nerve

Clinical features of supranuclear lesions:

Supranuclear control of the trigeminal motor function is bilateral, however, the contralateral hemisphere exerts predominant control on the voluntary activity of the muscles of mastication. Hence, a supranuclear lesion causes mild motor weakness with exaggeration of jaw jerk. Bilateral lesions (pseudo-bulbar palsy) cause profound motor weakness with exaggeration of jaw jerk. Fasciculations, profound bilateral weakness along with exaggeration of jaw jerkmay be noted in cases of amyotrophic lateral sclerosis. Thalamic lesions may cause anaesthesia of the contralateral face.

Clinical features of nuclear lesions:

The nuclear lesions of the trigeminal nerve often involve other adjacent brain stem structures and therefore, nuclear lesions of the trigeminal nuclei are diagnosed by "the company they keep" (e.g. long tract signs and other cranial nerve involvement).

Pontine lesions may affect the motor and main sensory nucleus of the trigeminal nerve. Involvement of motor nucleus results in to ipsilateral paresis, atrophy and fasciculation of the muscles of mastication. Involvement of sensory nucleus causes ipsilateral hemianeasthesia of face. Associated clinical findings may be contralateral hemiplegia (due to involvement of corticospinal tract in the basis pontis) and contralateral hemisensory loss of pain and temperature (due to involvement of spinothalamic tract). **Internuclear ophthalmoplegia** (due to involvement of medial longitudinal fasciculus) and ipsilateral Horner's syndrome (due to involvement of descending sympathetic pathway) may also occur.

Spinal nucleus of trigeminal nerve extends from the lower pons to the third or fourth cervical spinal cord level. Involvement of this nucleus leads to loss of pain and temperature sensations on the ipsilateral face. The lateral spinothalamic tract lies in the close association with the spinal nucleus of trigeminal nerve and often gets affected with the lesions affecting the spinal nucleus of trigeminal nerve causing associated loss of pain and temperature sensations over the contralateral trunk and limbs. With upper (rostral) spinal nuclear lesion, the entire trigeminal cutaneous distribution is affected. Caudal spinal nuclear lesion (at the level of upper cervical

spinal segment) results in loss of pain and temperature sensations over the peripheral (lateral) forehead, cheek and jaw, sparing the peri-oral region (onion-peel pattern of sensory loss). The onion-peel segmental distribution reflects the rostral-caudal somatotopic arrangement of the cutaneous distribution of the spinal nucleus (e.g., peri-oral area-rostral part of spinal nucleus; lateral face-caudal part of spinal nucleus).

Clinical features of preganglionic trigeminal nerve root lesions:

The lesions that involve the preganglionic trigeminal nerve root often also involve the neighboring cranial nerves (especially VIIth and VIIIth cranial nerves) and other structures such as cerebellum. A common example of such lesion is tumor of the cerebellopontine angle. There will be features of trigeminal root involvement- such as ipsilateral facial pain, paresthesia, sensory loss, masticatory paresis and depressed corneal reflex; associated with ipsilateral sensorineural hearing loss, tinnitus, vertigo (due to involvement of cranial nerve VIII) and facial paralysis, ipsilateralataxia and nystagmus (due to involvement of cranial nerve VII and cerebellum).

Another common lesion affecting the trigeminal root is trigeminal neuralgia where the root is often gets compressed by a vascular loop. The clinical feature is typical distinctive syndrome of sudden, excruciating, lancinating, paroxysmal pain in one or more of the division (often maxillary and mandibular) of the trigeminal nerve.

Clinical features of Gasserian ganglion lesions:

Lesion affecting the Gasserian ganglia causes ipsilateral facial pain, paresthesia, sensory loss, masticatory paresis and depressed corneal reflex. Sometimes, when the lesion is large (large trigeminal schwannoma), it is associated with ipsilateral Horner's syndrome due to involvement of sympathetic fibers around the internal carotid artery (Raeder's paratrigeminal syndrome).

Clinical features of lesions affecting the peripheral branches of the trigeminal nerve:
Lesion involving the ophthalmic or maxillary division causes sensory loss confined to the cutaneous supply of the ophthalmic or maxillary division respectively. Lesion involving the mandibular nerve causes sensory loss confined to the cutaneous supply of the mandibular nerve along with ipsilateral masticatory muscles paralysis.

Localization of lesion based on the sensory loss on the face:

- Complete loss of sensations over the distribution of one or two sensory divisions but not involving the entire trigeminal nerve suggests a lesion distal to the gasserian ganglia or a partial lesion of the gasserian ganglia or root.
- Complete loss of sensations over the entire distribution of the trigeminal nerve suggests a lesion in the gasserian ganglia or root or a more extensive lesion distal to the ganglia.
- Loss of pain and temperature with preservation of touch (dissociated sensory loss) suggests a lesion in the spinal nucleus of the trigeminal nerve. The sensory loss tends to follow an **onion-peel pattern** in the face.
- Loss of touch with preservation of pain and temperature suggests a lesion involving the main sensory nucleus of the trigeminal nerve in the pons.

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EXAMINATION OF FACIAL NERVE

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Related anatomy

The facial nerve contains not only motor fibres (somatic motor) supplying the muscles of facial expression, but also the visceral efferent (parasympathetic) secretomotor fibres to the lacrimal and salivary glands , and visceral afferent (taste) fibres from the taste buds of the anterior two thirds of the tongue. Facial nerve also carries cutaneous sensory impulses from the external auditory meatus and region back of the ear.

The motor nucleus lies in the caudal pontine tegmentum medial to the descending nucleus and tract of the fifth nerve. Axons from the motor nucleus project dorsomedially towards the floor of the fourth ventricle, where they loop around the Abducens nucleus (forming the genu of the facial nerve and the facial colliculus),and then project ventrolaterally to exit the brainstem between the pons and medulla at the lateral end of the pontomedullary sulcus just anterior to the entry of vestibulocochlear nerve and three millimeter rostral to the entry of glossopharyngeal nerve. The facial nerve and its visceral root (nervus intermedius) cross the cerebellopontine angle immediately adjacent to the eighth cranial nerve, enter the internal auditory meatus and passes through the facial canal of the temporal bone in close proximity to the inner ear and the tympanic membrane. The facial nerve gives off nerve to Stapedius 6mm above the stylomastoid foramen.

The branchial motor fibres exit the facial canal through the stylomastoid foramen and immediately innervate the stylohyoid, posterior belly of the digastrics and occipitalis muscles. Remaining fibres travel in the substance of the parotid gland to innervate the muscles of facial expression and the platysma.

The cortical fibers of the facial nerve proper originate from the lower third of the motor strip. They course in the genu of the internal capsule and the middle third of the cerebral peduncle, supplying the seventh nucleus in the lower pons. The supranuclear innervation is bilateral to the

muscles of the forehead and eyes but only contralateral to the muscles of the lower part of the face. This accounts for the sparing of the upper facial muscles with a contralateral cortical lesion.

The nervus intermedius joins the motor segment at the point where it exits from the pons. The intermediate nerve is composed of contributions from three areas:

1. The superior salivary nucleus in the pons supplies secretomotor fibers. They go to *(a)* the lacrimal, nasal, and palatine glands (via the greater superficial petrosal nerve) and *(b)* the submandibular and sublingual salivary glands (via the chorda tympani nerve).
2. The gustatory (solitary) nucleus in the medulla supplies sensory fibers. These fibers go to the taste buds on the anterior two-thirds of the tongue (via the chorda tympani nerve).
3. The dorsal part of the trigeminal tract supplies fibers that convey cutaneous sensation from the external auditory meatus and skin behind the ear (distributed with the facial nerve proper).

The general visceral efferent fibres which comes from the superior salivatory nucleues sends axons to the nervus intermedius. This courses **(1)**via the greater superficial petrosal nerve (GSPN) to the ptrerygopalatine ganglion and then to the lacrimal gland (lacrimation) and mucosa of the nose and mouth(secretion). GSPN exits the petrous temporal bone via greater petrosal foramen to the middle fossa passing deep to the trigeminal ganglion and down the foramen lacerum to the pterygoid canal (vidian vanal) where it joins the deep petrosal nerve (sympathetic fibres from the plexus that surrounds the ICA) to form the nerve of the pterygoid canal. This nerve goes to pterygopalatine fossa where the pterygopalatine ganglion is suspended from the V2 nerve. Fibres from this ganglia travel with V2 to the lacrimal gland and mucosa of the nose and mouth. and **(2)** via the chorda tympani nerve which arises just proximal to the stylomastoid foramen, travels through the petrotympanic fissure and joins the lingual branch of V3, one cm below the foramen ovale and then via submandibular ganglion (suspended from the lingual nerve) supplies the submandibular and sublingual glands for salivation.

The general somatic afferent fibres carries the sensation from the posterior wall of the external auditory meatus and the back of the ear via the geniculate ganglion, at the facial genu in the petrous temporal bone, to the spinal trigeminal tract.

The special visceral afferent fibres carry taste sensation from the anterior two thirds of the tongue to the geniculate ganglion via the chorda tympani nerve and then to the rostral part of the nucleus solitaris.

Taste sensibility is composed of four qualities: sweet, salt, sour, and bitter. Taste receptors are located on the tongue, palate, pharynx, and larynx. Although all four qualities can be perceived throughout, there is considerable localization. The tongue, especially at the tip and edges, is most sensitive to sweet and salt and the palate, to sour and bitter. The receptors are taste buds. Up to 8 receptors are present on each of the fungiform papillae on the anterior two-thirds of the tongue, and up to 100 on each of the circumvallate papillae on the posterior part of the tongue. The taste buds are barrel shaped with a pore/opening. Chemoreceptive taste hairs projecting into the barrel from neuroepithelial sensory cells. Impulses from these cells are transmitted to the brainstem. Afferent fibers from the anterior two-thirds of the tongue travel via the lingual nerve to the chorda tympani and then as described above to the gustatory nucleus. Taste fibers from the posterior third of the tongue, the palate, and the palatal arches travelling via the glossopharyngeal nerve and the nodosal ganglion, also end in the gustatory nucleus. There are two ascending pathways from the gustatory nucleus. One goes to the hypothalamus. The other goes to the thalamus and then to the gustatory center of the cortex, which is probably area 43 in the parietal operculum.

Summarizing,

The facial nerve should be considered to have **three** components, a brachiomotor nerve, a secretomotor nerve and a taste nerve. In addition to moving the face, the facial nerve mediates (1) *tearing*: the parasympathetic axons to the lacrimal gland via the pterygopalatine ganglion (2) *snotting*: parasympathetic axons to the nasal mucosa via the pterygopalatine ganglion(3) *tasting*: taste from the anterior two-thirdsof the tongue via the geniculate ganglion (4) *salivating:* parasympathetic axons via the submandibular ganglion.

Examination of facial nerve:

The muscles of the lower face are controlled by the contralateral hemisphere, whereas those in the upper face receive control from both hemispheres (bilateral representation). Hence a lower motor neuron lesion paralyses all facial muscles on that side, but an upper motor neuron (supranuclear) lesion paralyses only the muscles in the lower half of the face on the opposite side.. Subtle weakness is often difficult to confirm. Many, perhaps even most, normal individuals have mild lower facial asymmetries, making interpretation difficult.

Function of the facial nerve includes (a) expression of emotions, such as when frowning and smiling (b) compression of lips for whistling, blowing and spitting, labial sounds of speech, swallowing and other feeding actions (c) controlling and protecting facial apertures: the palpebral fissures, oral fissure, anterior nares, lips, and external auditory canals (d) dampening excessive movement of the ossicles by stapedius muscle contraction on hearing loud sounds. After stapedius paralysis, ordinary sounds may seem uncomfortably loud, a symptom called hyperacusis

Except for the mandible and eyelid elevation, facial nerve innervates every other movements that the face can make, Careful and thoughtful observation is the key to discerning subtle signs of weakness of muscles supplied by the motor portion. Note especially the blink, nasolabial folds, and corners of the mouth. Asymmetry is the clue to unilateral weakness and is best perceived during conversation when the patient is unaware of being observed.

1. Blink: The eyelid on the affected side closes just a trace later than the opposite eyelid.
2. Nasolabial folds: The weak one is flatter.
3. Mouth: The affected side droops and participates less in speaking.

Ask the patient to look up or wrinkle the forehead, inspect for asymmetry. Ask him or her to close the eyes tightly. Look for incomplete closure or incomplete "burying" of the eyelashes on the affected side. Observe the nasolabial folds and mouth while the patient is concentrating on the eyes. As the orbicularis oculi contract tightly, there are milder associated contractions of muscles about the mouth and nose, these milder contractions are better suited to displaying slight weakness than when these muscles are tested directly.

Ask the patient to smile, show teeth, or pull back the corners of the mouth. Look for asymmetry about the mouth.

The most subtle signs of mild facial weakness are the blink reflex and incomplete lid closure. Observe the blink reflex during conversation, or tap gently on the glabella with your index finger or reflex hammer in an attempt to bring out a mild asymmetry of blink. If you strongly suspect but are having difficulty confirming a mild facial weakness, ask the patient to lie flat on the examining table with face up. Slide the patient's head off the examining table so the head is below the body. This forces the eyelids to work against gravity. Now ask the patient to close both eyes and inspect for incomplete closure. Tap on the glabella and note asymmetry of blink.

Summary of tests of facial muscles

Examiners Command	Observation	Muscle tested
1.Wrinkle up forehead or look at the ceiling.	Inspect for asymmetry	Frontalis
2. Close the eyes tight and don't let examiner open.	Inspect for asymmetry of wrinkles	Orbicularisoculi
3. Pull back the corners of mouth, as in smiling.	Inspect for asymmetry of nasolabial fold	Buccinator

4.Wrinkle up the skin on neck or pull down hard on the corners of mouth.	Inspect for asymmetry	Platysma

By knowing the degree of unilaterality of various facial movements, one can unravel the pattern of UMN paralysis.

Tests for unilaterality of facial movements

Movement	Result
1. Retraction of one corner of mouth	Movement is unilateral. Every normal person can do it.
2. Wink one eye at a time	Most can do it, but some cannot wink one eye without the other. When one eye winks, the opposite orbicularis oculi contracts to some degree.
3. Elevate one brow at a time	Few can do it unilaterally, but everyone can elevate them together. The

movement is essentially bilateral.

Gradient of unilaterality

The most free unilateral facial movement normally is lip retraction. The least free unilateral facial movement normally is forehead elevation. The utility of various facial movements explains the gradient of unilaterality. When eating one makes unilateral movements to manipulate and clear food from ones cheeks (buccinators muscle). A major discomfort of facial palsy is that food lodges in the cheek. Unilateral forehead movements offer no such utility, and we usually activate both sides of forehead equally. Although the eyes usually blink together, sometimes you need to close only one. Thus the utility of unilateral eyelid action falls between that of mouth retraction and forehead wrinkling. Body parts such as hand and lip that have the most free, most independent unilateral movements receive their UMN innervations mainly from the contralateral hemisphere. Whereas proximally, symmetrical movements such as chewing and swallowing receive about same number of UMN axons from each hemisphere 50/50. The most free independent unilateral movements are innervated by crossed and uncrossed axons in a ratio 90/10 while for movements with an intermediate degree of unilateral independence the ratio will be 60/40 and so on. So the pattern of facial muscle weakness after unilateral destruction of one corticobulbar pathway will be maximum paralysis of lip retraction. The least paralysis will be that of forehead wrinkling.

Lesson learned After a large acute UMN lesion such as a massive cerebral infarct, eyelid closure is usually paretic (incomplete paralysis), along with paralysis of lip retraction. Rarely even the frontalis muscle is somewhat paretic. Because such a patient has eyelid closure, the examiner who does not understand the gradient of unilaterality of facial movement may erroneously diagnose a lesion of the ipsilateral facial nerve rather than the contralateral corticobulbar tract. In the acute phase shortly after a severe UMN lesion, lip retraction contralateral to the lesion will be paralysed during volitional movement and also during

emotional expression such as smiling. In chronic phase of the UMN lesion, lip retraction may remain weak during volitional action, but may be prominent during emotional expression.

Taste: The four primary tastes are bitter, sweet, sour, and salty. Screen for disorders of sweet or salty taste with salt and sugar. With the patient's eyes closed and tongue protruded, take a tongue blade and smear a small amount of salt or sugar on the lateral surface and side of the tongue. Instruct the patient to tell you the identity of the substance. Rinse the mouth thoroughly and repeat the test on the other side, using a different substance

The facial nerve participates in a number of **reflexes.** Assessment of these reflexes provides valuable additional information about facial nerve function. Some of these reflexes are listed below.

Orbicularis oculi reflex Percussion causes reflex contraction of the eye muscle. The reflex is known as the supraorbital, glabellar, or nasopalpebral reflex, depending upon the site of the stimulus. Both eyes usually close, with the contralateral response being weaker. The trigeminal nerve is the afferent and the facial nerve the efferent of the reflex. Light and sound can also produce the reflex, with the optic and acoustic nerves providing the afferent. The response is weak or abolished in nuclear and peripheral lesions, and present or exaggerated in supranuclear lesions. It is also exaggerated in Parkinsonism and cannot be voluntarily inhibited.

Palpebral – oculogyric reflex The eye balls deviate upward when the eyes are closed., both when awake and asleep. The afferent arc is proprioceptive impulses carried through the facial nerve to the MLF. The oculomotor nerve to the superior rectus muscles forms the efferent arc. In peripheral and nuclear lesions an exaggeration of this reflex is seen, known as Bell’s phenomenon.

Orbicularis oris reflex Percussion on the side of the nose or the upper lip causes ipsilateral elevation of the angle of the mouth and upper lip. The reflex arc is composed of the fifth and seventh nerves. Synonyms: nasomental, buccal, oral or perioral reflex This reflex disappears after about the first year of life, recurring with supranuclear facial nerve lesions and with extra-pyramidal diseases such as parkinsonism.

Snout reflex Tapping the upper lip lightly with a reflex hammer, tongue blade or finger causes bilateral contraction of the muscles around the mouth and base of the nose. The mouth resembles a snout. This is an exaggeration of the orbicularis oris reflex. It is present with bilateral supranuclear lesions and in diffuse cerebral diseases such as various causes of dementia.

Suck reflex Sucking movements of lips, tongue, and mouth are brought about by lightly touching or tapping on the lips. At times, merely bringing an object near the lips produces the reflex. Occurs in patients with diffuse cerebral lesions. The snout reflex occurs in similar circumstances.

Palmomental reflex A stimulus of the thenar area of the hand causes a reflex contraction ipsilaterally of the orbicularis oris and mentalis muscles. A number of normal individuals have this reflex, and also patients with diffuse cerebral disease. It is significant when other similar reflexes are also present.

Corneal reflex Stimulation of the cornea with a wisp of cotton produces reflex closure of both ipsilateral (stronger) and contralateral eyelids. The fifth nerve carries the afferent impulse, and the facial nerve the efferent impulse.

Summary

A lesion involving the nuclear or infranuclear portion of the facial nerve will produce a peripheral facial palsy. If all motor components are involved, there is complete paralysis of all facial muscles on the involved side. The brow is smooth, the eye does not close, the nasolabial fold is flat, and that side of the mouth droops. There is no movement at all. The paradigm of this type of involvement is Bell's palsy. Idiopathic Bell's palsy may strike at any age, often after a mild viral illness. Recovery is over a period of weeks to months and is variable. The cause of the idiopathic variety is unknown. Sequelae to Bell's palsy include the following:

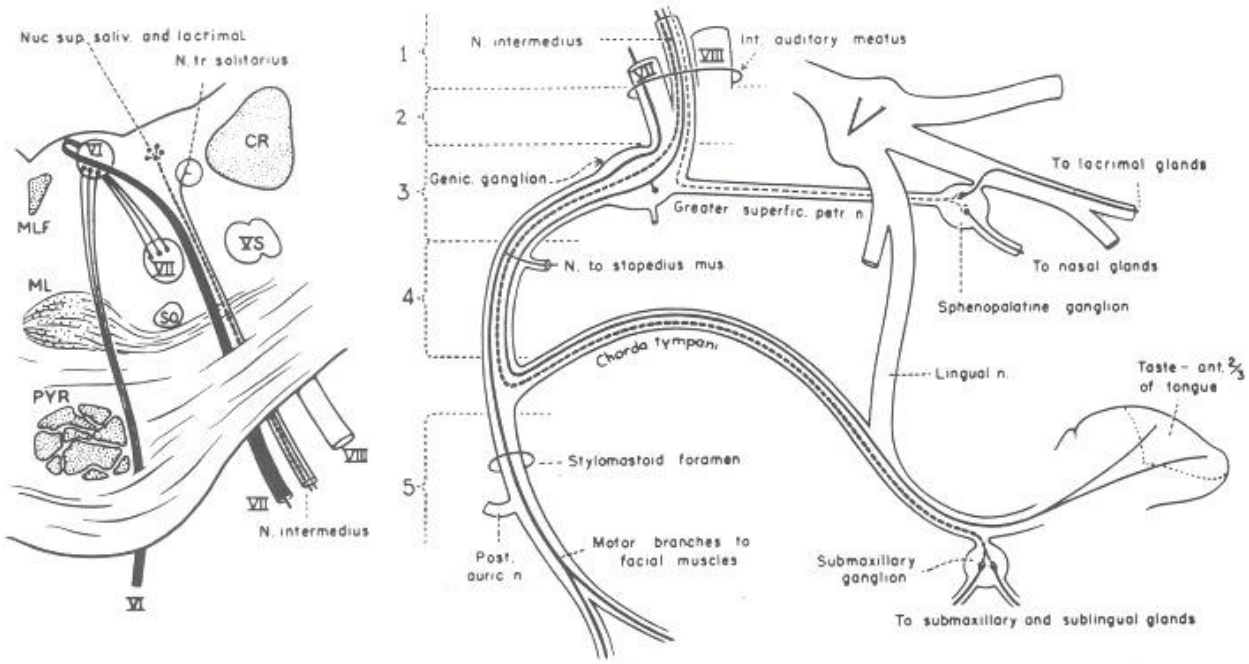
1. Interfacial synkinesis: When the eyes close, the mouth will twitch. This occurs when the regenerating nerve fibers do not grow back into the proper muscles. The synkinetic movements are almost always present on the involved side.

2. Because of the contractures, the face at rest may be more deeply etched on the side of the previous palsy. This can give a false impression of weakness on the opposite side.

Other causes of peripheral seventh nerve palsy include: neoplasm, trauma, middle ear infections, parotid gland surgery, granulomatous or carcinomatous meningitis, and diabetes. The disturbances of function produced by these lesions need not be complete. An important clinical point is that the clinical manifestations of these disorders are indistinguishable from idiopathic Bell's palsy.

Localization of lesion

Anatomic localization of lesions is made by the characteristics of the dysfunction and associated structures involved. Picture and the table given below depicts the location of lesions and the clinical manifestations.



Localization of Facial Nerve Lesions

Lesion location	Manifestations
Above the facial nucleus (Supranuclear lesion)	Contralateral paralysis of lower facial muscles with relative preservation of upper muscles. Lesion located either in brainstem or cortex.
Pons (nuclear or fascicular lesion)	Ventral pontine lesion (of Millard-Gubler): ipsilateral facial monoplegia, lateral rectus palsy (VI), contralateral hemiplegia (corticospinal fibers). Pontine tegmentum lesion (of Foville): ipsilateral facial monoplegia, contralateral hemiplegia (corticospinal fibers), paralysis of conjugate gaze to side of lesion (pontine paramedian reticular formation).
Cerebellopontine angle (peripheral nerve lesion). 1 in figure	Ipsilateral facial monoplegia, loss of taste to anterior two-thirds of tongue, impairment of salivary and tear secretion, hyperacusis (if VIII is not affected). Additional cranial nerves may be involved: deafness, tinnitus, vertigo (VII): sensory loss over face and absence of corneal reflex (V), ipsilateral ataxia (cerebellar peduncle).
Facial canal between internal auditory meatus and geniculate ganglion (peripheral nerve type lesion here and subsequently). 2 in figure	Same as above except cranial nerves other than VII are not involved.
Facial canal between geniculate	Facial monoplegia, impaired salivary secretion, loss of taste,

Lesion location	Manifestations
ganglion and nerve to stapedius muscle. 3 in figure	hyperacusis.
Facial canal between nerve to stapedius and leaving of chorda tympani. 4 in figure	Facial monoplegia, impaired salivary secretion, loss of taste.
After branching of chorda tympani. 5 in figure	Facial paralysis, distribution related to site of lesion.

Conclusion

Branches of the facial nerve (proximal to distal)

(a) greater superficial nerve (just before the geniculate ganglion)

(b) nerve to stapedius

(c) chorda tympani

(d) motor branches: **T**emporal, **Z**ygomatic, **B**uccal, **M**andibular, **C**ervical branches, (**Mnemonic**: "Ten Zebras Bit My Clock")

VII CN Lesions

(a) Upper motor neuron(UMN): Upper half of face has bilateral innervation. Therefore a UMN lesion affects only the contralateral face.

(b) Lower motor neuron: LMN lesion affects the entire ipsilateral face, decreased sensation and taste, impaired salivation and lacrimation, and hyperacusis depending upon exactly where the lesion is in relation to branches of the nerve.

(c) Mimetic or emotional innervations: involuntary contraction of the face can occur with emotion even after a corticobulbar lesion.

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BEDSIDE HEARING ASSESSMENT

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Bedside hearing assessment is useful for assessing

1. Type of hearing loss – conductive/ sensorineural/ mixed
2. Degree of hearing loss
3. Approximate site of pathology

Limitations

1. Not accurate
2. Subjective test
3. Requires patient's cooperation
4. Cannot be performed in unconscious patients and children <5 year olds.

I. VOICE TEST (FREE-FIELD SPEECH TEST)

- Can usually detect hearing loss >30 dB with a false positive rate of 13%
- Procedure:

  Test is done with patient facing forward and examiner stationed opposite to the test ear or behind the patient.

  Patient should not be able to see examiner's lip movements.

  Non-test ear should be masked by pushing opposite ear's tragus in and out or by rubbing paper between fingers over the non-test ear.

  Masking is done to prevent participation of the non-test ear in the test.

  Letter number combinations are used as this has a reasonable mix of consonants that allows a relatively broad range of frequencies to be tested.

  Test is performed at 2 feet (one arm distance) and then at 6 inches. And the voices used are loud voice, conversational voice and whisper.

  Whisper: at the end of forced expiration

  Conversation: at the end of normal expiration

  Loud voice: at the end of forced inspiration

If the patient can hear a whisper at 2 feet the hearing is considered normal.

- Interpretation:

Sound	Hearing threshold	
	2 feet	6 inches
Whisper	≥12 dB	≥36 dB
Conversation	≥48 dB	≥54 dB
Loud	≥72 dB	

- If the patient cannot hear whisper at 2 feet his hearing threshold will be greater than 12 dB and if he cannot hear loud sound at 2 feet his threshold will be greater than 72 dB.

II. TUNING FORK TESTS

Tuning forks used for hearing assessment: 256, 512 and 1024 Hz.

512 Hz tuning fork is the best as

- the frequency falls in the range of speech frequency
- sound lasts longer (1024 Hz has a faster decay)
- produces less overtones* (256 Hz produces more overtones)

*Overtones are frequencies above the fundamental frequency

Tones < 256 Hz tend to enhance perception by the production of vibrations.

Activation of tuning fork:

- Struck at a point about one-third of the length of the prong from the free end (to minimize overtones)
- Struck against a firm surface like the elbow or the thenar eminence of the palm and not a hard surface like a table (to minimize overtones and prevent internal fractures in the tuning fork)
- If the vibrations are felt in the stem of the tuning fork, it indicates production of overtones.

1. RINNE'S TEST

- False positive rate 20%

- Procedure:

  2 methods:

  i. Relative Loudness Method:

  256 Hz tuning fork is used first. Tuning fork is activated and placed in front of the external auditory canal (air conduction) and then over the mastoid (bone conduction) and the patient is asked which one was louder.

  Repeated with 512 and 1024 Hz

  ii. Threshold Comparison Method:

  Strike the tuning fork against the elbow and place it over the mastoid process of the patient and when he stops hearing the sound place it in front of the external auditory canal. If the patient can still hear it indicates that the air conduction is better than the bone conduction.

- Interpretation:
  - Rinne's positive: AC>BC →Normal &Sensorineural hearing
  - Rinne's negative: BC>AC →Conductive hearing loss

- False negative: Profound ipsilateral hearing loss (Patient does not perceive any sound by air conduction but responds to bone conduction due to transcranial stimulation of the contralateral cochlea.)

- Quantification of hearing loss based on Rinne's test:

Rinne Test Result	**Estimated Conductive Loss (dB)**
Negative, 256 Hz Positive, 512 Hz and 1024 Hz	Mild conductive loss of 20-30
Negative, 256 Hz and 512 Hz Positive, 1024 Hz	Moderate conductive loss of 30-45
Negative, 256 Hz, 512 Hz, 1024 Hz	Severe conductive loss of 45-60

2. WEBER'S TEST

- Low sensitivity and specificity

- Procedure

  Only 512 Hz tuning fork used. Activated tuning fork is placed on the vertex/ root of nose/ upper central incisors. The patient is asked which ear hears the sound better.

- Interpretation

  Normal: Central

  Conductive deafness: lateralized to the worse ear

  Sensorineural deafness: lateralized to the better ear

- Weber's lateralizes at a difference in threshold of only 5 dB between the two ears.

- Causes of lateralization in conductive deafness
    1. Ambient noise theory:

       In conductive hearing loss ambient sounds present in the atmosphere are not heard and hence, the tuning fork is heard better.
    2. Theory of dispersion:

       When sound from the vibrating tuning fork reaches the middle ear it disperses in all directions – towards the cochlea and towards the external auditory canal. In conductive hearing loss, the sound does not get dispersed to the exterior due to the middle ear pathology.

3. ABSOLUTE BONE CONDUCTION TEST

- Prerequisite: Examiner has normal hearing

- Procedure

  Press the tragus and place the vibrating tuning fork on the mastoid. Ask the patient to raise his hand when he stops hearing the sound and then the tuning fork is transferred to the mastoid of the doctor.
- Interpretation

  Doctor can hear the sound → bone conduction of patient is reduced (SNHL)

  Doctor cannot hear sound → bone conduction is normal

EXAMINATION OF THE VESTIBULAR SYSTEM

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INTRODUCTION

The ability to stand, walk and hold the eyes and head steady and with grace is one of the important abilities that defines us as *homosapiens sapiens*. Various neural circuits are at work, most of them at reflex level to keep us constantly upright and to hold the eyes steady when the head and body are in motion. These reflexes have evolved considerably in us. The various structures which are involved in the balancing act of the body with respect to the environment are grouped together and referred to as the vestibular system. This system is in turn intricately associated with the cerebellum, which also has a refining action on the vestibular system.

Various disease processes may affect this system which gives rise to symptoms and signs. These symptoms may be as common as vertigo which may have an underlying benign pathology such as BPPV (benign positional paroxysmal vertigo) or much serious condition such as acoustic neuroma or lesions of the CNS. Therefore it is essential to determine the cause of vertigo and to localise its origin to either peripheral or a central dysfunction. This requires a clinician to be able to take a good and relevant history and then to perform a few simple tests to examine the vestibular apparatus.

The examination of the vestibular system however requires a lot of dedication right from the beginning of history taking to the performance of bedside tests.

This chapter tries to highlight the important and relevant clinical methods to examine this unique balancing system of our body. To proceed further it is necessary to briefly recapitulate the salient anatomical and physiological features of the vestibular system. Following which the clinical approach to examine the vestibular system are highlighted.

BASIC ANATOMIC AND PHYSIOLOGICAL REVIEW

The vestibule, the vestibular nerves and their central connections are the relevant structures in the act of balancing. However the role of the cerebellum and the cortex as a whole to fine tune the system cannot be overlooked.

The *utricle, saccule and the three semicircular canals* in the inner ear(membranous labyrinth) communicate with each other via the vestibule of the labyrinth. The sensory specialised epithelium of the labyrinth is present as raised structures at the ampulla of the semicircular canals and the utricle and saccule. At the opening of the semi-circular canals these are called the *cristae ampullaris* and at the opening of the utricle and the saccule they are called the *maculae acusticae*. The specialized sensory epithelium of the labyrinth has microvilli called the hair cells which are embedded in the otolithic membrane of the maculae and in the cupula of the cristae. The otolithic membrane is made of calcium carbonate crystals and the cupula is composed of a gelatinous mass. The endolymph continuously bathes the utricle, saccule, the semicircular canals and the sensory epithelium with its hair cells. Any change in the direction or speed of flow is transmitted to the hair cells as kinetic energy, which in turn converts it to action potential to be carried and transmitted via the vestibular nerve.

The vestibular nerve consists of axons of bipolar neurons, the cell bodies of which are located in the Scarpa's ganglion situated inside the internal auditory meatus (IAM). The peripheral processes originate in the cristae and the maculae of the inner ear. The central processes of the bipolar cells continue as the vestibular nerve proper traversing the IAM along with the cochlear nerve, the 7^{th} cranial nerve and the intermediate nerves. Within the meatus the vestibular and the cochlear fibres together form the 8^{th} cranial nerve. The 8^{th} nerve complex exits the IAM, traverses the Cerbello-Pontine angle space and enters the brainstem at the junction of the pons and the medulla, more precisely between the inferior cerebellar peduncle and the olive. The central connections of the vestibular nerve end in the vestibular nuclei which are located in the floor of the 4^{th} ventricle.

Not all fibres of the nerve end at the nuclei, some directly pass onto the cerebellum via the juxtarestiform body forming the vestibulocerebellar tract. This tract terminates in the flocculonodular lobe and the vermis of the cerebellum. Fibres from here now crisscross via the juxtarestiform body to the ipsilateral vestibular and fastigial nuclei as well as to the opposite vestibular nuclei thereby creating a system via which each cerebellum controls and fine tunes both vestibular nuclei inputs and outputs.

The vestibular nuclei are four in number. They are named as superior (angular nucleus of Bechterew), lateral (nucleus of Deiters), spinal (descending nucleus of Roller) and medial (triangular nucleus of Schwalbe). The superior and the medial vestibular nuclei are involved in vestibuloocular reflexes. The fibres from these nuclei as well as some from the remaining two nuclei pass along the Medial Longitudinal Fasciculus (MLF) to connect with 3rd, 4th and 6th cranial nerve nuclei to control eye movements in conjugation with the changes in the endolymph flow in the inner ear. The head and neck movements are believed to be due to the connections of the vestibular nuclei with the 11th cranial nerve.

The medial vestibular nuclei and the lateral via the uncrossed lateral vestibulospinal tract and crossed medial vestibulospinal tracts play an important role in maintaining posture. It is postulated that the medial tract acts on the axial muscles and the lateral tract acts on the limb muscles to bring about proper posture and tone in the balancing act of the body. The descending nuclei are closely associated with the cerebellum by providing information to the cerebellar nuclei.

All vestibular nuclei send efferents diffusely to the pontine reticular formation. They also receive many inputs from the reticular formation.

The projections of the vestibular nuclei further rostrally to the cortex is established, however the exact sites of these projections are still debatable. Most anatomists and physiologists agree that many projections terminate at the superior *sylvian gyrus* and the *intraparietal sulcus*. Some argue that the vestibular nuclei projections also involve the thalamus.

With all the above connections in place and working fluently, when there is a linear acceleration (detected by the maculae) or an angular acceleration (detected by the cristae of the semicircular canals) the impulses are picked by the peripheral fibres of the bipolar neurons and transmitted rostrally. The action of each vestibular apparatus on either side may be summarised as pushing the eyes, limbs and other structures the nuclei innervate, to the opposite side. With both the sides acting correctly in tandem the structures are kept in balance in the midline. In the event of destruction or dysfunction of the system on one side the opposite side dominates and hence the eyes, limbs and other structures deviate towards the diseased labyrinth.

This is the basic physiology in a simplified form which helps in understanding the various symptoms and signs during examination of the vestibular system and assists, a clinician, in accurately localising diseases or lesions of the vestibular system.

EXAMINATION OF THE VESTIBULAR SYSTEM

History

Like in all other systems the clinical examination begins with a good history taking of the patient's complaints. The problem with vestibular system is that the most important presenting history is vertigo, the description and definition of which is extremely varied and subjective. It becomes very important for the clinician to sieve out the correct associations of the patient's complaints and thus arrive at the conclusion as to whether the patient has features of vertigo or pseudovertigo (giddiness).

Vertigo may be defined as *any illusion of motion*, whether subjective or objective that a patient may perceive. The cause of vertigo, may be, due to an imbalance between the various sensory inputs that the proprioceptive organs such as retina, labyrinth, muscles and joints, are subjected to, leading to an illusion of movement. A good example being, what is felt when sitting in a stationary train compartment when the adjacent train starts moving.

Sometimes it is evident from the history itself when the patient states that the environment seems to spin around or vice versa. Many clinicians make a distinction between subjective vertigo with the feeling of the self-spinning or objective vertigo when the objects in the surrounding environment seem to move around the patient. When patients are not as articulate as above then they may use various phrases such as felling of to and fro movements, imbalance while walking, felling of being drawn towards one side as if by a magnet, illusion of the walls or the ceiling tilting up down or sideways. Subtle points during the history narrative such a desire to lie down or sit down or keep still or avoid certain forms of postures like bending or leaning forwards or sideways are often clues to a diagnosis of vertigo. A history of worsening of symptoms on, turning in bed, sudden turns while walking may also help. However a patient complaining of dizziness which does not worsen on sudden movements of the head is unlikely to be vertigo. True vertigo is also associated with symptoms such as tinnitus, hearing loss, vomiting and is relieved by keeping still.

Pseudovertigo on the other hand may be associated with conditions of anxiety, panic attacks, hyperventilation. Patient's do not complain of the characteristic features of vertigo such as

pulsion, rotation, up and down and other abnormal types of motion along with associated symptoms as mentioned above. Vague terms such as light headedness, giddiness, drunkeness are often described by the patient.

Thus, a careful history is of utmost importance in differentiating pseudovertigo and vertigo and is the first critical step in testing the vestibular system.

Examination

Vertigo as a symptom is primarily a disorder of the vestibular apparatus, the vestibular nerve and the vestibular nuclei. Although lesions of the cerebral cortex or the eyes or the muscles or the peripheral nerves may all have vertigo but vertigo is not the dominant symptom and other constellation of symptoms predominate. Tests to detect vestibular dysfunction include tests for vestibuloocular reflexes (VOR), vestibulospinal reflexes (VSR) and observation of nystagmus. Together they are often termed as tests to detect Labyrinthine dysfunction.

Tests for VOR

The VOR has evolved in order for the eyes to remain fixed at an object of interest when the head and/or body are in motion. This can be tested by:

1. Caloric tests**:** As observed earlier the vestibular apparatus works such that the right side pushes the eyes, limbs and the body to the opposite side. The patient's head is positioned such that the horizontal semicircular canal is vertically aligned. This is achieved by tilting the head forwards by 30 degrees. The ear under evaluation is first irrigated with water at 30 degrees Celsius for 30 seconds followed by a gap of 5 minutes and then by water at 44 degree Celsius. In normal individuals when the right ear is irrigated with cold water at 30 degree Celsius then the effect produced is similar to creating a lesion of the right labyrinth. As a result of the absence of the right vestibular apparatus the eyes are slowly pushed towards the right side by the left sided functioning labyrinth. This aberrant movement is corrected within 20 seconds by a fast movement of the eyeballs to the opposite side-- nystagmus to the opposite side. Warm water induces similar movements to the same side, that is the fast component or correcting nystagmus is towards the side of lesion and the slow component to the opposite side of the lesion. The pneumonic COWS---COLD OPPOSITE WARM SAME helps to remember the above. In case both the ears are irrigated with cold water together

then the eyes move downwards slowly with nystagmus upwards and the opposite for warm water. In comatosed patients large volumes of waters around 30-40 cc are used to ensure absolute success of stimulation. This test is one of the most reliable tests to elicit labyrinthine dysfunction and also to discriminate the side of lesion.

2. Doll's eye reflex or oculocephalic reflex**.** This test is used to identify whether the pathways from the vestibular nuclei via the MLF in the pons to the oculomotor nuclei in the midbrain are functioning or not. Especially helpful in comatosed patients. The test is performed by slow sideways movements of the head of the patient and watching for the movement of the eyes. In patients with intact pathways the eyes move towards the opposite direction.
3. Head thrust**:** This test is done in awake patients. Here the subject is asked to fix his gaze at a particular object and the examiner, after explaining the procedure, turns the head rapidly sideways. In normal individuals the gaze is fixed at the object. In subjects where there is a dysfunction, the eyes lag behind and a rapid compensatory movement is often seen to fix the object.
4. Dynamic visual acuity**:** The normal VOR allows us to read while the head is in motion, to some extent. This is objectively tested here by asking the patient to read from a Snellen's test chart while the head is being moved to and fro. Any fall of visual acuity values below three Snellen's chart lines from, the baseline for, the patient is indicative of labyrinthine dysfunction.

Tests for VSR

The tests described here are based on the same principle that the vestibular system normally pushes the body or the limbs to the opposite side. The tests described, detect movements of the body or the limbs in labyrinthine dysfunction. In all these tests the eyes are kept closed as visual fixation is often able to suppress and compensate for the labyrinthine dysfunction.

1. Past pointing**:** The subject is asked to outstretch his arm in front and touch the examiners tip of the index finger with his index finger. After he has brought his index finger in contact with the examiners, he is asked to close his eyes and raise his arm up straight above and then bring it down to touch the tip of the examiners finger. In patients with acute labyrinthine dysfunction the arm sways towards the

diseased labyrinth and completely misses the target. The test may not be positive in patients with chronic labyrinthine dysfunction where patient has learned to compensate by other means.

2. Romberg's test**:** Here the patient is asked to stand with heels and feet close together and asked to close his eyes. In case of labyrinthine dysfunction patient will fall towards the side of the lesion. If the dysfunction is in the right ear and patient falls towards the affected ear. The position of the head changes the direction of fall, in case of right ear if head is turned to the left patient falls forwards and so forth.
3. Star walking**:** The patient is asked to close his eyes and walk several steps forward and backwards. A patient with labyrinthine dysfunction will sway and lean towards the side of lesion thereby creating a star shape with multiple prongs.
4. Fukuda' stepping test**:** Patient is asked to march in the same place with eyes closed. Affected patients tend to turn and face the side of the lesion. In compensated cases this may not be always positive.

Observation for nystagmus

The characteristic feature of vestibular nystagmus is again based on the same physiological principle as above.The slow phase being towards the lesion and the fast phase away. The nystagmus of vestibular origin is typically fine and may be missed if not observed closely. Furthermore it is inhibited by visual fixation and therefore fixation needs to be removed. To achieve this **Frenzel lenses** are employed. These are convex lenses with very high power which not only block fixation but also magnify the eyes for closer observation. Vestibular nystagmus may be spontaneous as above or it may be positional. Positional nystagmus can be elicited by various manoeuvres like the Dix-Hallpike manoeuvre.

Dix-Hallpike manoeuvre: Patient is initially seated and the head is turned to the by 45 degrees in order to align the posterior semicircular canal in the sagittal plane. Patient is rapidly made to lie down with the examiner holding his head at 45 degrees until the head hangs off the examining couch. Within a few seconds patient experiences vertigo and nystagmus is seen. The nystagmus elicited has fast component towards the dependent ear. This type of positional nystagmus is fatigable and is seen in labyrinthine dysfunction such as BPPV.

Labyrinthine dysfunction can also be quantitatively measured by recording the labyrinthine eye responses while performing the tests. This recording can be done with Electronystagmography(ENG) where the patient is either rotated in a swivel chair with acceleration controls or ENG performed along with caloric tests.

All the above tests if performed with dedication and accuracy can correctly diagnose labyrinthine dysfunction and help the examiner clinically localise the site of lesion.

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2. Paul.W.Brazis, *Localisation in clinical neurology*,pg 343-345
3. Allan.H.Ropper, Robert.H.Brown, *Adams and Victor's Principles of Neurology,* pgs 258-268.
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THE CLINICAL EXAMINATION OF LOWER CRANIAL NERVES

(9^{th}, 10^{th}, 11^{th})

Dr Altaf Umar Ramzan, Sher e Kashmir Institute of Medical Sciences, Srinagar, J & K

The glossopharyngeal (CN IX) and vagus (CN X) nerves are intimately related and similar in function. Both have motor and autonomic branches with nuclei of origin in the medulla. Both conduct general somatic afferent (GSA) as well as general visceral afferent (GVA) fibers to related or identical fiber tracts and nuclei in the brainstem; and both have a parasympathetic, or general visceral efferent (GVE) and a branchiomotor, or special visceral efferent (SVE) component. The two nerves leave the skull together, remain close in their course through the neck, and supply some of the same structures. They are often involved in the same disease processes, and involvement of one may be difficult to differentiate from involvement of the other. For these reasons the two nerves are discussed together.

THE GLOSSOPHARYNGEAL NERVE

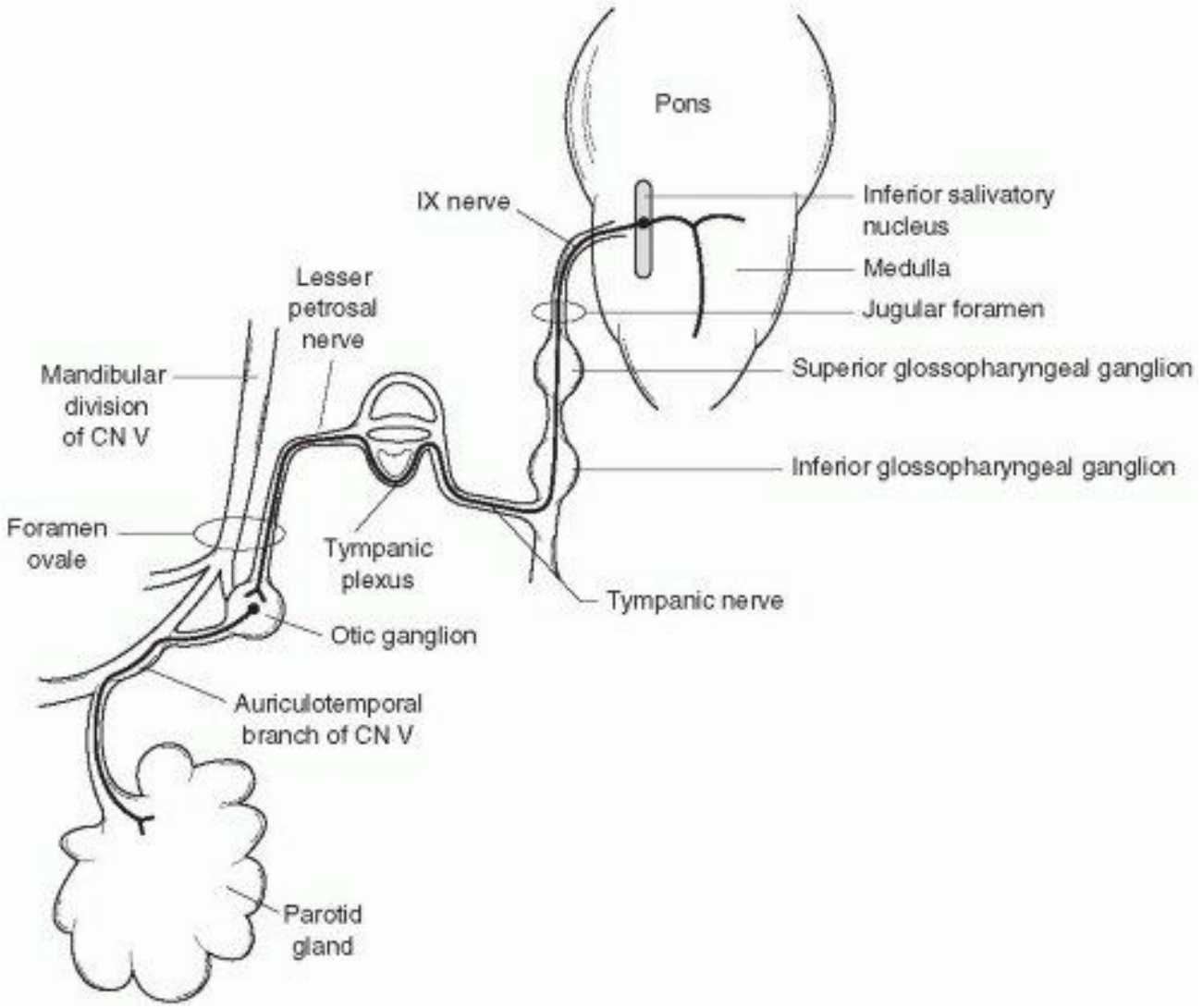


FIGURE: Peripheral distribution of the parasympathetic branches of the glossopharyngeal nerve.

Clinical Examination

Cranial nerve IX is difficult to examine because most or all of its functions are shared by other nerves and because many of the structures it supplies are inaccessible. It is possible to examine pain and touch sensation of the pharynx, tonsillar region and soft palate, and the gag reflex.

MOTOR FUNCTION:

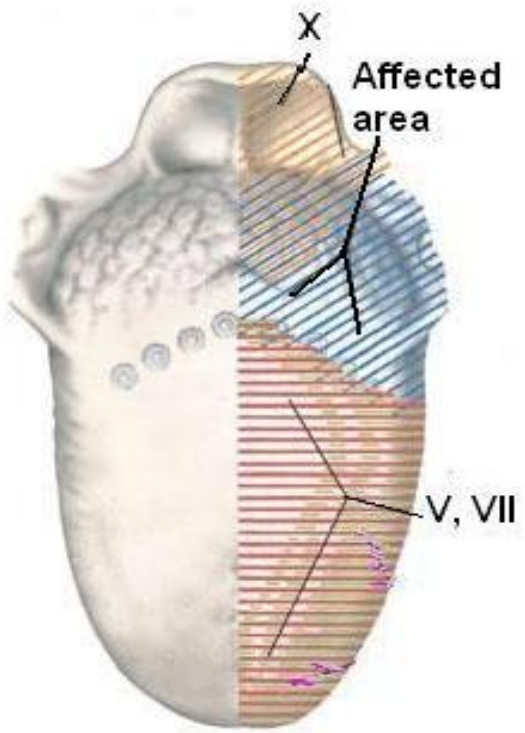
The only muscle to receive its motor innervation purely from CN IX is the stylopharyngeus. The only deficit that might be detectable is a slight lowering of the palatal arch at rest on the involved side.

AUTONOMIC FUNCTION:

The salivary reflex is flowof saliva from the parotid duct after gustatory stimuli. The afferent limb is through taste fibres and theefferent through the parasympathetic outflow of the superior and interior salivatory nuclei.

If highly seasoned foods are placed on the tongue, a copious flow of saliva may be seen to issue from Stensens duct : this may be called the salivary reflex

SENSORY FUNCTION



9th nerve supplies exterioceptive sensations to posterior portion of the tympanic membrane and posterior wall of external auditory canal these regions are supplied by 5th,7th,and 10th cranial nerves also so ,making it irrelevant for 9th c.n examination. However 9th c.n supplies special visceral sensation to the posterior third of the tongue. To test the taste quinine soaked cotton bud is applied to the posterior third of the tongue which gives a bitter taste.

Alternatively taste may be tested by the use of a galvanic current, applying the naked copper electrode to the tongue. The anode is applied to the tongue and a current of 2-4 milliamperes is used which gives a sour metallic taste. Anelectrogustometer,which regulates the galvanic taste stimulus with a potentiometer can be used to determine taste thresholds and carry out quantitative testing.

THE REFLEXES:

The gag reflex is elicited by touching the pharynx or palate. Some sources make a distinction between the pharyngeal reflex and the palatal reflex, referring only to the former as the gag reflex. In common clinical usage, no distinction is made between these two and either is referred to as the gag reflex. The reflex is elicited by touching the lateral oropharynx in the region of the anterior faucial pillar with a tongue blade, applicator stick, or similar object (pharyngeal reflex), or by touching one side of the soft palate or uvula (palatal reflex). The pharyngeal reflex is the more active of the two. The reflex also occurs with touching the base of the tongue or posterior pharyngeal wall. The afferent limb of the reflex is mediated by CN IX and the efferent limb through CNs IX and X. The reflex centre is in the medulla. The motor response is constriction and elevation of the oropharynx.

This causes the midline raphe of the palate and the uvula to elevate, and the pharyngeal constrictors to contract. The activity on the two sides is compared. The gag reflex is protective; it is designed to prevent noxious substances or foreign objects from going beyond the oral cavity. There are three motor components: elevation of the soft palate to seal off the nasopharynx, closure of the glottis to protect the airway, and constriction of the pharynx to prevent entry of the substance.

When unilateral pharyngeal weakness is present, the raphe will deviate away from the weak side and towardthe normal side. This movement is usually dramatic. Minor movements of the uvula and trivial deviations ofthe midline raphe are not of clinical significance. In normal adults, both palatal and pharyngeal reflexes areusually present but there may be inter- and intraindividual variation in the intensity of the stimulus required.

The gag reflex may be bilaterally absent in some normal individuals. Unilateral absence signifies a lower motor neuron lesion. Like most bulbar muscles the pharynx receives bilateral supranuclear innervation, and a unilateral cerebral lesion does not cause detectable weakness.The gag reflex is often used to predict whether or not a patient will be able to swallow. A poor gag reflex in an awake patient with an acute deficit may be a predictor of swallowing difficulties. In fact, the gag reflex has little to do with normal swallowing.

Normal deglutition is a smooth coordinated sequence of muscle contractions that propel a bolus of food from the mouth into the esophagus. A normal swallow bears little resemblance to the chaos of a gag reflex. Higher cortical centers have to inhibit the gag response during normal swallowing. The gag reflex is useful but limited in assessing airway protection.A decreased gag reflex in a patient with depressed consciousness may portend inadequate guarding of the airway and increased aspiration risk, but the status of the gag reflex is not a completely reliable indicator.Patients with an apparently intact gag reflex may still aspirate, and a patient with a depressed gag reflex may not. The trigeminal nerve contributes to palatal sensation, andmay allow for paradoxical preservation of the gag reflex in the face of a CN IX lesion.The gag reflex may be hyperactive in some normal individuals, even to the point of causing retching and vomiting. A hyperactive gag reflex may occur with bilateral cerebral lesions, as in pseudobulbar palsy and amyotrophic lateral sclerosis (ALS).

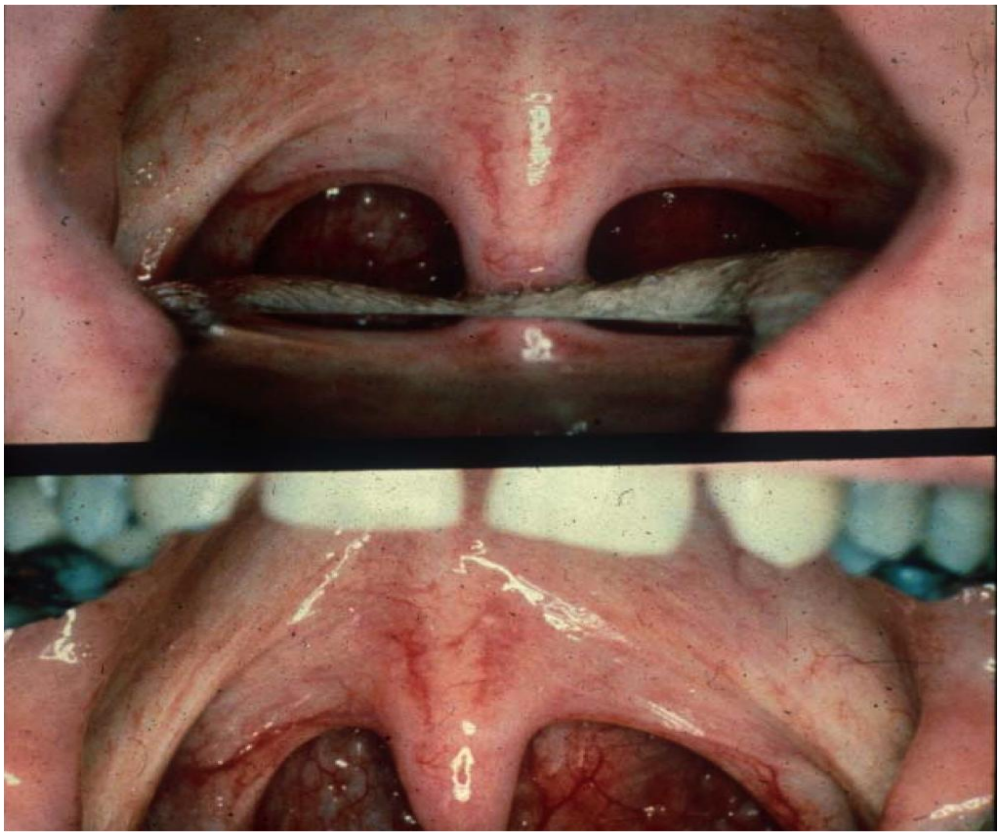


Figure: Lowering of left palatal arch paralysis of left 9th c.n.

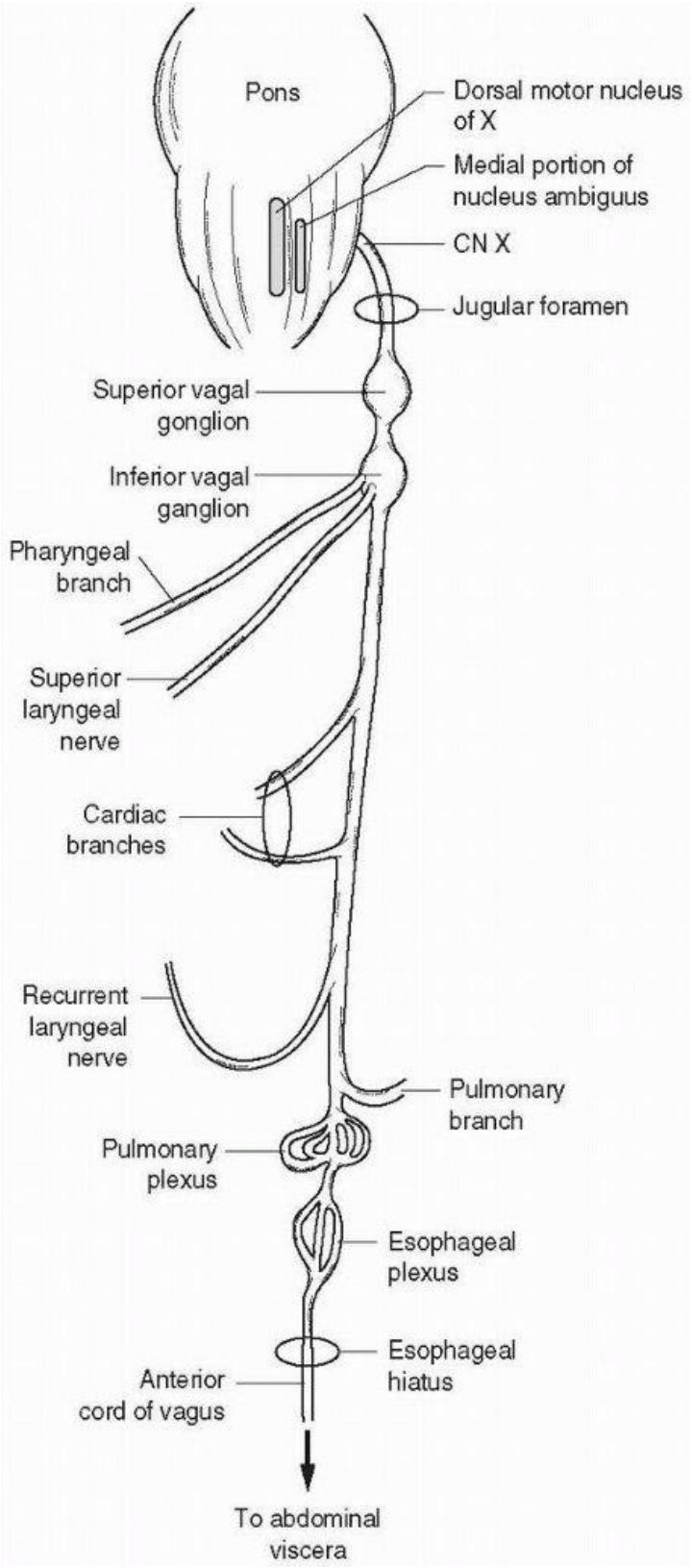


Figure: Peripheral distribution of the branches of the vagus nerve.

THE VAGUS NERVE

Clinical Examination

Examination of the Motor Functions

The motor branches of CN X supply the soft palate, pharynx, and larynx in the same distribution as for CN IX, and are examined in the same manner. The gag reflex is discussed in the section on CN IX.The character of the voice and the ability to swallow provide information about the branchiomotor functions of the vagus. With acute unilateral lesions the speech may have a nasal quality and dysphagia is often present; this is more marked for liquids than solids with a tendency to nasal regurgitation. Examination of the soft palate includes observation of the position of the palate and uvula at rest, and during quiet breathing and phonation. The median raphe of the palate rises in the midline on phonation. With a unilateral lesion of the vagus there is weakness of the levator veli palatini and musculus uvulae, which causes a droop of the palate and flattening of the palatal arch (Figure).Preserved function of the tensor veli palatini (innervated by CN V) may prevent marked drooping of the palate. On phonation, the medial raphe deviates toward the normal side. The palatal gag reflex may be lost on the involved side because of interruption of the motor rather than sensory path.

With bilateral vagus involvement the palate cannot elevate on phonation; it may or may not droop depending on the function of the tensor veli palatini. The palatal gag reflex is absent bilaterally. The tendency toward nasal speech and nasal regurgitation of liquids is pronounced. The speech is similar to that of a patient with cleft palate

Weakness of the pharynx may also produce abnormalities of speech and swallowing. With pharyngeal weakness, dysarthria is usually minimal unless there is also weakness of the soft palate or larynx. Spontaneous coughing and the cough reflex may be impaired. Dysphagia may occur but without the tendency to greater difficulty with liquids and to nasal regurgitation that occurs with palatal weakness. Dysphagia is marked only in acute unilateral or in bilateral lesions. Examination of the pharynx includes observation of the contraction of the pharyngeal muscles on phonation, notation of the elevation of the larynx on swallowing,

and testing the pharyngeal gag reflex. Unilateral weakness of the superior pharyngeal constrictor may cause a “curtain movement” (Vernet's rideau phenomenon), with motion of the pharyngeal wall toward the nonparalyzed side on testing the gag reflex or at the beginning of phonation. The normal elevation of the larynx may be absent on one side in unilateral lesions, and on both sides in bilateral lesions.

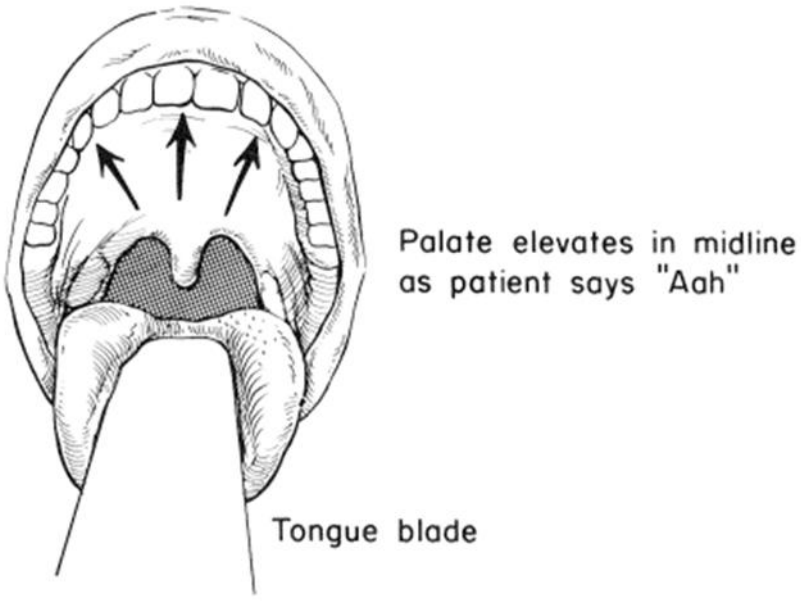


Figure: Normal, symmetrical elevation of soft palate.

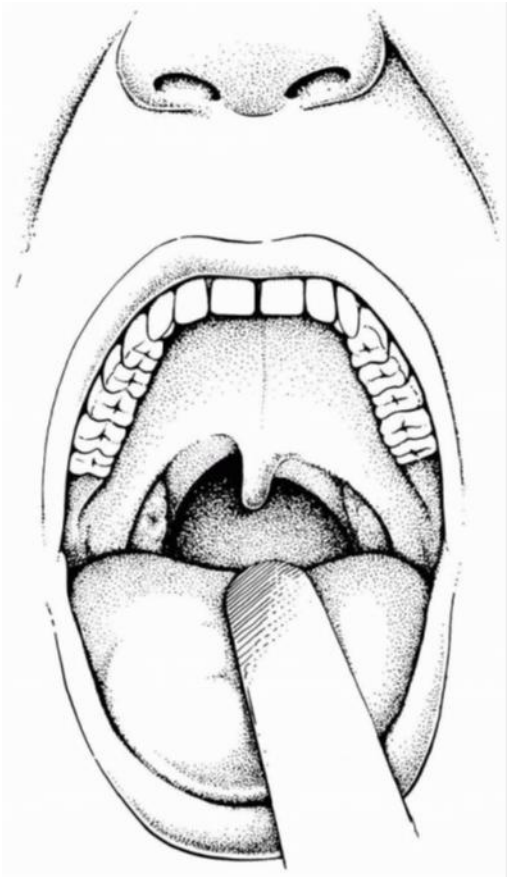


FIGURE: Unilateral paralysis of the soft palate.

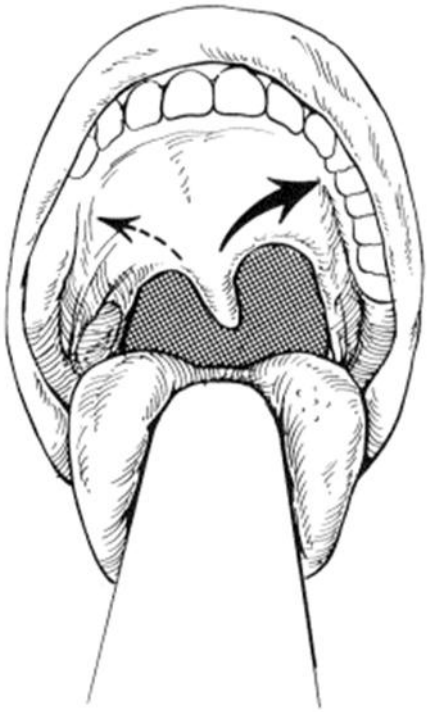


FIGURE: Elevation of palate away from weakened right side (Rideau or curtain sign).

Cranial nerve X innervates the vocal cords. Normal movement of the vocal cords is necessary for three vital functions: breathing, coughing, and talking. During inspiration and expiration, the cords abduct to allow for free air flow; when speaking the cords adduct and vibrate to accomplish phonation. The cords are also adducted when coughing. Movements of the myriad small muscles that control the larynx are complex and have different effects on laryngeal function (Table 18.1). The effects of weakness of the different laryngeal muscles is summarized in Table 18.3. A unilateral lesion of the vagus may cause cord weakness or paralysis.

Vocal cord dysfunction alters the character and quality of the voice, and may produce abnormalities of articulation, difficulty with respiration, and impairment of coughing.

Spasmodic dysphonia is a common focal dystonia that involves the vocal cords and causes characteristic voice changes. Spasmodic dysphonia most often causes abnormal adduction spasms of both vocal cords, and the voice is strained and high-pitched.

Abductor dysphonia is due to spasmodic contraction of the posterior cricoarytenoid, which causes a failure of normal adduction on phonation; the voice is breathy and hoarse. This type of spasmodic dysphonia is most likely to be confused with a lesion of CN X.

The most common cause of vocal cord paralysis is a lesion of one recurrent laryngeal nerve. The

paralysis may evolve from mild abduction impairment due to isolated involvement of the posterior cricoarytenoid to complete paralysis with the cord in the cadaveric position. With slight weakness of the vocal cords or pharynx,hoarseness and dysphagia may be apparent only when the head is turned to either side. Occasionally, even severe weakness of a vocal cord causes little appreciable effect on the voice because of preserved movement of the normal cord.

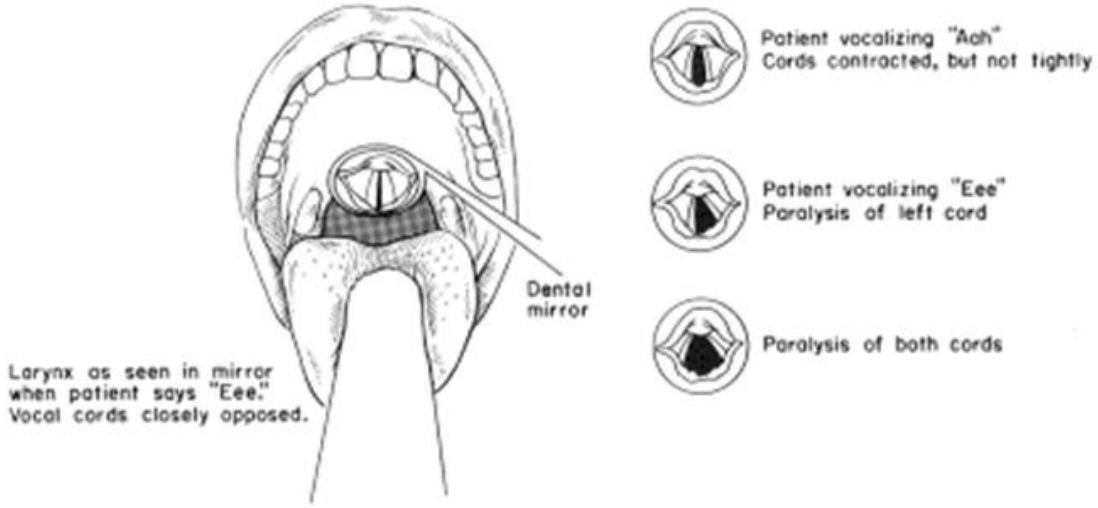


FIGURE: Indirect (through mirror) laryngoscopy. Mirror warmed to avoid fogging by patient's breath.

Examination of the Autonomic Functions

Autonomic Function Testing

Many different procedures have been developed to test the sympathetic and parasympathetic nervous systems.

Tests of cardiac vagal tone include assessment of heart rate variability to deep breathing, standing, and performing Valsalva. The beat-to-beat changes in heart rate in response to autonomic reflexes occur quickly,often too quickly for bedside assessment to be accurate. It is possible at the bedside to determine if heart rate variability with respiration or to Valsalva is present and obvious (probably normal), present but minimal (possibly abnormal), or absent (abnormal). More precise testing requires equipment, and may include an indwelling arterial catheter to follow BP changes. Normal sinus arrhythmia is the beat-to-beat variability in heart rate that occurs with respiration. It is most prominent in healthy young people. Sinus

arrhythmia normally becomes less prominent with age, and it may be markedly impaired or abolished when vagal innervation of the heart is compromised. The heart rate response to deep breathing (HRDB) shows maximal variability at a breathing rate of 5 to 6 per minute. The HRDB can be assessed at the bedside simply by noting pulse variability; it can be measured more quantitatively by measuring the R-R interval with cardiac monitoring. The expiratory to inspiratory ratio quantitates the variability in HRDB. The heart rate response to standing (30:15 ratio) is another method of evaluating the baroreflex arc. The most dramatic changes in HR normally occur in the first 30 seconds after standing, with an initial tachycardia,followed by bradycardia about 20 seconds later. The 30:15 (tachycardia: bradycardia) ratio is the ratio of the R-R interval at beat 30/R-R interval at beat 15; normal is >1.04.The respiratory variability in heart rate is exaggerated when a Valsalva maneuver is performed. The cardiovascular responses to Valsalva are divided into four phases. Phases I and II occur during breath holding,phases III and IV after release. The BP and HR responses are mirror images: when BP increases, HR reflexly decreases. Measuring HR alone is adequate for some aspects of the Valsalva response, but a complete evaluation requires measurement of BP. In phase I there is a brief rise in BP because of increased intrathoracic pressure constricting the great vessels; in phase II, there is a gradual fall in BP because of impaired venous return that reaches a plateau because of peripheral vasoconstriction, with a compensatory tachycardia; in phase III, there is a brief fall in BP because of removal of the intrathoracic pressure constricting the great vessels. Phase IV occurs after the Valsalva is released and the patient resumes normal breathing, the BP begins to recover and slowly rises. About 15 to 20 seconds after release, there is a rebound overshoot of BP to a level above baseline, accompanied by a reflex bradycardia with an HR below baseline, lasting for approximately 1 minute. The Valsalva ratio is the ratio of the fastest HR during phase II to the slowest HR during phase IV, or the longest R-R interval during phase IV to the shortest (R-R) interval during phase II. Normal is approximately ≥1.45, but age specific reference values are more precise. A lack of rebound overshoot of BP during phase IV is an early

indicator of autonomic dysfunction. A lack of overshoot can also occur in some non-neurologic conditions, such as congestive heart failure. The BP changes occur quickly and it is not possible to follow the complete cycle at the bedside with a BP cuff. The rebound overshoot in phase IV, however, can be detected by inflating a cuff to just at SBP and then having the patient Valsalva. Without changing the cuff pressure, the sounds will disappear during breath holding, and on release the sounds will return and can befollowed up to detect the rebound overshoot in BP.Tilt-table testing evaluates the integrity of autonomic reflexes. Autonomic laboratories use different degrees of tilt, but usually in the range of 60 to 80 degrees and for different durations. In neurocardiogenic (vasovagal, vasodepressor) syncope, or fainting, hypotension is accompanied by bradycardia, rather than the tachycardia that should occur. It occurs in response to emotional upsets such as fear, stress, or the sight of blood, occasionally in relation to micturition (micturition syncope) or coughing (cough syncope), and sometimes without identifiable provocation. Tilttable testing has shown that a neurocardiogenic mechanism is responsible for a large proportion of the patients with recurrent, unexplained syncope.

Examination of the Sensory Functions

The somatic sensory elements of CN X are discussed above. They are not clinically important and cannot be adequately tested.

Examination of the Reflexes

Cranial nerve X plays a part in several autonomic, or visceral, reflexes; loss of these reflexes may follow a lesion of the tenth nerve. In some of these reflexes, such as the sternutatory, sucking and yawning, the vagus plays a supportive role.. Afferent impulses are carried over CN V to the reflex center in the brainstem and upper spinal cord, with efferent impulses primarily by CN VII with some overflow to CNs IX and X and the phrenic nerve. In other reflexes, such as swallowing, vomiting, and coughing, the vagus is central.

The Spinal Accessory Nerve

CLINICAL EXAMINATION

The functions of the cranial portion of CN XI cannot be distinguished from those of CN X, and examination islimited to evaluation of the functions of the spinal portion. The examination of spinal accessory nerve is limited to an evaluation of the functions of the sternocleidomastoid(SMC)and trapezius muscles.

One SCM acts to turn the head to the opposite side or to tilt it to the same side. Acting together, the SCMs thrust the head forward and flex the neck. The muscles should be inspected and palpated to determine theirtone and volume. The contours are distinct even at rest. With a nuclear or infranuclear lesion there may be atrophy or fasciculations.

To assess SCM power, have the patient turn the head fully to one side and hold it there, then try to turn the head back to midline, avoiding any tilting or leaning motion. The muscle usually stands out well, and its contraction can be seen and felt (Figure). Significant weakness of rotation can be detected if the patient tries to counteract firm resistance. Unilateral SCM paresis causes little change in the resting position of the head. Even with complete paralysis, other cervical muscles can perform some degree of rotation and flexion;

only occasionally is there a noticeable head turn. The two sternocleidomastoid muscles can be examined simultaneously by having the patient flex his neck while the examiner exerts pressure on the forehead, or byhaving the patient turn the head from side to side. Flexion of the head against resistance may cause deviation of the head toward the paralyzed side. With unilateral paralysis, the involved muscle is flat and does not contract or become tense when attempting to turn the head contralaterally or to flex the neck against resistance. Weakness of both SCMs causes difficulty in anteroflexion of the neck, and the head may assume an extended position. The sternocleidomastoid reflex may be elicited by tapping the muscle at its clavicular origin. Usually there is a prompt contraction. The reflex is mediated by the accessory and upper cervical nerves, but has little significance in neurologic diagnosis.

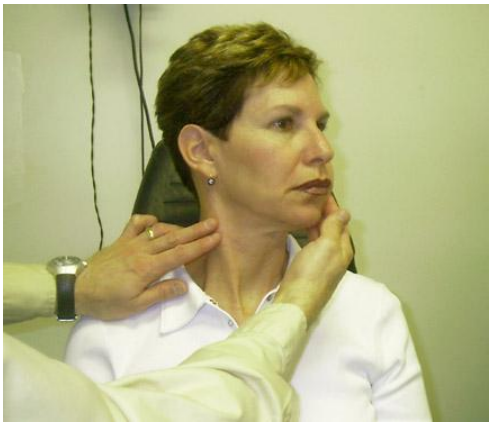
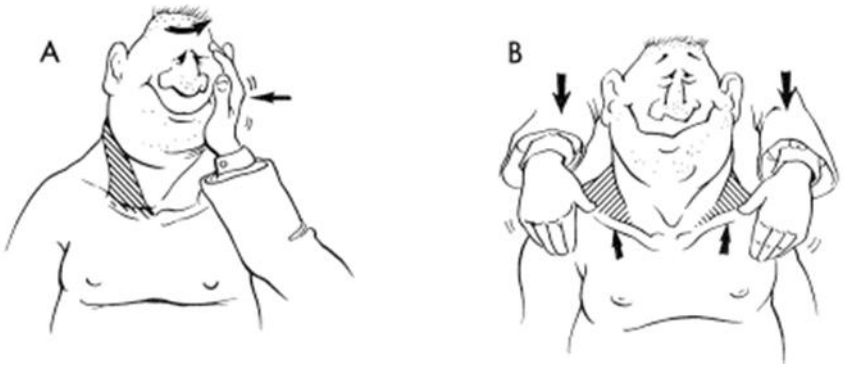


FIGURE*:* Examination of the sternocleidomastoid muscle. When the patient turns his head to the right against resistance, the contracting muscle can be seen and palpated.

With trapezius atrophy the outline of the neck changes, with depression or drooping of the shoulder contourand flattening of the trapezius ridge (Figure). Severe trapezius weakness causes sagging of the shoulder, and the resting position of the scapula shifts downward. The upper portion of the scapula tends to fall laterally, while the inferior angle moves inward. This scapular rotation and displacement are more obvious with arm abduction. The strength of the trapezius is traditionally tested by having the patient shrug the shoulders against resistance (Figure). However, much of shoulder shrugging is due to the action of the levator scapulae.

A better test of the upper trapezius is resisting the patient's attempt to approximate the occiput to the acromion. The movement may be observed and the contraction seen and palpated. To examine the middle and lower trapezius, place the patient's abducted arm horizontally, palm up, and attempt to push the elbow forward. Muscle power should be compared on the two sides. In unilateral weakness of the trapezius, these movements are impaired.



FIGUR A, Testing of right sternomastoid muscle. B,Testing upper portion of trapezius muscles.

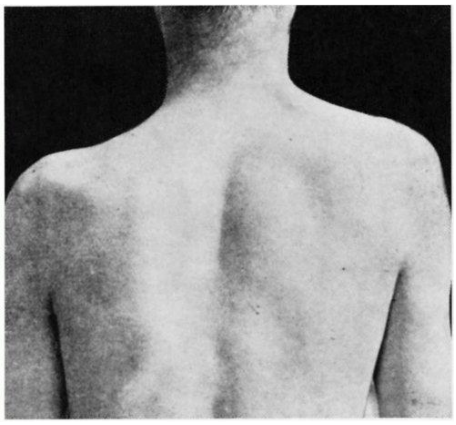


Figure*:* Paralysis of the left trapezius muscle. There is a depression in the shoulder contour with downward and lateral displacement of the scapula.

The trapezius is one of several muscles that act to stabilize the scapula and create a platform for movements of the humerus. The serratus anterior protracts the scapula, moving it forward as in a boxing jab. The trapezius is a synergist to the main mover, the rhomboids, in retracting the scapula. The trapezius and serratus anterior act in concert to rotate the scapula when the arm is abducting. The trapezius brings the glenoid fossa progressively more cephalad so that the abduction motion is unrestricted. In addition,contraction of the upper trapezius adds the final few degrees of abduction, after the glenohumeral and acromioclavicular ranges of motion are exhausted, so that the arm can be brought directly overhead (Figure).

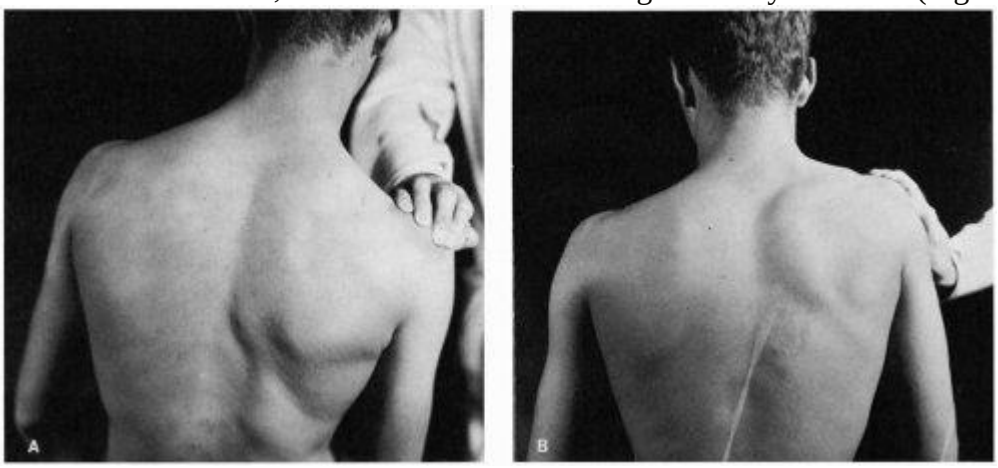
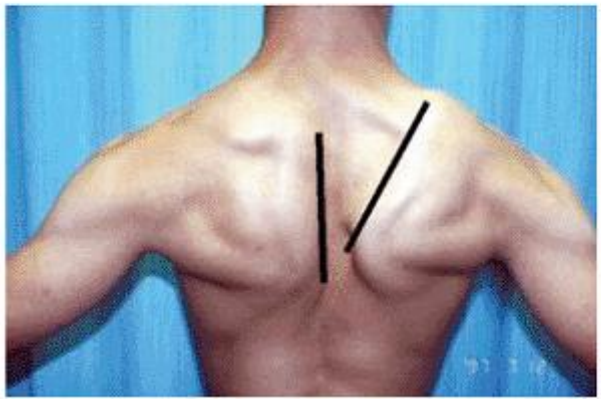


Figure: Examination of the trapezius muscle. **A.** Examiner pressing shoulder down against patient's resistance. **B.** Patient attempting to elevate shoulder against examiner's resistance.

Weakness of the trapezius disrupts the normal scapulohumeral rhythm and impairs arm abduction. Impairment of upper trapezius function causes weakness of abduction beyond 90 degrees. Weakness of the middle trapezius muscle causes winging of the scapula. The

winging due to trapezius weakness is more apparent on lateral abduction in contrast to the winging seen with serratus anterior weakness, which is greatest with the arm held in front. In fact, with winging due to trapezius weakness, the jutting of the inferior angle lessens when the arm is raised anteriorly; in winging due to serratus anterior weakness, it worsens. When the trapezius is weak, the arm hangs lower on the affected side, and the fingertips touch the thigh at lower level than on the normal side. Placing the palms together with the arms extended anteriorly and slightly below horizontal shows the fingers on the affected side extending beyond those of the normal side. The drooping of the arm and shoulder caused by trapezius weakness may lead to pain and subjective sensory complaints in the extremity due to traction on musculoligamentous structures and possibly sensory nerves. Loss of shoulder mobility may result in a secondary adhesive capsulitis, which further restricts motion.

The two trapezius muscles can be examined simultaneously by having the patient extend his neck against resistance. Bilateral paralysis causes weakness of neck extension. The patient cannot raise his chin, and the head may tend to fall forward (dropped head syndrome). The shoulders look square or have a drooping, sagging appearance due to atrophy of both muscles.



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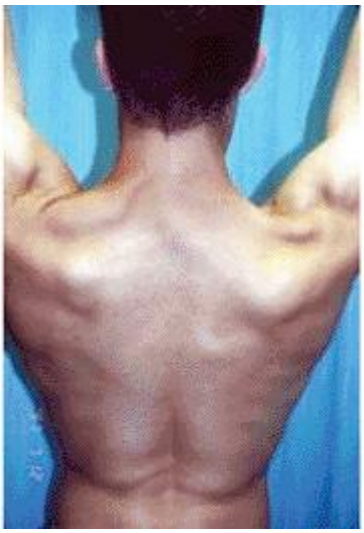

B

EXAMINATION OF MUSCLE TONE

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Introduction

The word 'tonus' was first used by Muller (1838) to designate the state of contraction of muscles.

Muscle tone is defined as the resting state of contraction of a muscle or muscle group. Muscle tone affects muscle contractibility even if it is not involved in active voluntary contraction. Muscle tone is attributable to the intrinsic properties of muscle such as elasticity.

Physiology

Muscle tone or the state of resting contraction of muscle is maintained by the involuntary gamma loop. The muscle spindles which lie within striated muscles are receptors for stretch. These muscle spindles are innervated by gamma afferents which synapse with gamma efferent nerve cells located in the anterior horn of spinal cord. The gamma efferents innervate the muscle and keep it contracted. This segmental loop is essentially an involuntary phenomenon. Various supraspinal tracts affect this local segmental loop either positively or negatively thereby modifying the tone accordingly.

The muscle tone does not exist without reason. This tone helps maintain balance and posture in the passive state. It also prevents dislocations and subluxations of joints and keeps muscles prepared for active contraction if needed.

Disorders of tone

Hypertonia:

Hypertonia means that the resting state of contraction in a particular muscle or muscle group is more than normal. Such a condition usually arises from disinhibition of the spinal reflex arc due to supraspinal lesions. Conditions giving rise to hypertonia include pyramidal tract lesions in the brain and spinal cord. Muscle tone is also increased in extrapyramidal tract

diseases like Parkinson's disease & Huntington's chorea, and in other diseases like CJD, decortication syndromes, stiff man syndrome and cerebellar atrophy.

Hypotonia:

Hypotonia means loss of normal tone in muscles. When extreme, the limb becomes flaccid. Hypotonia occurs due to disruption of the reflex arc (gamma loop). Examples include brachial plexus and other peripheral nerve palsies, anterior horn cell diseases, Poliomyelitis, Tabes dorsalis, cerebellar lesions, etc.

Conditions mimicking hypertonia

There are certain conditions that mimic hypertonia such as paratonia or "gegenhalten", Myotonia (Thomson's disease), Neuromyotonia, torticollis, muscle spasm due to inflammation, blepharospasm, some tics, functional (hysterical) rigidity, etc.

Clinical Examination of Muscle tone

Assessment of tone is difficult. There is a lot of inter-observer variability. This is one examination that requires extreme patient cooperation, gentleness on the part of the examiner and a good rapport between doctor and patient. Talking to the patient, explaining what is going to be done and assurance that he/she is not going to be hurt are very essential to maximize the yield from clinical examination of tone. Examination of tone should be done across all joints starting distally in fingers & toes and progressing proximally to the shoulder joint. Examination of tone of extremities is easier than evaluation of truncal muscle tone. Examination of muscle tone in infants is difficult.

Pieron (1920) and Foix (1924) examined muscle tone in three aspects, i.e.

a) Resting muscle tone
b) Attitudinal muscle tone and
c) Action Tone

A. Resting Muscle tone

Resting muscle tone, as the name suggests, is examined when the muscle is at rest. This again has three aspects, i.e.

a) Consistency (deep palpation): This is tested by deep palpation of the muscle between two fingers. This can be compared with the normal side.
b) Extensibility (Stretching): When joint movement is normal, this is examined by flexing or extending joints.

   Some of the tests usually performed are:

   i. Babinski Tonus test: With arms at the level of shoulders the forearms are passively flexed at the elbow joint and the tone is compared on both sides. In hypotonia the involved elbow joint is more acutely flexed than the normal side.
   ii. Shoulder-shaking Test: The examiner places his hands on the patent's shoulders and shakes them vigorously to and fro, thereby assessing tone.

c) Resistance (to passive movements): This is performed with passive stretching of the muscle by moving the joint in alternate (to & fro) directions while keeping the proximal segment of the joint fixed.

B. <u>Attitudinal (postural) muscle tone</u>

When a muscle group is moved passively and immobilized in a new position which is not the *resting position* of the limb; the tone adapts to this new position in normal conditions. However, in certain conditions with extrapyramidal rigidity such as Parkinson's disease, this reflex is exaggerated. Some of the classical postures and gait like festinating gait, hemiparetic gait and spastic gait suggest hypertonia, while others, like the flail limb of brachial plexus injury, depict hypotonia.

Following are some examples of tests for attitudinal muscle tone:

i. Arm-dropping test: The patient's arms are briskly lifted to shoulder level and suddenly dropped. In hypertonia there is delayed descent of the arm (Bekhterew's sign).
ii. Hand position: Hypo-tonicity due to Sydenham's chorea assumes a characteristic limb position with the arms and fore-arms outstretched, flexion at the wrists and hyper-extension of the fingers.

C. <u>Tone of action</u>

Tone of action is tested when the patient is asked to do some known activity, like buttoning-unbuttoning, combing, etc. Normally, these movements are regular, harmonious and purposeful. In hypertonia, the movements are jerky and clumsy with over- or under-shooting of movements. Similarly, in hypotonia, the movements are slow and incomplete.

On assisted movements of joints in cases with hypertonia two situations exist:

I. In pyramidal weakness (spasticity): on passive firm movement of the concerned joint there is sudden release of resistance after a peak, known as 'Clasp-knife' phenomenon.

II. In extra-pyramidal type of weakness (rigidity): on passive firm movement of the concerned joint there is continuous ('Lead-pipe') or multiple short resistance ('Cog-wheel') rigidity. This is because of the sustained tonic state in extra-pyramidal lesions.

Further Reading

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2) DeJong's The Neurologic Examination edited by W.W. Campbell, seventh edition (2014).

GUIDE TO EXAMINATION OF THE MOTOR SYSTEM- MUSCLE POWER

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Examination of muscle power involves INSPECTION, PALPATION, MOVEMENT and MEASUREMENT in that order. This is a brief discussion of the details.

INSPECTION

STAGE 1

- Examination of the motor system starts during the history taking itself. Assess the patient's articulation, content of speech, and overall mental status
- Inspect the patient's facial features, facial movement and note any asymmetry. Note the patient's eye movements, blinking, and the relation of the palpebral fissures to the iris, look for enophthalmos or exophthalmos.
- Observe how the patient swallows saliva and breathes. Inspect the posture and look for tremors or involuntary movements

STAGE 2

Inspection of the limbs and position of the trunk is the first step in assessment of power. Look for any twitches, tremors, abnormal movements or postures.

- Look carefully for hypokinesia, decreased eye blinking or staring which could be indicative or an extrapyramidal disorder such as Parkinson's disease.
- In suspected lower motor neuron disorders, look for muscle wasting, hypertrophy or fasiculations .
- In case there is wasting, the pattern and extent of wasting is of utmost importance: whether the wasting is proximal or peripheral, unilateral or bilateral holds a key to the anatomical localization of the lesion.

- Remember that the position of the limbs is a clue to power in a patient who is bedbound and supine. An externally rotated lower limb for example is indicative often of a marked weakness in that limb as a whole (when local hip causes are ruled out!) One needs to observe the shape, size and position of limbs.

PALPATION- LOOKING FOR TONE

This has been discussed by the previous author.

EXAMINATION FOR POWER- MOVEMENT

While assessing power it is important to look at the limbs as a whole (in cases of upper motor deficits) or at individual muscle groups (in lower motor deficits). The order of examination must necessarily be from limb to muscle.

- Hence, Test for subtle weakness first by checking pronator drift, finger tapping, pronation/supination movements and toe tapping.
- Then check individual muscles for strength using the MRC scale to rate strength
- The MRC(Medical Research Council, UK) established a grading system for muscles as follows:

1 Flicker or trace contraction

2 Active movement with gravity eliminated

3 Active movement against gravity

4 Active movement against gravity and resistance

5 Normal power

0 No contraction

The following facts have to be understood about each muscle:

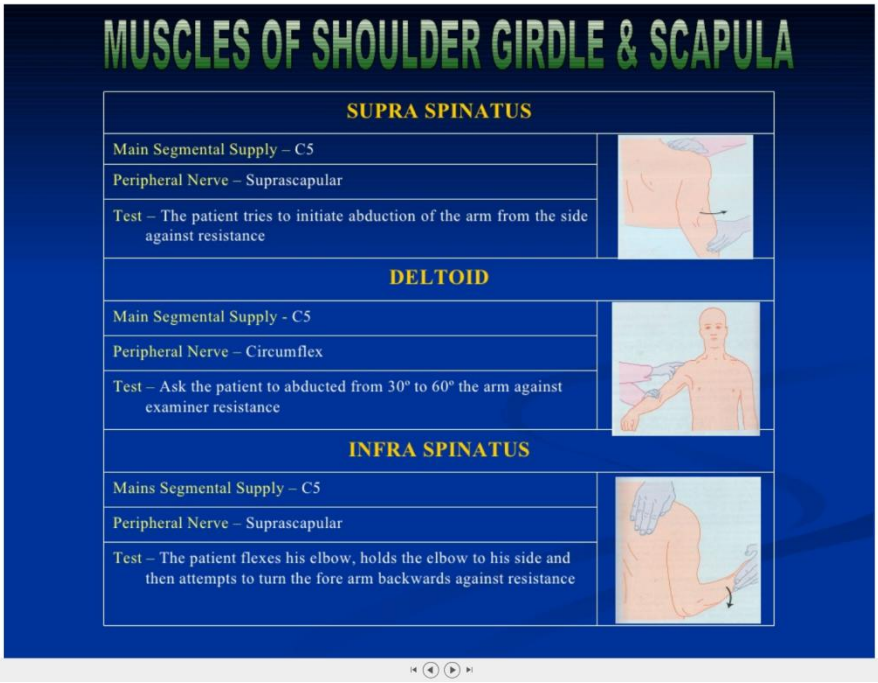
1) The peripheral nerve supply.

2) The segmental nerve supply.

3) The action of the muscle.

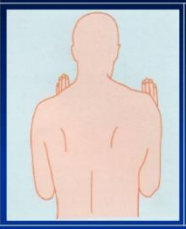
It needs to be reiterated that during assessment of power, the muscle needs to be fully exposed, palpated, and compared to its similar muscle/group ON THE OTHER SIDE. In neurological convention, the healthy side is always examined first.

The charts below are a guide to individual muscles/groups that need to be examined.



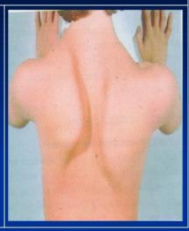
SERRATUS ANTERIOR

Main Seg supply C5C6C7 P. Nerve – N. to serratus anterior



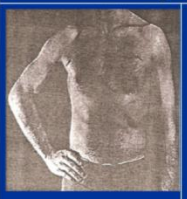
Test
Push the wall by his hands keeping elbow extended

If seratous anterior is weak, scapular medial borden will become prominent – winging of scapula



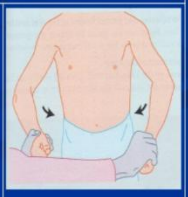
PECTORALIS MAJOR

Main Seg supply C5C6C7 P. Nerve – Lat. & medial pectoral N.



Test-1
Placing the hand on hip and pressing in wards

Test-2
Strech the arm out infront & than to clasp the hands together while examiner endeavour to hold them apart



LATISSIMUS DORSI

Main Seg supply C7 P. Nerve –Nerve to L. Dorsi

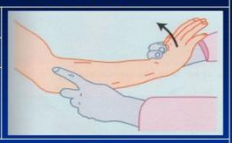
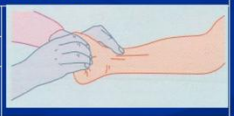
Test-1
While palpating the muscle, ask the patient to cough

Test-2
Resist the patient's attempt to adduct the arm when abducted to above 90°

RHOMBOIDS

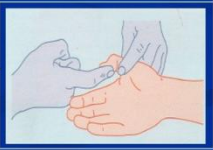
- SEGMENTAL SUPPLY C5
- PERIPHERAL NERVE NV TO RHOMBOIDS
- METHOD ASK THE PATIENT TO PLACE HIS HANDS ON HIP AND TRY TO FORCE HIS ELBOW BACKWARDS.

MUSCLES OF FOREARM & WRIST JOINT

EXTENSOR CARPI RADIALIS LONGUS	
Main Segmental Supply –C6C7	
Peripheral N. – Radial	
Test – Pt. holds the fingers partialy extended and dorsiflexes the wrist towards the radial side against resistance	
EXTENSOR DIGITORUM	
Main Segmental Supply-C7	
Peripheral Nerve – Radial	
Test – Examiner attempt to flex the patients extended fingers at metacarpophalangeal joints	
FLEXOR CARPI RADIALS	
Mains Segmental Supply – C6C7	
Peripheral Nerve – Median	
Test – Examiner resist the patient attempt to flex the wrist towards the radial side	
FLEXOR CARPI ULNARIS	
Mains Segmental Supply – C8	Peripheral Nerve - Ulnar

MUSCLES OF THUMB

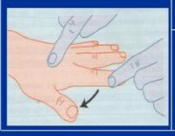
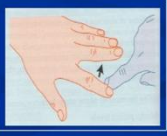
ABDUCTOR POLLICIS LONGUS	
Main Segmental Supply –C8	
Peripheral N. – Radial	
Test – Pt. attempts to maintain his thumb in abduction against the examiner resistance	
EXTENSOR POLLICIS BREVIS	
Main Segmental Supply - C8	
Peripheral Nerve – Radial	
Test – Pt. attempts to extend the thumb while the examiner attempts to flex it at MCP joint	
EXTENSOR POLLICIS LONGUS	
Mains Segmental Supply – C8	
Peripheral Nerve – Radial	
Test – Patients attempt to extend the thumb while the examiner attempts to flex it at the interphalangeal joint	

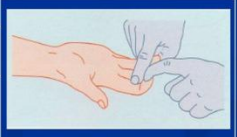
OPPONENS POLLICIS	
Main Segmental Supply –T1	
Peripheral N. – Median	
Test – Pt. attempts to touch the little finger with the thumb	
ABDUCTOR POLLICIS BREVIS	
Main Segmental Supply - T1	
Peripheral Nerve – Median	
Test – Ask the patient to abduct the thumb in a plane at right angle to the palmar aspect of index finger against the resistance of examiner thumb	
FLEXOR POLLICIS LONGUS	
Mains Segmental Supply – C8	
Peripheral Nerve – Median	
Test – Patient attempt to flex the distal phalanx of the thumb against the patient resistance (Hold the proximal phalanx)	

Adductor pollicis

- T1
- ULNAR NERVE
- The patient attempts to hold a piece of paper between the thumb and the palmar aspect of forefinger.

MUSCLES OF HAND & FINGERS

LUMBRICAL & INTEROSSI		
	Main Seg supply C8 T1 Median – Lumbical I & II Ulnar – lumbrical III & IV & interossi	
	A. Lumbricals – Pt. Tries to flex the extended fingers at the MCP Joints	B. Interossi – Pt. attemps to keep the fingers abducted against resistance
IST DORSAL INTEROSSI & IST PALMAR INTROSSI		
	Main Seg supply T1 Peripheral nerve ulnar	
	Pt. Tries to abduct the fore finger against resistance	Pt. Tries to Adduct the fore finger against resistance

FLEXOR DIGITORUM SUBLIMIS	
Main Segmental Supply –C8	
Peripheral N. – Median	
Test – Pt. flexes the finger at the proximal inter phalangel joint against resistance (examiners fingers placed on middle phalanx)	
FLEXOR DIGITORUM PROFUNDUS	
Main Segmental Supply - C8	
P. N. – Median Ist & IInd Ulnar IIIrd & IVth	
Test – Pt. Flexes the terminal phalanx of the fingers against resistance, the middle phalanx being supported	
ABDUCTOR DIGITI MINIMI	
Mains Segmental Supply – T1	
P. N. – Ulnar	
Test – Back of hand is placed on the table & little finger abducted against resistance (sign of ulnar N. lesion)	

Abdominal muscles

- T5- L1
- Intercostal, ilioinguinal and iliohypogastric nerves
- The patient lies on his back and attempts to raise his head against light resistence. Watch movement of the umbilicus.
- ***Beevor's sign*** -with paralysis of the lower segment the umbilicus moves upwards but when upper segment is affected, umbilicus is pulled downwards.

- Ask the patient to sit up in bed from the supine position without the aid of arms.
- Inability to do so indicates severe weakness of the muscles of abdomen and the hip flexors.
- ***Babinski's rising up sign*** –in spastic paralysis of the leg the affected limb will rise first, but in hysterical paralysis this does not occur.

- Ask the patient to sit up in bed from the supine position without the aid of arms.
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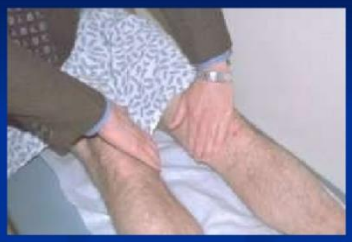
Hip extension

- instruct the patient to press down on the examiner's hand which is placed underneath the patient's thigh. This tests the gluteus maximus
- Inferior gluteal nerve [l4,L5]
- Observe the pt standing from a low chair

Hip adduction

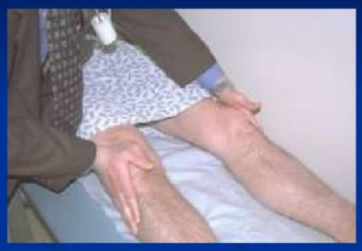
- place your hands on the inner thighs of the patient and asking them to bring both legs together. This tests the adductors of the medial thigh.
  the L2, L3 and L4 nerve roots.



Hip abduction

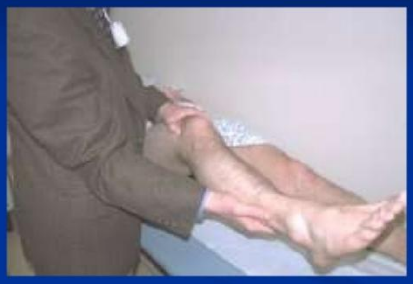
- place your hands on the outer thighs and asking the patient to move their legs apart. This tests the gluteus maximus and gluteus minimus.

  Abduction of the hip is mediated by the L4, L5 and S1 nerve roots.



Knee flexion

hold the knee from the side and applying resistance under the ankle and instructing the patient to pull the lower leg towards their buttock as hard as possible. This tests the hamstrings. L5 and S1 nerve roots via the sciatic nerve.



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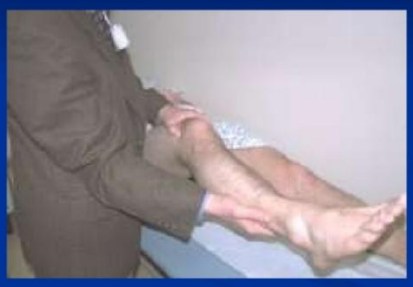
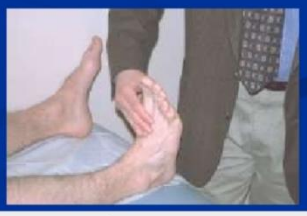


Ankle dorsiflexion

Hold the top of the ankle and have the patient pull their foot up towards their face as hard as possible.This tests the muscles in the anterior compartment of the lower leg.

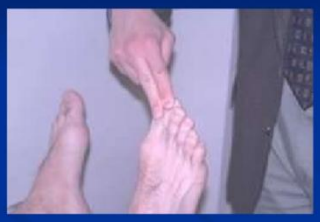
L4 and L5 nerve roots via the peroneal nerve

Ask the patient to walk on heels



Extension of great toe

Ask the patient to move the large toe against the examiner's resistance "up towards the patient's face". The EHL muscle is almost completely innervated by the L5 nerve root.



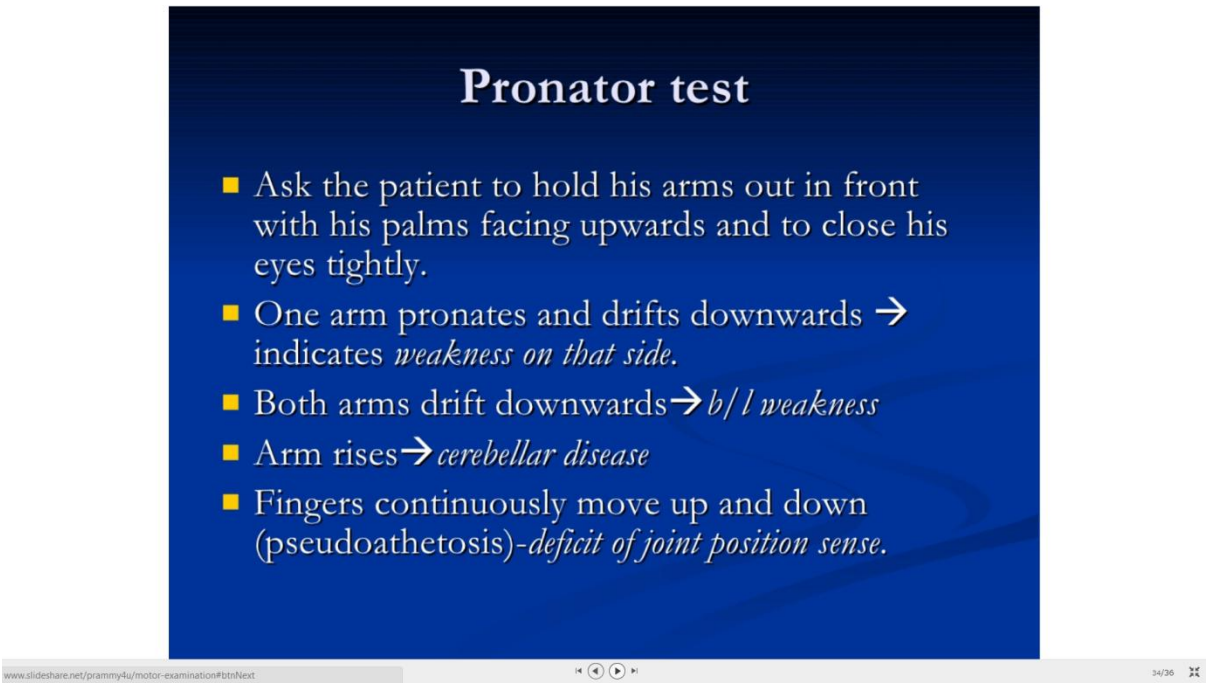
Summary- upper limb

ROOT	MOVEMENTS	REFLEX
C5	Shoulder abduction, elbow flexion	Biceps
C6	Elbow flexion (semipronated)	supinator
C7	Finger extension, elbow extension	Triceps
C8	Finger flexors	
T1	Small ms of hand.	

Lower limb

Root	Movement	Reflex
L1, L2	Hip flexion	
L3, L4	Knee extension	Knee reflex
L5	Dorsiflexion of foot, inv & eversion of ankle, extn of great toe.	
S1	Hip extension, knee flexion, plantar flexion.	Ankle reflex

One final test needs to be mentioned: the so called test for PRONATOR DRIFT which records small "cortical" weakness of the upper limb.



MEASUREMENT

Muscle bulk is measured by recording diameter of the limb at a fixed distance from a joint, e.g. decreased girth of the thigh 8 cm above the knee joint on one side could signify wasting of quadriceps on that side. Measurements are thus an additional reinforcement of inspectory findings of wasting.

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EXAMINATION OF ABNORMAL MOTOR MOVEMENTS

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Abnormalities of movements also called movement disorders, may involve any part of the body and are due to disease involving various parts of the motor system due to varying etiologies.

CLASSIFICATION: Hypokinetic

Hyperkinetic

DEFINITIONS:

Parkinsonism: A clinical syndrome with bradykinesia, accompanied by rigidity and tremor

Dyskinesia: Involuntary movements usually drug-induced (may be chorea or dystonia)

Tremor: A rhythmical, involuntary oscillatory movement of a body part; may be resting or postural

Chorea: A quick, irregular, semi-purposive and predominantly distal involuntary movement

Dystonia: An abnormal movement characterised by sustained muscle contractions, frequently causing twisting and repetitive movements or abnormal postures

Ballism: A proximal, high amplitude movement, often violent and flinging in nature; usually unilateral in nature.

Tic: An abrupt, jerky non-rhythmic movement (motor tic) or sound (vocal tic) that is temporarily suppressible by will power; tics may be simple or complex.

HISTORY:

Time course / functional disability / effect upon quality of life

Past medical history, including infections & toxin exposure

Musculoskeletal symptoms (e.g. frozen shoulder with early PD)

Drug history

Alcohol consumption & responsiveness

Family history

Neuropsychiatric features

Autonomic symptoms

Sleep problems (REM sleep behavioural disorder suggests PD, Dementia with Lewy Body or Multi-System Atrophy)

EXAMINATION:

Observation of involuntary movements:

1. **Specific distribution**: e.g. restless legs syndrome (RLS) and painful legs and moving toes (PLMT). Parkinson's disease (PD) is typically asymmetric in onset.
2. **Specific actions:** e.g. action tremor and dystonia. The patient may be asked to write or pick up a cup of water
3. **Speed:** Slow - parkinsonism, dystonia
   Intermediate - chorea, tremor
   Fast - myoclonus,
4. **Rhythm:** Continuous – tremor, PLMT
   Intermittent - asterixis

5. Relation to posture: e.g- Orthostatic tremor

6. Relation to sleep: few movement disorders persist during sleep; e.g. palatal tremor and segmental myoclonus

7. Relation to voluntary movement: e.g. action tremor and action dystonia.

8. Associated sensory symptoms: PLMT, RLS and phantom dyskinesias; tics may be associated with a vague discomfort or unusual sensation in the prodrome before the movement

9. Suppressibility: Volitional in tics (but associated with increasing unease and rebound worsening of tics upon release), by sensory tricks in dystonia and by activity in rest tremor

10. Aggravating or precipitating factors: stress and anxiety worsen all movement disorders; myoclonus may be worsened by specific stimuli e.g. sudden, loud noise

11. Ameliorating factors: alcohol may relieve essential tremor and myoclonic-dystonia, sometimes quite dramatically; walking backwards or running may improve a dystonic gait, leading the unwary to suspect a non-organic cause.

Cognitive assessment (subcorticofrontal vs cortical problems – MMSE, verbal fluency test, Luria, go/no-go task)

Cardiovascular – lying & standing blood pressure, cool periphery (MSA)

Gait, postural reflexes (pull test) & **axial tone**

Eye movements especially saccadic speed & latency & blink frequency

Limb examination (include specimen of writing & observe posture)

- tremors / dystonic posturing (including postural & action)
- tone – use reinforcement if in doubt
- power & co-ordination

- fine finger and rapid alternating movements

Reflexes including plantars / primitive reflexes

GAIT AND STANCE

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Gait abnormality is a common clinical problem, which may have neurological and non-neurological origins. Careful clinical examination is required to know the cause of gait abnormality. Station is the way the patient stands and gait is the way the patient walks. Both require number of factors and reflexes, which includes proprioceptive sensations, basal ganglia, cerebellum and its connections, intact skeletal muscles, tonic neck and labyrinthine reflexes.

Examination of station:

Station is patient's attitude, posture and way he stands. Asking the patient to stand with feet closely together tests station. Note any instability and unsteadiness. Further examination is carried out by asking the patient to close eyes, stand on one foot at a time, stand on toes and then on heels. He or she may be given a gentle push to see whether patient falls to one side, forward or backward.

Unsteady patients always compensate by placing feet wide apart. In vermian cerebellar lesions, patient will always tend to sway forward or backward. In unilateral cerebellar lesions, patient will tend to fall on the side of the lesion. In these patients, if a gentle push is given on either side, they tend to lose balance more on affected side. In unilateral vestibular lesions, the patient will also tend to fall on affected side. If a patient with unilateral cerebellar lesion is asked to stand on one foot, the patient will loose more balance on ipsilateral involved side.

Apart from this, skeletal changes like kyphosis, scoliosis, lordosis, any sciatic list, position of head, shoulder, hips and extremity is noted. If a patient is unable to stand by himself, notice how much support the patient needs for the same.

Romberg Sign:

When proprioception is impaired, patient may be able to stand with eyes open, but sways or falls when asked to close eyes. This is Romberg sign. To demonstrate this sign, the patient must have a stable stance with eyes open and then demonstrate the loss of balance with eyes closed. Many patients sway slightly with eyes closed especially elderly patients. This is of no significance.

Romberg sign is a test of proprioception and not of for cerebellar function. Even in cerebellar disease, particularly with disorders of spinocerebellum and vestibulocerebellum, there may be some degree of instability with eyes closed but not to the degree with impaired proprioception.

Examination of Gait:

Gait requires coordinated action of sensory and motor functions. In some patients, gait may be the only abnormality on clinical examination that may point to clinical diagnosis.

Various parameters are used to measure and characterize gait which include gait velocity, stride time, step time, stride length and step length. An adult walking normally on a level surface, walks with velocity of 80 m/sec, 113 steps / min, stride length of 1.41 meter. Around 60% of gait cycle is spent on stance and 40% on swing. Body's center of gravity is located just anterior to S2 vertebra.

Ask the patient to walk and make sure to see the arms and legs clearly. Now you have to ascertain whether the gait is symmetrical or asymmetrical.

If the gait is symmetrical:

Check the paces whether small or normal

If paces are small*:*

Look the patient's posture and arm swing

1. Stooped with reduced arm swing: Parkinson disease (Gait difficult to start and stop-festinant gait- may be worse on one side)
2. Upright with marked arm swing-march a petit pas

If normal paces:

Look at the lateral distance between the feet

1. Normal
2. Widely separated- broad based
3. Legs uncoordinated- cerebellar
4. Crossing over, toes dragging- scissoring

Look at the knees

1. Normal
2. Knees lifted high- high stepping gait

Look at the pelvis and shoulders

1. Normal
2. Marked rotation of pelvis and shoulders on either sides while walking- waddling gait

Look at the whole movement

1. Normal
2. Disjointed as if forgotten to walk, patient appears to be fixed at the spot- apraxic gait
3. Bizzare, elaborate and inconsistent - functional gait

If gait is asymmetrical

Is patient in pain?

1. Yes- painful or antalgic gait

Look at the bony deformity

1. Orthopedic gait

Does one leg swing out to the side?

1. Yes- hemiplegic gait

Look at the knee heights

1. Normal
2. One knee lifts higher- foot drop

Further examination of gait

Ask the patient to walk as if on tight rope

1. If patient falls consistently- unsteady
2. Patient may fall on either side
3. Elderly patient may be slightly unsteady

Ask the patient to walk on heels

1. If unable to do- foot drop

Ask the patient to walk on toes

1. Unable to do so- gastrocnemius weakness

Interpretation*:*

1. **Parkinsonian Gait**: due to basal ganglia dysfunction

2. **March a petit pas**: indicates bilateral diffuse cortical dysfunction. Common causes- diffuse cerebrovascular disease

3. **Scissoring gait**: indicates spastic paraparesis, common causes- cerebral palsy, multiple sclerosis, spinal cord compression

4. **Sensory ataxia**: indicates loss of joint position sense (Romberg's positive sign) common cause: peripheral neuropathy, Posterior column involvement.

5. **Cerebellar ataxia**: patient veers towards the side of lesion,common cause- phenytoin toxicity, alcohol intoxication, tumors, cerebrovascular disease.

6. **Waddling gait:** indicates the weakness of proximal hip girdle muscles, causes- mayopathies, hip dislocation.

7. **Apraxic gait**: due to loss of cortical integration of movements, usually frontal lobe pathology. Common causes- normal pressure hydrocephalus, cerebrovascular diseases.

8. **Hemiplegic gait:** Unilateral upper motor neuron lesions. Common cause- stroke.

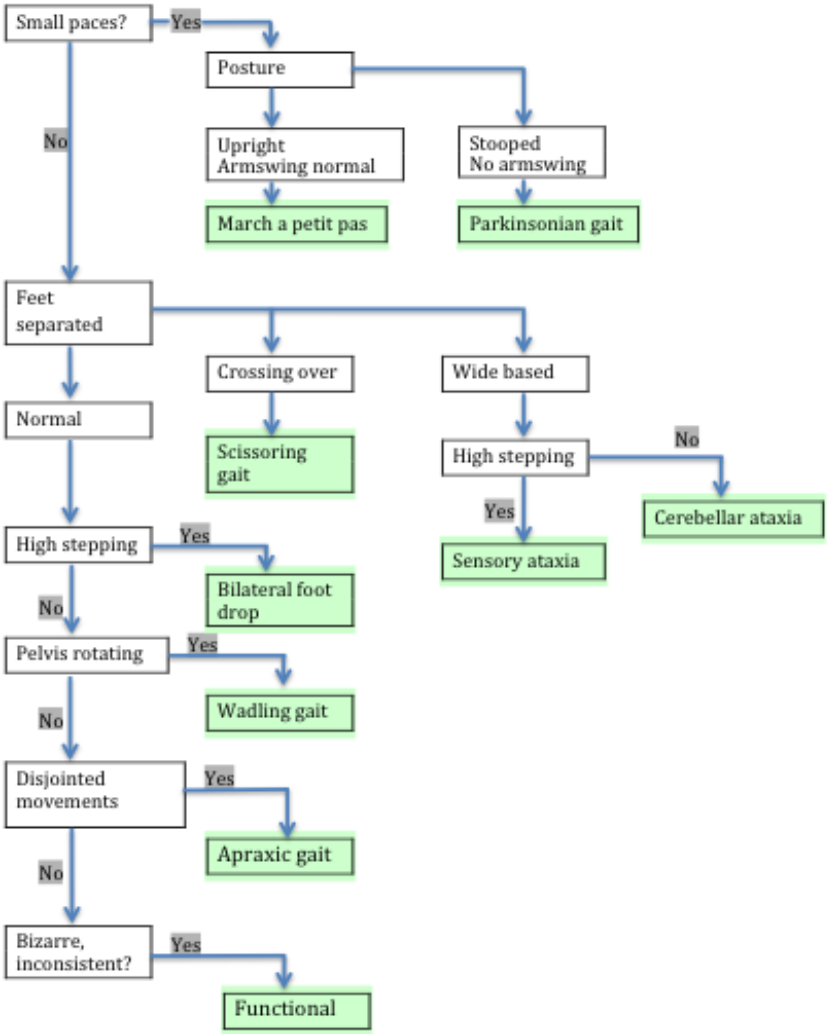
9. **Foot drop**: common causes- Unilateral- common peroneal nerve palsy, pyramidal lesion, and L5 radicular lesion. Bilateral: peripheral neuropathy.

10. **Functional gait:** usually inconsistent with rest of the neurological examination. It is worse when watched.

Non-neurological gaits:

1. Painful gaits: common causes- arthritis, trauma
2. Orthopedic gait: due to shortened limbs, previous hip surgery, obvious injuries to the patient.

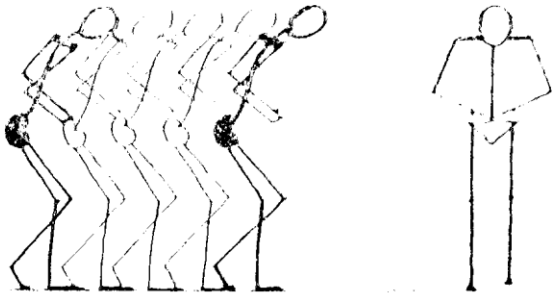
Flow Chart for gait abnormality



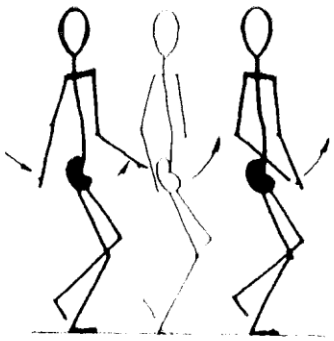
Symmetrical Gaits

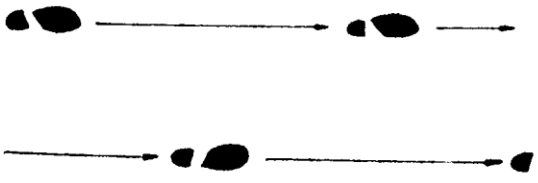
Normal

Parkinsonian gait

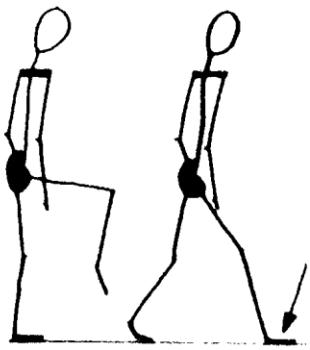


Marche à petit pas





Wide based



High stepping

Asymmetrical Gait

Hemiplegic gait

Asymmetrical Gait

Hemiplegic gait

Clinical Examination and Localisation of Brachial Plexus Injuries

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Anatomy of the Brachial Plexus (BP) (Figure 1, 2)

The BP is formed mainly by the ventral rami of 5 spinal nerves: C5, C6, C7, C8 and T1.If there is a major contribution from C4 and minor contribution from T1, it is called prefixed brachial plexus and if there is a major contribution from T2 and a minor contribution from C5, then it is a post-fixed plexus. The plexus is divided into roots, trunks, divisions, cords and major terminal branches (proximal to distal).

Roots and Trunks

The upper two roots (C5 and C6) join to form the upper trunk while the lower two roots (C8 and T1) join to form the lower trunk. C7 root continues as the middle trunk.

Of all the roots and trunks, clinically significant branches arise from C5, 6 and 7 roots and from the upper trunk. The dorsal scapular nerve arises from the C5 root and supplies the rhomboid muscles. The phrenic nerve (C3-5) receives contribution from C5 root and supplies the diaphragm. The long thoracic nerve (of Bell) is formed by the roots of C5, 6 and 7 and supplies the serratus anterior muscle. The only branch of significance, arising from the trunk, is the suprascapular nerve (C5, 6). This nerve arises from the upper trunk and supplies the supra and infraspinatus muscles.

Divisions

The divisions bridge the gap between the trunk and the cord. Each of the three trunks divides into an anterior and posterior division. All the three posterior divisions join to form the posterior cord. The anterior divisions of the upper and middle trunks join to form the lateral cord while the anterior division of the lower trunk continues as the medial cord. There are no named branches from these divisions.

Cords and Major terminal branches

The cords are named according to their relationship to the axillary artery lying deep to the pectoralis minor muscle (lateral, medial and posterior). As the cords pass distal to the pectoralis minor muscle, it's anatomical relationship changes and it is no longer lateral, medial or posterior.

Branches from lateral cord:

The musculocutaneous nerve (C5,6) is the distal continuation of the lateral cord which contains fibers of the upper trunk. It supplies the coracobrachialis, biceps brachii and brachioradialis muscles. It receives sensory information from the skin on the lateral aspect of the forearm through the anterior and posterior divisions of the lateral cutaneous nerve of the forearm.

The lateral cord gives a branch which merges with a branch from the medial cord to form the median nerve (C6-T1). The lateral cord component (C5 to C7) mainly carries the sensory input from the median nerve from the lateral palm and first three digits. There is also a smaller motor component which supplies the pronator teres and flexor carpi radialis muscles.

The lateral pectoral nerve (C5, 6) arises from the proximal part of the lateral cord and innervates the pectoralis major muscle (predominantly the clavicular part)

Branches from the Medial cord:

The Ulnar nerve (C8-T1) is the major terminal branch of the medial cord.

The medial pectoral nerve arises from its proximal part. It innervates the pectoralis minor and the sternal head of pectoralis major muscles.

The third and fourth branches include the medial cutaneous nerve of arm and forearm respectively. They carry sensory information from the medial half of arm and forearm. Loss of sensations in this area indicates medial cord involvement.

The medial component of the median nerve (C8-T1) is derived from the medial cord. It is a pure motor nerve with no sensory component. It supplies the intrinsic muscles of the hand and variably the long finger flexors.

Branches from the Posterior cord:

The radial nerve is the direct continuation of the posterior cord (C5-T1).
The axillary nerve (C5, 6) arises from the posterior cord, deep to the axillary artery. It has an anterior division which supplies the deltoid and a posterior division which supplies the teres minor. It also receives sensory fibers from the upper lateral arm through the upper lateral cutaneous nerve of arm.
The upper and lower subscapular nerves (C5, 6) supply the subscapularis muscle.
The thoracodorsal nerve, arising between the upper and lower subscapular nerves, supplies latissimus dorsi muscle (C6, 7).

Clinical Evaluation

With a thorough understanding of the anatomy and systematic examination of the upper limb, brachial plexus injury localization can be done with relative ease. The examination can be started off by dividing BP palsies into proximal i.e roots (C5-T1) and upper trunk palsies (note there are no significant branches from middle and lower trunk) and distal i.e cord and nerve palsies.

Proximal Brachial plexus palsies

C5 root

It contributes mainly to shoulder abduction and external rotation. This is brought about by the axillary nerve supplying the deltoid (30-90 degrees abduction of shoulder) and the suprascapular nerve which supplies the supra (0-30 degree abduction of shoulder) and infraspinatus (important external rotator muscle) muscles. Hence, an overall function of

C5 mediated actions can be assessed by the arm raise, in which, the arms are kept by the side of the body and abducted to 90 degrees while simultaneously externally rotating the arm so that the palm faces the roof **(Figure 3)**.
The C5 root receives sensory supply from the lateral portion of the arm upto the elbow (via the upper and lower lateral cutaneous nerve of arm which join the axillary and radial nerves respectively). So, sensory loss can be detected in this area.

C6 root

This root plays a major role in arm extension and adduction along with forearm supination and flexion. Arm extension and adduction is mediated by C6 through the latissimus dorsi while flexion and supination of the forearm is mediated through the radial nerve (supinator and brachioradialis muscles) and the musculocutaneous nerve (biceps brachii muscle). An underhand chin-up movement (when a person tries to lift himself up while holding a bar and tries to bring his chin to the bar) causes extension and adduction of the arm with flexion and supination of the forearm thus assessing the function of C6 **(Figure 4)**. The thumb and the lateral aspect of the forearm are the sensory territories of C6 (via the lateral cutaneous nerve of forearm and the sensory branches of the median and radial nerves) where sensory impairment can be looked for.

Upper trunk

Palsies of the upper trunk results in weakness of C5 and C6 innervated muscles with sensory loss along the lateral aspect of the arm, forearm including the whole thumb.
The affected limb is adducted and internally rotated (because of unopposed action of pectoralis major), forearm is extended and pronated (pull of triceps and pronator teres) and the wrist and fingers are flexed (C6 is responsible for wrist and finger extension). This position is known as *policeman's tip deformity or waiter's tip position.*

Weakness of the rhomboids (dorsal scapular nerve), serratus anterior (long thoracic nerve) and /or diaphragm (phrenic nerve) helps localize the injury to the C5 and C6 roots rather than upper trunk.

Middle Trunk

As the C7 root continues as the middle trunk, no distinction can be made between C7 root injury and an injury involving the middle trunk. Its major contribution is to the elbow extension via triceps (radial nerve), wrist flexion via flexor carpi radialis (median nerve) and ulnaris (ulnar nerve) and pronation of forearm via pronator teres (medial nerve). C7 also provides variable innervation to the wrist extensors as well as finger flexors and extensors. The triceps pushdown test assesses the C7 root/middle trunk function. In this test, one tries to get up from a chair by pressing hands on a table placed in front. In doing so, the wrist has to be flexed, forearm pronated and the elbow to be extended **(Figure 5)**. A person with C7 or middle trunk injury cannot perform this action. In these lesions, there is sensory loss over the entire long fingers supplied by the median and radial nerves.

C8 root

An easy way to assess the function of C8 root is to ask the patient to grasp and then let go the examiners fingers. One has to pay special attention to the first three digits. This movement is impaired in C8 lesions. The C8 provides major innervation to the long finger flexors/extensors and along with T1, supplies the intrinsic hand muscles. Commonly, C8 palsies cause significant weakness of the long flexor (profundi) to the index and long fingers, thenar muscles and extensors to the thumb, index and long fingers. Thus, the hand grasp test can pick up a C8 lesion. Sensory loss along the lateral third of hand including the entire little finger (superficial branches of lunar nerve) occurs in C 8 lesions.

T1 root

The dorsal interossei muscles are supplied predominantly by T1 root via the ulnar nerve. Power is tested by asking the patient to spread the fingers. Atrophy of the first dorsal interosseous is easily observed if present. Sensory innervation from the medial half of the forearm is carried through T1 by the medial cutaneous nerve of forearm, a branch of the medial cord. T1 root avulsions may be associated with Horner's syndrome. Damage to both C8 and T1 roots causes Klumpke's palsy which causes grip weakness.

Lower Trunk Palsies will have features of C8 and T1 motor and sensory deficits.

Distal Brachial plexus palsies

This includes cord lesions. Clinical identification of isolated lesions involving the BP divisions is not possible. It resembles either trunk or cord lesions.

Lateral cord

The lateral cord consists of fibers from C5, 6 and 7. The main terminal branches include musculocutaneous nerve and lateral branch of median nerve (C5-7). Hence, there will be weakness of forearm flexion along with loss of sensations along the lateral aspect of the forearm (musculocutaneous nerve and lateral cutaneous nerve of forearm). Due to the lateral component of median nerve involvement, there will be sensory loss on the lateral aspect of the palm and the first three digits along with weakness of pronation of forearm and flexion at the wrist. Weakness of clavicular head of pectoralis major may also be present due to involvement of lateral pectoral nerve.

Medial Cord

It contains fibers of C8 and T1 as a continuation of the lower trunk. Lesions of medial cord will cause ulnar nerve palsy and median nerve weakness of C8-T1 distribution.

There will be weakness of medial wrist flexion, flexor digitorum profundus of the fourth and fifth fingers, all movements of little finger, finger abduction and adduction (ulnar motor) along with sensory loss over the medial palm (ulnar sensory). Involvement of the medial component of the median nerve (C8-T1) causes weakness of the intrinsic thumb muscles and first two lumbricals (proximal interphalangeal joint extension weakness). Involvement of the other side branches of the medial cord leads to sensory loss over the medial aspect of the arm and forearm (medial cutaneous nerve of arm and forearm) and weakness of the sternal component of the pectoralis major (medial pectoral nerve)

Posterior cord

It contains fibers from C5 to C8 with variable contribution from T1. The signature injury of the posterior cord is radial-axillary nerve palsy. Radial nerve palsy will cause forearm extension and supination weakness, wrist, finger and thumb extension weakness with sensory loss over the posterior aspect of arm and forearm(posterior cutaneous nerve of arm and forearm), lower lateral aspect of arm (lower lateral cutaneous nerve of arm) and dorsolateral hand (superficial sensory radial nerve). Axillary nerve palsy will cause shoulder abduction weakness with sensory loss over the upper lateral aspect of arm. Due to involvement of the subscapular and thoracodorsal nerves, there will also be weakness of shoulder adduction and internal rotation. This will help confirm injury to the posterior cord.

Diagnosis of Preganglionic Injury

It is important to determine whether a lesion is pre or post ganglionic. This is important from treatment and prognosis point of view. Root avulsions of the BP may cause wasting and weakness of the rhomboids and levator scapulae (C5), serratus anterior (long thoracic nerve) and hemidiaphragm palsy (C3-5). There may also be paraspinal muscle wasting. T1 lesion can cause Horner's Syndrome. However, ENMG (presence of sensory nerve action potentials) and MRI (pseudomeningoceles) help in clinching the diagnosis.

Summary

A thorough understanding of the anatomy of the BP is of paramount importance for proper evaluation and planning management of brachial plexus injury. A systematic approach of examination must be adhered to for the same. Initially, examine the back to look for the paraspinal muscles, trapezius, rhomboids, serratus anterior and latissimus dorsi and then proceed examination from the proximal shoulder joint then elbow, wrist and fingers respectively. Muscle power across all joints must be tested. Finally a sensory examination must be done. Knowing the deficits and applying the knowledge of BP anatomy should help in accurate localization of the lesion.

Suggested Reading

1.Russell SM. Examination of Peripheral Nerve Injuries: An Anatomical Approach.Thieme Publication 2006.

2.Kilne and Hudson's Nerve Injuries: Operative results for Major Nerve Injuries,Entrapments and Tumors.Saunders 2nd edition, 2008

Examination of sensory System

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Overview

Classification of sensation

Anatomists divide sensations into:-

1. **Somatic Sensations –**
    a. General Sensations (pain, touch, temperature, vibration, joint and position etc),

    b. Special Sensations (Smell, Vision, Taste, Hearing & Vestibular sensation.)
2. **Visceral Sensations –**
    a. General Sensations,
    b. Special Sensations

Sherrington divided Sensations into (Based on location of end organs):-

1. **Exteroceptive –**
    a. Originate in skin or mucous membrane
    b. Stimulus is – external agent or Environment
    c. 3 types – pain / temperature / light touch
2. **Interoceptive**
3. **Proprioceptive**
4. **Combined**

Another way of classification:

1. Cortical sensory system
2. Non-cortical sensory system

Sensory pathway

1. In general the sensory pathway consists of: - Stimulation of receptors – passage of impulse through sensory nerves via dorsal root ganglia – enters specific fiber tracts in spinal cord / brain stem - central sensory areas of brain for conscious recognition.

2. The impulses pass through dorsal root ganglia which are situated just outside the cerebro-spinal axis (spinal cord & brain stem). From dorsal root ganglia the fibers are grouped in medial (towards posterior column) and lateral divisions (towards spinothalamic tract).

Abnormalities of sensation and related Symptoms

1. **Increased sensation** – e.g. pain – due to excessive stimulation or damage or irritation of fibers, tracts or receptors
2. **Perversion of sensation –**
    a) **Paraesthesia** - feeling abnormal sensation without any stimulus e.g.
        i. Sensation of Itching,
        ii. Sensation of "Pins & needles",
        iii. Sensation of crawling of insects (formication),
        iv. Sensation of Burning or hotness, coldness, thickness of skin,
        v. Sensation of weight, constriction, Pressure, Distention,
        vi. Feeling as if blood supply has been stopped(Tingling)

    b) **Dysesthesia** - perverted sensation of feeling burning on touch.
    c) Phantom sensation
3. **Decreased sensation** – Decreased sensation of pain, touch, temperature sensation etc.
4. **Delayed sensation** – as in tabes dorsalis.
5. **Dissociation of sensation** – loss of one type but not of others.
6. **Mixture of above.**
7. **Asymptomatic** - temperature, vibration, joint and position loss is often asymptomatic.

Evaluation of sensory symptoms

1. Must determine what the patient means by "numbness."
2. Onset: when did the symptom begin? Duration?
3. What Type of sensation is affected?
4. How much sensation is affected (Quantity)?
5. Exact distributions of affection- are there sharp borders?
6. Change is subjective (Dysesthesia) or spontaneous (Paraesthesia)?
7. Periodicity?
8. Has it been steady, progressive or does it wax and wane?
9. Accentuating, Precipitating, Relieving factors?
10. Are there additional accompanying symptoms that are present?
11. Other diseases: does the patient have symptoms of other underlying diseases?

Interesting facts

1. Sensory examination is a simple procedure **if** patient is co-operative and alert.
2. At times the findings may be confusing & may require a **repeat evaluation after some time.**
3. Pain may coexist with numbness.
4. Vibration and joint and position test are usually quick and easy –**always test them first.**
5. **Always move from affected area to normal area**.
6. Sensory signs are considered "softer" as compared to reflex and motor signs, and less weight is given to them while localizing the lesion.

Requisites for a satisfactory sensory examination

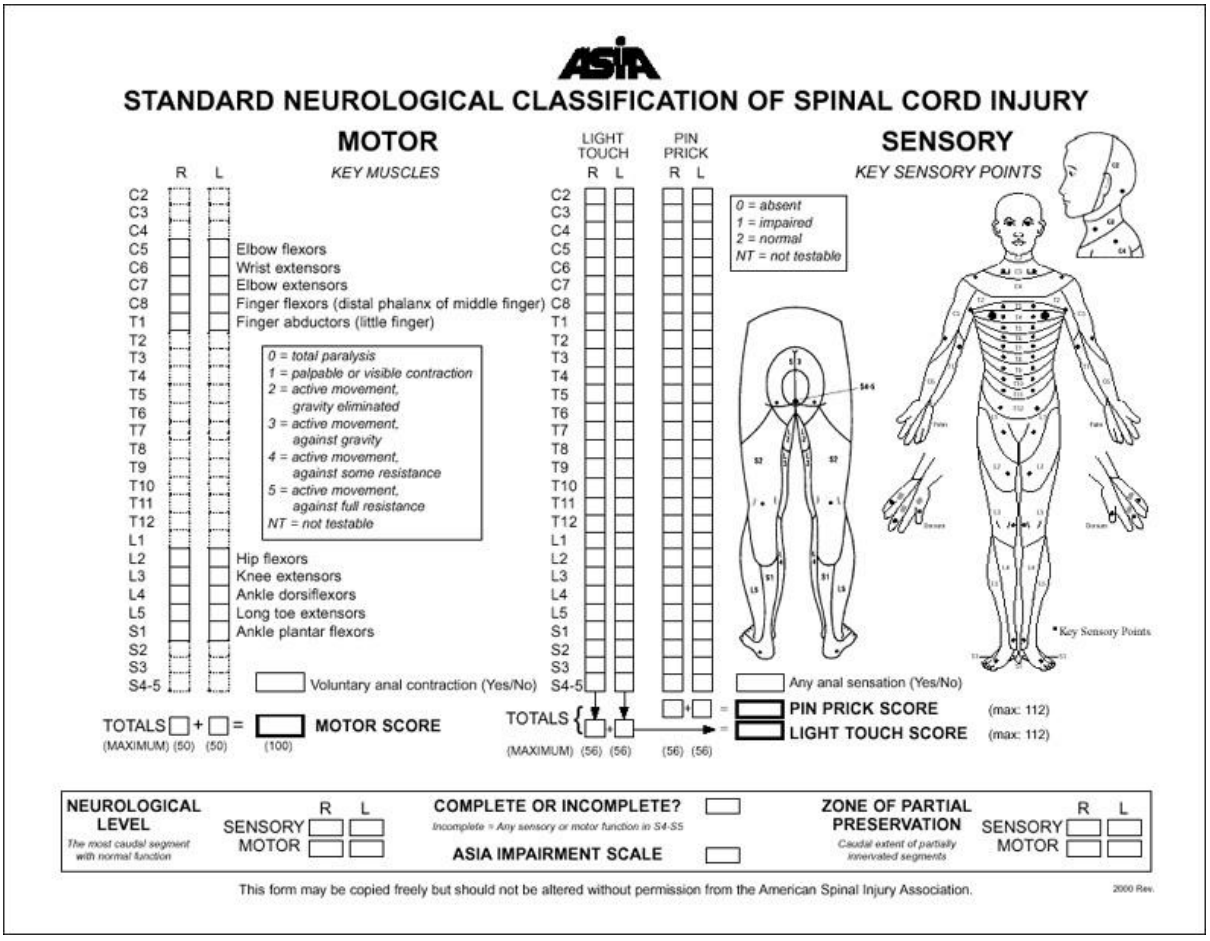
1. Patient must understand the procedure – explain the procedure to the Patient.
2. Patient should be comfortable & at ease (avoid noisy surroundings, ask whether the patient is thirsty or hungry, temperature should not be too cold, maintain privacy, exposure should be as little as required).
3. Compare symmetric areas of body.

Preliminary and Detailed examination

1. **Preliminary examination** - If patient does not have sensory symptoms – one can test entire body rapidly. (Preliminary examination).
2. **Detailed examination** - If sensory symptoms are presnt and if abnormality is found on Preliminary examination, then detailed examination must be performed.

Recording of findings of examination

1. Put your findings either in **sensory pictorial charts** or in **text form**.
2. Mention whether the loss is in **dermatomal pattern** or **peripheral nerve distribution pattern**
3. Mention the **quantity** of loss
4. Mention **reliability** of your findings



Superficial pain sensation

Anatomy (Superficial pain sensation)

1. Fibers for Superficial pain sensations travel along **unmyelinated & thinly myelinated** nerve fibers.
2. From Dorsal root ganglion they pass through lateral root to dorsolateral fasciculus of spinal cord
3. Within one or two segments of their point of entry into the cord they synapse on the stellate cells or funicular cells or both.
4. 2^{nd} order neuron from these cells cross the midline of cord **anterior** to the central canal and ascend in **lateral spinothalamic tract**. (Lower limb fibers are lateral & dorsal to upper limb fibers).
5. Location of **Lateral ST Tract** at different levels –
    a. **in medulla** - It is Dorsolateral to inferior olivery body Lateral to medial leminiscus & medial to middle cerebellar peduncle in pons
    b. **in mid brain** - Dorsal to medial leminiscus & dorsolateral to red nucleus
6. 2^{nd} order neuron of Lateral ST Tract terminate in **nuclei Ventralis posterolateralis & posteromedialis** of thalamus along with pain fibers from 5^{th} nerve in ventral secondary ascending tract of 5^{th} nerve.
7. Lower limb fibers are lateral & rostral and upper limb fibers are medial & caudal.

Clinical examination (Superficial pain sensation)

1. Use pin- **do not use hypodermic needle**
2. Pin should be sterile & for single use only
3. Keep patient's eyes closed
4. Explain everything to him

5. Alternate stimulation with head & the point of pin has to be done & patient with closed eyes has to say **'sharp' or 'dull'**.
6. Proceed from areas of lesser sensitivity to those of greater sensitivity.
7. Test in dermatomal pattern as well as PN pattern.
8. Distance (variable) & time gap (2-3 seconds) between two stimulus should be enough to **not to allow the summation of stimulation**.
9. **Do not forget to examine sacral sensations.**

Temperature sensation

Anatomy (Temperature sensation)

1. Two types of temperature sensations– warm & cold
2. Pathway of fibers for temperature sensations is **same as pain Fibers**
3. In lateral spinothalamic tract Fibers of temperature are little dorsal & medial to pain, but there is much overlapping & dissociation is very rare.

Clinical examination (Temperature sensation)

1. Use: -
    a. Test tubes with cold & warm water
    b. Metal tubes with cold & warm temperature
    c. Tuning fork in air conditioned room can be used for testing (screening) cold sensation
    d. Beam of light for (screening) warm sensation
2. Patient should respond by **'warm' or 'cold'**
3. Temperature should be 5 – 10 degree C for cold testing 40 – 45 degree C for warmth testing - beyond this range pain fibers get stimulated.
4. Loss of one (warm or cold sensation) in usually associated with loss of other.
5. Usually area for loss of 'warm' sensation in more than area for loss of 'cold' sensation.
6. Sometimes patient feels warm & cold both stimulus as warmth.

Tactile sensation

Anatomy (Tactile sensation)

Two types –

a. Light touch (crude touch)

i. Impulses travel in **myelinated fibers**

ii. Axons enter through **medial division** of dorsal root – ascend & descend up to many segment & 2nd orders fibers cross to opposite side in **ventral S.T. tract.**

b. Fine touch –

i. Impulses travel in **myelinated** fibers

ii. **Medial division** of dorsal root ganglia

iii. Ascend in fasciculus gracilis & cuneatus

iv. End in nucleus gracilis & cuneatus is medulla

v. 2nd order neurons cross opposite as **arcuate** fibers & ascend is **medial leminiscus**

vi. Face sensation through dorsal secondary ascending tract of 5th nerve are in close proximity.

vii. In thalamus all tactile fibers are **caudal to pain & temperature fiber**

viii. In cortex they are **posterior to pain & temperature fibers.**

ix. **Do not forget to examine sacral sensations.**

Clinical examination (Light Touch)

1. Use
    a. Camel's hair brush
    b. Wisp of cotton
    c. Feather
    d. Tissue paper
    e. Light touch with fingers tip
    f. Sharp & blunt ends of a pin for simultaneous examination of pain & light touch.

2. Stimulus should be so light that no pressure is produced on subcutaneous tissue. **Do not move/drag the stimulus on skin, only dab/touch lightly it on skin.**
3. Patient should say – 'yes' for feeling the stimuli and should tell area where stimulated (keep changing from upper limb, lower limb, trunk etc)
4. **Do not forget to examine sacral sensations**.

Clinical examination (Fine touch)

1. Use hairless skin area for fine touch
2. Touch but do not move the stimulating object
3. Two point discrimination testing is good enough for fine touch testing.

Proprioceptive sensation (Sense of motion & position, vibration, Pressure and deep pain)

Anatomy (Proprioceptive sensation)

1. Arise from deeper tissue – muscles, ligament, bones, tendons & joints
2. Sensation of joint & position, vibration & pressure & deep pain are included.
3. Impulses travel along **heavily myelinated fiber**
4. **Medial division** of dorsal root – ipsilateral fasciculus gracilis & cuneatus -> gracilis & cuneatus nuclei in medulla -> cross as internal **arcuate fiber** -> medial laminiscus -> thalamus.
5. Fasciculus gracilis & cuneatus - Lower limb fibers are medial, upper limb fibers are lateral.
6. Medial laminiscus – on both sides of median raphe in medulla - Lower limb fibers – ventral, upper limb fibers dorsal
7. In pons lower limb fibers lateral, upper limb medial
8. In thalamus – lower limb fibers lateral upper limb fibers medial.
9. via post limb of internal capsule to cortex

Clinical examination (Sense of motion & position)

1. Explain and demonstrate the procedure to patient with eyes open
2. Ask him to close eyes

3. Passively move the digits up and down
4. Ask patient –
    a. Whether he can appreciate movement?
    b. Whether he can recognize direction of movement?
    c. Whether he can recognize 1 – 2 degree movement at interphalyngeal joint.
5. Digits should be **grasped laterally with minimal pressure & moved gradually.**
6. Separate digits from other digits
7. Patient **should not do any active movement** during testing.
8. Start in distal joints and proceed to proximal

Other ways of examination (Sense of motion & position)

1. Put patients finger in a particular position with his eyes closed - ask him to
    a. Describe the position of finger
    b. Imitate the position of finger with other hand
    c. Put his leg in certain position with eyes closed & ask him to show his great toe
2. Ask him to stand with outstretched hand with eyes closed
    a. See if one hand droops
    b. Change position of one hand & ask him to change position of other hand to same level.
3. Finger nose test with eyes open & eyes closed
4. **Romberg's test** is a test of Sense of motion & position

Clinical examination (Sense of vibration)

1. T F of **128 H_z** is used
2. Explain and demonstrate the procedure to patient
3. Make sure that he responds to vibration and not to touch of T F.
4. Ask him to close eyes
5. Start at toe tips – mp joint- medial malleolus – tebial tuberosity –ASIS – spine – arms – wrist – elbow – shoulders – compre right with left.

6. **If sensations are normal distally in a limb, no need to test proximally in the same limb.**
7. Note whether patient can feel or not when T F is vibrating maximally
8. Then when he stops feeling it, check it on clinician's bone to compare
9. IF patient can not feel check at a proximal point or upper limb
10. It is carried mostly in post column & partly through ST tract – therefore rarely one can find dissociation of joints & position sense & vibration sense.
11. **DM, pernicious anemia, hypothyroidism – there may be marked loss of vibration sense.**

Clinical examination (Pressure sensation)

1. Tested by pressure on subcut structure by blunt object carried via post column.
2. Patient is asked-
    i. To feel the stimulus
    ii. To tell the location of stimulus

Clinical examination (Deep pain sensation)

1. **These are the only Proprioceptive sensation that travel in Lateral spinothalamic tract along with superficial pain**
2. Tested by squeezing muscle, testicles, eyeballs
3. Early loss in tabes dorsalis
4. Preceded by delayed pain response

Interoceptive or visceral sensation

1. Vaguely localized
2. Parietal pleura sensitive to pain
3. Vessels of heart sensitive to pain
4. Travel through dorsal root ganglia to spinal cord & then
    a. I/L ST tract
    b. C/L ST tract medial to pain & temperature fiber
    c. Some reach to thalamus without decussating

5. End station may be gyrus rectus rather than sensory cortex.
6. Cordotomy for intractable visceral pain has to be
    a. Deeper as the fibers lie medial to other fibers in ST tract
    b. Higher as the fibers ascend greater distance before dicussation.
    c. bilateral as the fibers may travel in both crossed and uncrossed tracts
7. Visceral pain may be referred to cutaneous area of their dermatome (viscerocutaneous reflex).
8. Visceral pain may be referred to muscles leading to their spasm due to visceromotor reflex

Cortical sensations

1. These are combined sensations
2. They use Proprioceptive, Exteroceptive inputs along with cerebral component from parietal lobe to analyze the sensation

Stereognosis

1. To identify an object's form, size, shape and nature by touch.
2. **Loss of stereognosis** is known as **astereognosis or tactile agnosia.**
3. Can be tested in hand only.
4. For size testing – use smaller and bigger rounded cutouts of stiff paper
5. For shape testing – use cicular, triangular or square cutouts of stiff paper
6. For form testing – use cubes, pyramids or plasic balls.
7. For nature testing – use wood, glass and metal.
8. Touch sensation must be normal for stereognosis.
9. If motor power is weak then examiner may have to manipulate the object in patient's hand.
10. Occasionally seen in cervical lesions too.

Barognosis

1. Ability to recognize and differentiate weight.
2. **Loss of Barognosis** is known as **baragnosis.**
3. Can be tested in hand only.

4. Use two coins of same shape and size but different weight.

Topognosia

a. Ability to locate a tactile sensation.
b. Loss of Topognosia is known as **Topagnosia.**
c. Can be tested in hands as well as legs.

Graphesthesia

1. Ability to recognize numbers and letters written on hands.
2. Loss of Graphesthesia is known as **Graphanesthesia.**
3. Letters of 1 mm on finger tips, 4 mm on forearm and legs can be appreciated.
4. Written letters should be dissimilar and easily identifiable 3& 4 rather 3& 8.
5. Letters are written using pencils.
6. Unilateral loss is particularly important.

Two point discrimination

1. Ability to recognize cutaneous stimulation by one blunt object from two blunt objects
2. Tested by using compass. Patient is stimulated randomly by a single point and by two points. Gradually reduce the distance between two points.
3. Normal discrimination distance is 2- 4 mm at finger tip, 4-6 mm at dorsum of finger, 8-12 mm on palm, 20-30 mm on dorsum of hand.
4. Compare with other side of body.

Sensory inattention

1. Touch patient on one side and then on other side. If he is able to recognize each side independently, then touch him on both sides at the same time.
2. If he is able to recognize left and right sides when stimulated independently but recognized only one side when both sides stimulated at the same time this is called sensory inattention
3. It indicates parietal lobe lesion.

Autotopagnosia/somatotopagnosia

1. Loss of ability to identify the body schema/parts.
2. Occasionally seen in cervical lesions too.

Finger Agnosia: Inability to name individual fingers of his own or of examiner.

Anosognosia: inability to perceive the existence of disease such as hemiplegic limbs.

Localization of site of lesion

Peripheral nerve –

1. Loss will follow PN distribution & not dermatomal pattern
2. Loss will be equal for all modalities – no dissociation
3. Area of actual loss in usually in smaller than anatomical distribution of that nerve
4. Borders are always graded & not very sharp
5. **Some important Peripheral nerve distribution to remember**
    a) Hand – medial 1.5 finger – **Ulnar Nerve**
    b) Hand – lateral 3.5 fingers – **Median Nerve**
    c) Hand –anatomical snuff box – **Radial Nerve**
    d) Arm – lateral aspect where deltoid inserts – **Axillary Nerve**
    e) Thigh – anterolateral aspect upper 2/3 – **Lateral Cutaneous Nerve Of Thigh** (meralgia parasthetica)
    f) Leg – anterolateral leg and dorsum of foot sparing lateral 2 toes – **Common Peroneal Nerve**
    g) Anteromedial thigh and anteromedial leg – **Femoral Nerve**
    h) Sole and dorsum and most of posterior leg and thigh – **Sciatic Nerve**

Dorsal root ganglion affection occurs in

a) Tabes dorsalis
b) Hereditary sensory neuropathy

Rhizotomy –

a) For relief of intractable pain.

b) Lesion is between Dorsal root ganglion and cord.

c) If lesion is distal to Dorsal root ganglion regeneration with return of pain may occur.

Nerve root

1. Loss will be in segmented distribution
2. Loss will be for all modalities
3. May be associated with radicular pain
4. **Some important dermatome to remember**

a) C1 - no sensory supply

b) Intermeatal Line over vertex – 5th nerve meet C2

c) C4 -5 meet T1 – 2 on upper chest

d) L1-2 meet S2 – 3 on medial thigh near genetalia

e) C5 –6 - lateral arm, forearm & hand

f) C7 – Middle Finger

g) C8 T1 - medial arm, forearm & hand

h) T4 - nipple level

i) T10 - umbilicus level

j) T12 & L1 – groin

k) L1 –2 – 3 – anterior thigh

l) L4 –5 – anterior & lateral leg

m) L5 – great toe

n) S1– little toe,

o) S1-2 – **little toe**, most of sole, posterior leg& thigh

p) S3 –5 – perineum

q) Sole – L5-S1

r) Heel – **S1**

Spinal cord, brain stem

1. Sharp level – with pain & temperature loss
2. No level – with touch loss
3. Level may be 1 – 2 level below or above the actual level.

Parietal lobe –

a) Threshold for stimulation of sensation on opposite half of body is increased.
b) **Agnosias** – loss of cortical sensations
c) Sensory affection is more in upper than lower limbs, distal than proximal parts of the limb
d) Perception become normal before midline is reached due to opposite normal parietal lobe.

Thalamus

a) All forms of sensation on opposite side are blunted.
b) **Hyperpathia** –marked discomfort by light stimulation of affected side of body
c) **Anesthesia dolorosa** – diminution of sensation accompanied by intractable pain in affected side of body
d) Hemiplegia,hemianopia, hemiataxia, choreoathetosis, unmotivated emotional response may also occur.

Interesting facts

1. **Peripheral neuritis –**
    a. Loss of sensation but increased pain
    b. Nerves that can be felt under skin – ulnar & Radial, Peroneal nerve may feel tender
    c. Passive stretching of limb may cause pain in nerve
    d. Vibration may be lost in early course of disease.

2. **Cordotomy –** Lesioning of lateral spinal thalamic tract between dentate ligament & motor root to relieve intractable pain
3. **Posterior cordotomy** - For relief of pain in phantom limb

4. **Causalgia** is a neuritis characterized by parastesias and trophic changes usually in median and sciatic nerve distribution.
5. **Phantom sensation** is spontaneous sensations in anesthetic areas.
6. **Phantom limbs** are continued sensations of pain, movement or parastesias in an absent part of body.
7. **Cutaneous sensory extinction**
8. **Hysteria – Psychogenic pain –**
    a. Vague,
    b. Associated with inappropriate affect,
    c. Inconsistent - Varies with each examination
    d. Sensory loss up to exactly midline or quite beyond it
    e. 'Yes' or 'No' test
    f. Pure 'glove' & 'stocking' distribution of anesthesia
    g. Sharp defined areas of loss
    h. Loss is absolute & not gradual
    i. Loss of vibration in midline bone – skull, sternum

EXAMINATION OF CEREBELLAR SYSTEM

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Cerebellum is an important part of the brain which is involved in maintaining balance tone and co-ordination. It is customary to speak about cerebellar examination after motor and sensory examination. Though examination of the cerebellar functions or examination starts as soon as the patient enters the OPD room still is it advisable for the students to test each and every function independently in detail.

Cerebellum is examined under the following heads:

1. **Gait-**

The patient is closely observed while enter the OPD room it can itself give a clue. Next step is to ask the patient to walk as normally as he can. The examiner must watch gait under the following headings :

a. If the gait is broad based or not?
b. What happens when he takes turn to return back?
c. Does the patient sway while walking?
d. Does the patient walk through narrow space?

2. **Speech** -

Best thing would be to ask the patient to tell about himself and listen for delivery of words and flow of words. In cerebellar speech there would be explosive speech. Next step is to ask patient to repeat words like " Kurukshetra, Amritsar, British Constitution", etc.

3. **Examination of Eyes –**

Examination of eyes is to check for nystagmus. While describing nystagmus the examiner has to report about the axis of the nystagmus i.e. horizontal or vertical, direction of the fast component and slow component and degree of nystagmus. In the lesions of cerebellopontine angle the nystamus is typically is horizontal nystagmus with fast component to the opposite side.

4. **Tone-**

Tone can be tested while the patient is sitting or lying or even standing. While standing patient can be asked to keep hands out stretched with fingers open and eyes closed . Because of hypotonia there will be drift which will be out and own.

Alternative the patients both arms can be pushed down by the examiner in this case the affected side will further go down.

While sitting patient can be asked to keep arms on table with elbows resting on the table and one observes for attitude of hand . In case of hypotonia the hands will be pointing down.

With the patient lying down the arms and legs can be rolled or moved to check for hypotonia.

5. **Delayed Check (Rebound) responses**

To check Holmes rebound phenomenon examiner stands in front of the patient. Patient is asked to flex his arm and is explained that the examiner will undo what the patient will be doing. The examiner keep his hand little way from patients face so that while over shooting the patient's hand does not accidently hit patient's face.

6. **Truncal titubation** –

Truncal titubation seen in severe cerebellar lesion. Patients body keeps on oscillating, even on sitting.

7. **Abnormal rapid alternating movements (Dysdiadochokinesia)** –

Patient is asked to clap one hand with other . In this procedure one hand is kept flat and with the other alternating movements are done and the examiner observes for speed of movements and if there is any slowing or faltering of movements indicating ipsilateral cerebellar movements.

Dysdiadochokinesia for lower limbs can be tested by asking the patient to rapidly tap the cemented floor with each foot and then lift the foot up. The same movement to be repeated rapidly in a to and fro motion. Both feet are checked alternately.

8. **Action tremors (Active, akinetic, terminal)** -

Evident on purposeful movement, absent at rest. Irregular to and fro jerky movement. Oscillation of outstretched extremity, Increases in amplitude on reaching objects.

9. **Ataxia on Finger Nose Finger -**

This is a test to see ataxia as well as co-ordination. The patient is asked to keep him arm stretched at shoulder and parallel to the ground. He is asked to touch the observer's finger with the tip of the finger so that arm is externally rotated at shoulder and then to bring the finger in to touch his nose tip the process he keeps his arm flexed and palm pointing towards the nose. To make the test even more difficult and meaningful the examining doctor shifts his finger at different places. Commonest mistake in this test is the examining doctor does not keep his finger at arm length.

10. **Heel-knee- shin test** –

This is the equivalent of the finger nose test. The patient is asked to touch shin of tibia with heel at the tibial tuberosity and then to drag it over the shin till ankle and then to lift it in air and again to put it at the shin to repeat the same procedure. Observer looks for ataxia and clumsiness of movements. Commonest mistake that is done is that patient is not explained to lift the leg up after reaching the ankle joint.

11. **Pendular knee jerk -**

It is important to make patient seated in couch in such a manner that knees are about five six inches from the edge of the couch. Patella is struck and oscillation movements are recorded. Only if the oscillations are more than two and half the knee jerk is called pendular. If the thighs are closer to the couch the oscillations will be hampered.

12. **Writing –**

Patient is asked to write few lines and formation of letters and size of letters are observed carefully. In addition one observes for any intentional temors. In cerebellar lesions, there is macrographia.

ANATOMY OF CEEBELLUM

Anteriorly – Pons and Medulla (separated by fourth ventricle and cistern magna)
Superiorly – Tentorium
Inferiorly – cervicomedullary junction

Cerebellar lobes includes:

a. Anterior
b. Posterior
c. Flocculonodular lobe

Functional distribution:

Hemispheres responsible for appendicular coordination

Anterior vermis responsible for gait and other axial functions

Flocculonodular lobe responsible for vestibular and balance function (phylogenetically oldest)

Archiocerebellum (flocculonodular lobe) also called the vestibulocerebellum because it has a number of connections with the vestibular system. It also receives input from areas of the brain concerned with eye movements

Paleocerebellum - vermis of the anterior lobe, the pyramid, the uvula, and the paraflocculus. Also known as the spinocerebellum because it receives input mainly from the spinal cord, it plays a role in the control of muscle tone and the axial and limb movements.

Neocerebellum (corticocerebellum), or cerebrocerebellum- middle portion of the vermis and most of the cerebellar hemispheres. Because it receives projections from the pons, it is also termed the pontocerebellum.

Cerebellum - connected to the brainstem by three large cerebellar peduncles:

1. Inferior cerebellar peduncle (restiform body)
2. Middle cerebellar peduncle (brachium pontis)
3. Superior cerebellar peduncle (brachium conjunctivum)

Most afferent fibers that project to the cerebellum do so through the inferior and middle cerebellar peduncle, whereas efferent fibers from the cerebellum traverse the superior and the inferior cerebellar peduncles

Figure 1 FINGER NOSE TEST

Figure 2 FINGER NOSE TEST

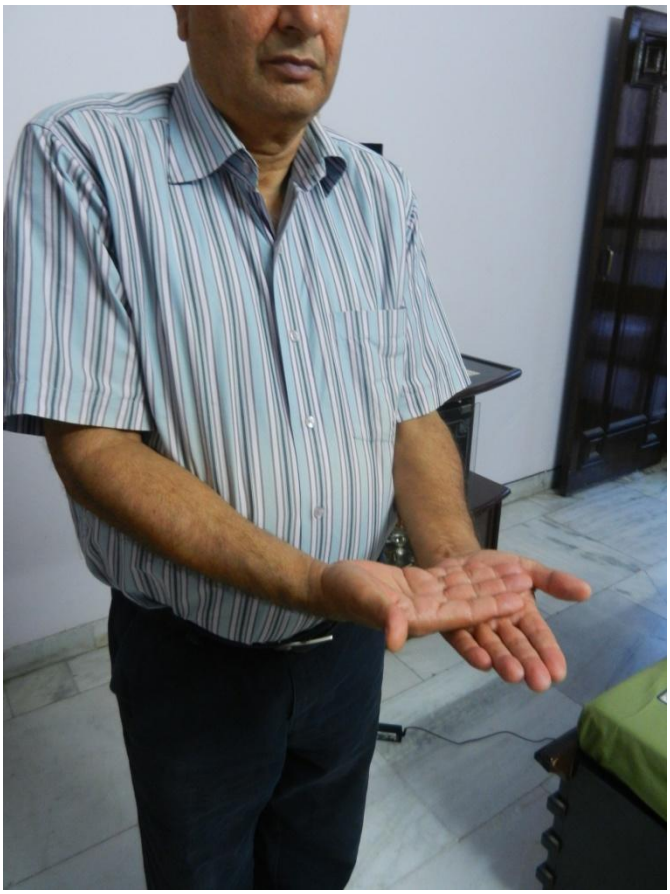
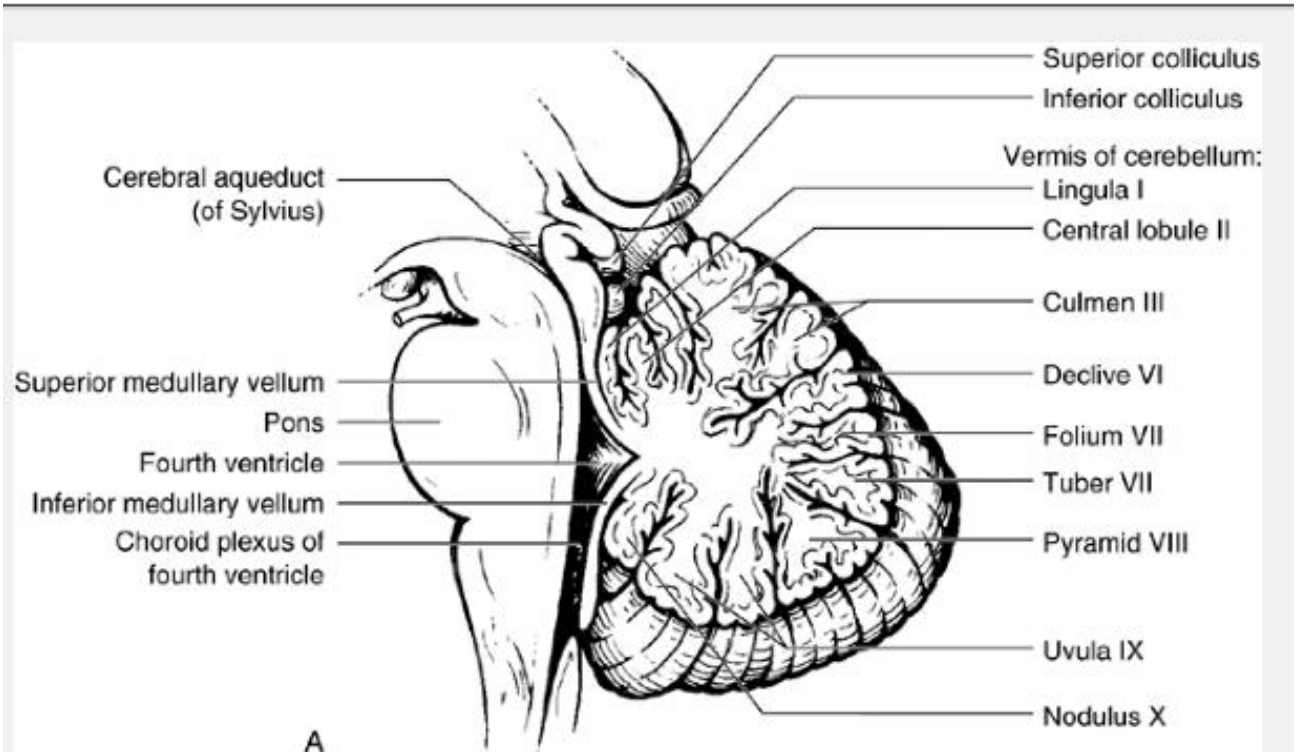


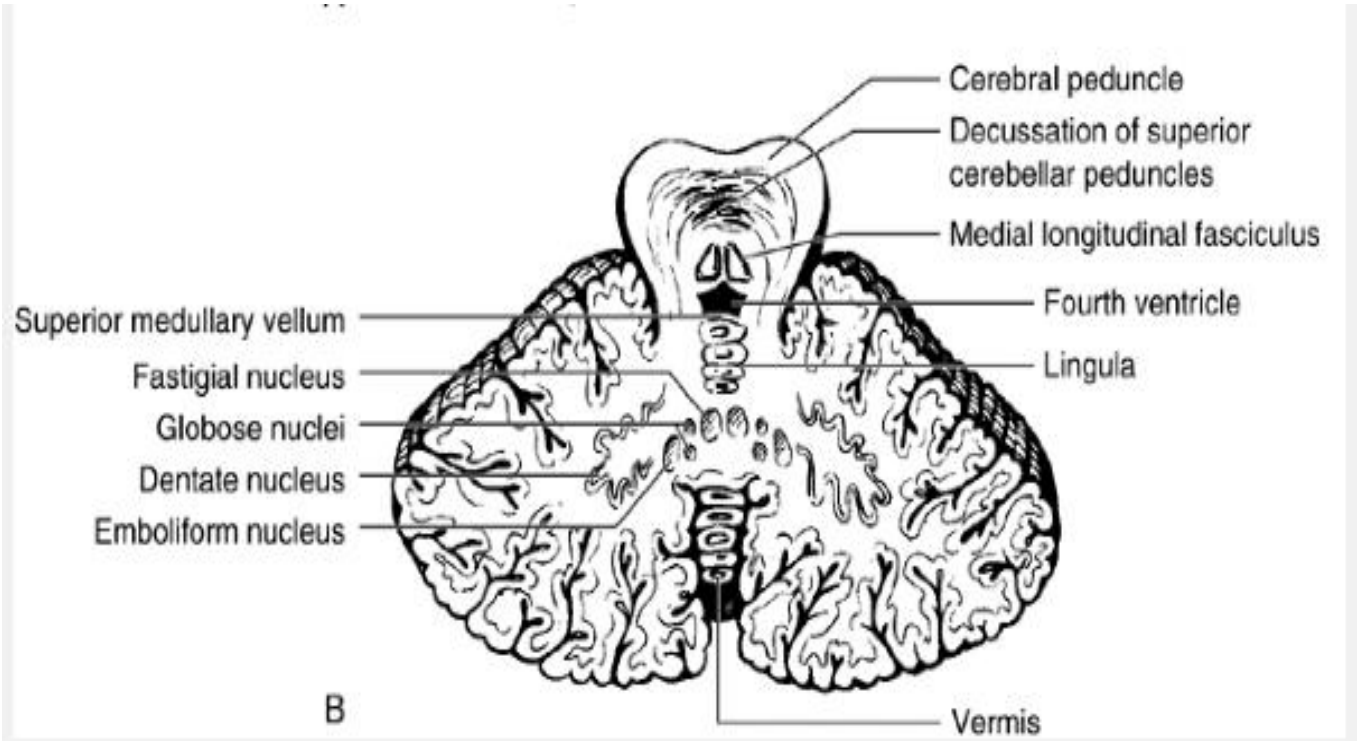
Figure 3 DYSDIADOKOKINESIA

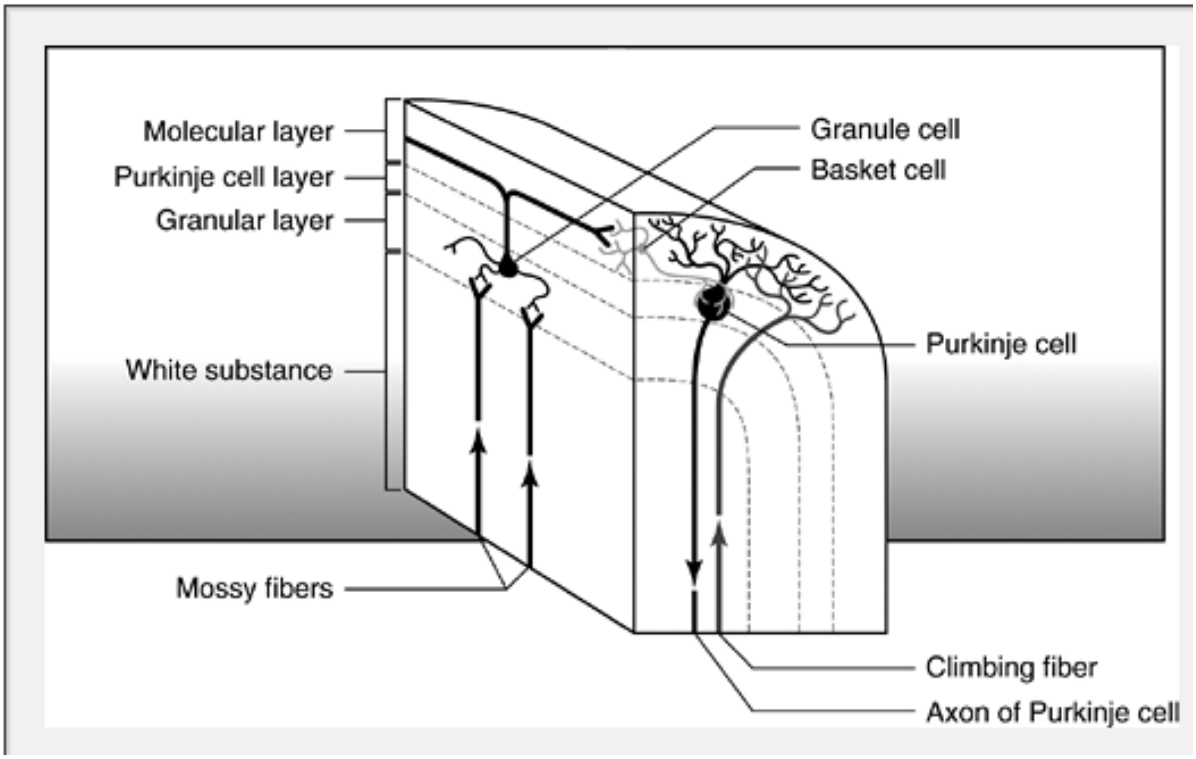
Figure 4 KNEE HEEL TEST

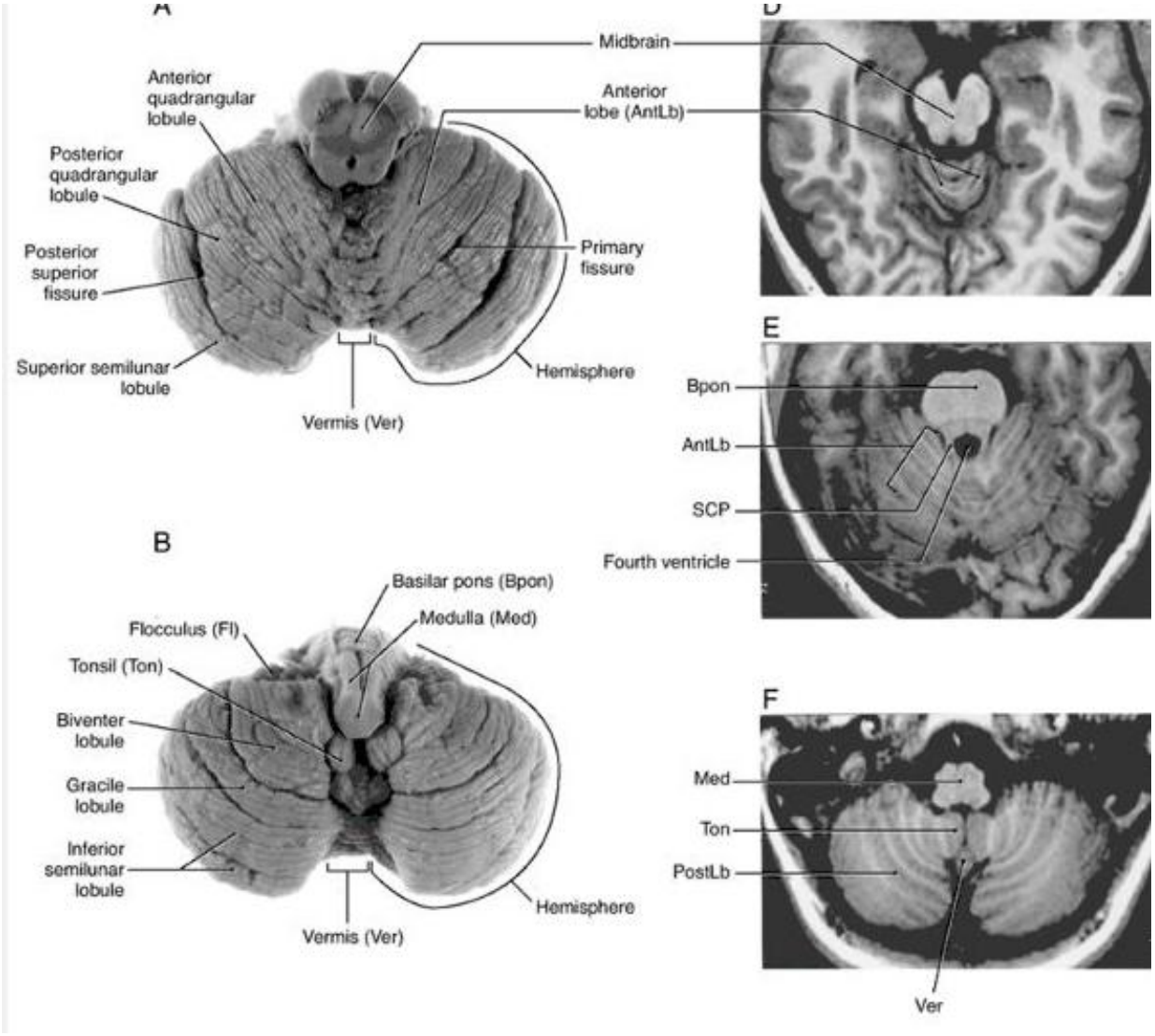
Figure 5 TANDEM WALK

ANATOMY CEREBELLUM




Cerebral peduncle
Decussation of superior cerebellar peduncles
Medial longitudinal fasciculus
Fourth ventricle
Lingula
Superior medullary vellum
Fastigial nucleus
Globose nuclei
Dentate nucleus
Emboliform nucleus
Vermis
B


Molecular layer
Purkinje cell layer
Granular layer
White substance
Mossy fibers
Granule cell
Basket cell
Purkinje cell
Climbing fiber
Axon of Purkinje cell


A
Midbrain
Anterior quadrangular lobule
Anterior lobe (AntLb)
Posterior quadrangular lobule
Primary fissure
Posterior superior fissure
Superior semilunar lobule
Hemisphere
Vermis (Ver)
D
E
Bpon
AntLb
SCP
Fourth ventricle
B
Basilar pons (Bpon)
Medulla (Med)
Flocculus (Fl)
Tonsil (Ton)
Biventer lobule
Gracile lobule
Inferior semilunar lobule
Hemisphere
Vermis (Ver)
F
Med
Ton
PostLb
Ver

EXAMINATION OF RELEASE REFLEXES AND LOBAR SIGNS

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Release Reflexes

Release reflexes or release signs refer to a group of neurological phenomena where primitive reflexes that are suppressed by the cerebral cortex during development are "released from its control" during disease states and can be elicited in adulthood. They are normally present in infants and in a small group of normal adults. These reflexes are seen in unilateral or bilateral frontal lobe disease and aid in localization of disease to the frontal lobe. Although their pathways reside in the parietal lobes, it is the control of the frontal lobe that inhibits them in the normal state. They are not restricted to a specific pathology. The sucking, snout, palmomental and grasp reflexes are the main release reflexes that are tested in routine neurological examination.

The *sucking reflex* consists of sucking movements by the lips when they are stroked or touched. The suck reflex is elicited by lightly touching or tapping on the lips with an object such as a reflex hammer, or the examiner's finger. At times the reflex is obtained merely by approaching the lips with an object.

The *snout reflex* involves puckering or protrusion of the lips with percussion and is elicited by tapping the upper lip. The muscles around the mouth and base of the nose contract to resemble a snout.

In *palmomental reflex,* a stimulus to the thenar area of the hand causes ipsilaterally reflex contraction of the orbicularis oris and mentalis muscles.

The *grasp reflex* is obtained when the examiner's hand is gently inserted into the palm of the patient's hand and the palmar surface is stroked or touched (with the stimulus in a proximal to distal direction) while distracting the patient. The flexor surfaces of the fingers may be stimulated also by the examiner's fingers. With a positive response, the patient grasps the examiner's hand with variable strength and continues to grasp as the examiner's hand is moved. Some clinicians maintain that the reflex is brought out more easily if the patient is lying on the side with the hand to be tested uppermost. The grasp reflex is elicited specifically in patients who have lesions localised to the supplementary motor area.

A *foot grasp reflex* can be elicited by stroking gently the plantar surface medially with a blunt object such as the handle of a reflex hammer. The lateral surfaces of the foot bend as if to make a cup out of the plantar surface. The toes adduct and there is hollowing of the sole with some wrinkling of the skin. If the toes also flex, this is called the tonic foot response.

The *groping reflex* is elicited by repeatedly touching the side of the palm of the patient. In pathological states, a reflex response of the patient's hand following the stimulus and attempting to grasp it is seen. This reflex has no localizing value.

Lobar signs:

Frontal lobe

The clinical effects of lesions of the frontal lobe can be grouped into:

1) Motor abnormalities due to involvement of precentral gyrus and the subcortical white matter
2) Speech and language disturbances when the dominant hemisphere is involved
3) Impairment of cognitive function involving a lack of initiative, diminished attention, concentration and diminished ability to shift from one step to another when a sequence of steps need to be performed to complete a task
4) Lack of initiative and spontaneity
5) Personality and mood changes, disinhibition of behaviour
6) Incontinence of bladder and bowel

The motor abnormality due to a lesion in the *precentral gyrus* results in contralateral spastic monoparesis depending on the area of the lesion. If the lesion is large enough to extend subcortical region, spastic hemiparesis ensues, with significant weakness and functional disability. Involvement of the *premotor cortex* results in less severe degree of paresis, with a delay in executing commands and more spasticity. There is a release of the sucking, groping and grasping reflexes. Lesions of the *supplementary motor area* can result in inability to respond voluntarily to a command to perform movements, but function of the limbs in serial automatic activities such as dressing and walking are preserved.

When the *dominant inferior frontal gyrus* and the Broca's area is involved, there is reduced word output, decrease in spontaneous speech with intact

comprehension and inability to repeat what is presented, resulting in motor or expressive aphasia. Involvement of the *nondominant inferior frontal gyrus* and perisylvian areas results in lack of intonation of speech called dysprosody.

The involvement of the *anterior frontal lobe (prefrontal lobe)* results in various non motor deficits such as diminished attention, vigilance, impaired concentration, impaired performance in tasks that test kinetic melody such as reproducing diagrams of alternating patterns or performing alternating steps such as the fist-ring-palm test. The inability to execute sequential steps to complete a task results in perseveration. There is a lack of initiative or interest in surroundings (abulia and apathy). Akinesia refers to an extreme form in bilateral prefrontal lobe lesions when the patient remains still with no spontaneous movement with no verbalisation (mutism), in the absence of motor weakness. This lack of initiative and slowness is seen with lesions that affect the orbital and dorsolateral frontal lesions.

Personality and mood changes in the form of inappropriate jocularity, social disinhibition and are seen with lesions of the medial and orbital parts.

Bilateral frontal lobe lesions result in involvement of gait referred to as *apraxia of gait*, described by Bruns as an "ataxia". These patients have normal motor power but have a difficulty in initiating walking, take short stuffled steps with imbalance. There may or may not be spasticity of the limbs.

Patients with unilateral (right or left) or bilateral lesions involving the *superior frontal and anterior cingulate gyri* and the intervening white matter have loss of control of micturition and defecation. The earliest symptom is that of urge incontinence. In the complete form of the syndrome, the patient has sudden incontinence due to emptying of bladder/bowel and may be embarrassed by it by posteriorly placed lesions. If the anterior nonmotor parts of the frontal lobe are involved the resultant incontinence will be accompanied by an indifference to the incontinence. The clinical manifestations of frontal lobe involvement have been summarized in Table 1.

Table 1. Clinical manifestations of frontal lobe disease

Unilateral	**Side specific**	**Bifrontal**
Contralateral spastic hemi- or monoparesis, incontinence	*Dominant (usually left)* – Aphasia	Bilateral hemiparesis with pseudobulbar palsy, emotional incontinence
Increased talkativeness, loss of initiative	*Right* – Dysprosody	Apraxia of gait, urinary and faecal incontinence
Release of grasp, sucking reflexes		Abulia, akinesia, apathy
Anosmia if orbitofrontal lobe is involved		Release of primitive reflexes

Temporal lobe

The clinical effects from lesions of the temporal lobe can be classified into those affecting: 1) language; 2) cognitive manifestations and seizures; 3) visual functions; and 4) auditory functions.

Lesions of the *dominant posterosuperior temporal lobe* involving the Wernicke's area results in the syndrome of *receptive aphasia*, wherein there is impaired comprehension of spoken or written commands with speech being fluent and paraphasic with substitution of one word for another. In pure word deafness, there is impairment of auditory comprehension and inability to write to dictation but spontaneous writing and ability to comprehend written language is preserved.

One of the most important symptoms that help localisation of lesions in the temporal lobe is the semiology of seizures that arise from the temporal lobe. Seizures with olfactory hallucinations point to a *medial temporal lobe lesion*. Seizures with vertigo as an aura can aid in localizing the lesion to the *superior temporal gyrus* posterior to the primary auditory cortex. Complex partial seizures of other types with automatisms can also point to a lesion in the temporal lobe.

Lesions of the medial temporal lobe involving the *fornix* can result in memory disturbances, particularly for recollection of recent events. There may be impaired

recollection of verbally presented memory in dominant forniceal lesions and impaired recollection of visually presented memory in nondominant forniceal lesions.

Involvement of the lower arching fibres of the geniculocalcarine pathway *(Meyer's loop)* in the white matter of the central and posterior part of the temporal lobe results in contralateral congruent superior homonymous quandrantanopia. Visual hallucinations of a complex form, where well constructed images are seen by the patient, can occur in lesions of the middle and inferior temporal gyri that have connections with the occipital cortex. There may be alterations in perception of the objects, with the object appearing too large (macropsia) or too small (micropsia).

Bilateral temporal lobe lesions involving the *transverse gyri of Heschl* (situated in the depth of the sylvian fissure on the posterior superior surface) result in cortical deafness with elevation of threshold of hearing. Some of these patients may not be aware of their deafness. It has been found that in unilateral lesions, there could be impairment of hearing in the contralateral ear that may not be recognised by the patient, except if specific audiometric tests are performed.

Lesions of the *secondary auditory cortex* result in *auditory agnosias*. The patient has no difficulty in perceiving the sound and pure tones but has difficulty in recognising what the sound is. Amusia, is a specific defect with impaired appreciation of music. In *nondominant hemispheric involvement*, there is impaired recognition of harmony and melody (in the absence of words) of music. Appreciation and naming of the musical scores, writing and reading of music requires intact functioning of the *dominant temporal lobe*.

Lesions of the superolateral part of the dominant temporal lobe can result in auditory illusions or hallucinations. In auditory illusions, words are perceived to be too loud or soft than normal. There may be a sense of repetition of the words being heard. In auditory hallucinations, elementary sounds such as murmurs, blowing of air, whistles etc. may be heard. Complex hallucinations involve musical themes and voices. The clinical manifestations of involvement of temporal lobe have been summarized in Table 2.

Table 2. Clinical effects of temporal lobe disease

Unilateral (Either)	**Side specific**	**Bilateral**
Contralateral superior quadrantanopia	*Dominant* – Wernicke's aphasia, amusia, Impaired naming, visual agnosia, Impaired performance in tests of verbal material presented through auditory sense	Korsakoff's amnesia Apathy, placidity, increased sexual activity, sham rage (Kluver Bucy syndrome)
Auditory, visual, olfactory hallucinations, complex partial seizures, Delirium	*Nondominant* – Inability to judge spatial relationships, Agnosia for sounds, Impaired performance in tests of visually presented nonverbal material	

Parietal lobe

The clinical effects of parietal lobe dysfunction may be grouped into: 1) cortical sensory deficits; 2) impaired perception of the body's image and its relation to the surroundings; and 3) visual symptoms.

Patients with unilateral parietal lobe dysfunction involving the *postcentral gyrus* have contralateral impairment of perception of cortical sensations such as impaired sensory discrimination, inability to distinguish objects by their size, shape and texture (astereognosis), inability to recognise figures written on the skin, impaired two point discrimination and impaired ability to detect direction of movement of tactile stimulus. To diagnose cortical sensory deficit, it is mandatory that the primary modalities of touch, pain and temperature are perceived normally by the patient.

In lesions usually of *nondominant (right) parietal lobe*, there is profound neglect of the opposite side of the body and the environment. With this hemi-neglect (anosognosia), patient may ignore objects on the side opposite the involvement as well as his own body parts. This is one of the manifestations of unilateral asomatognosia (Anton-Babinski syndrome) seen more commonly with right sided lesions than with left sided ones. "Dressing apraxia" is a condition with neglect of one

side of the body in dressing and grooming. These patients also have difficulty in relating to their extrapersonal space and have difficulty in copying diagrams, drawing a clock or geometric figures (constructional apraxia). They also may have impaired topographic and geographic memory impairment with resultant difficulties in finding their way around.

Bilateral asomatognosia is also called Gerstmann syndrome and consists of finger agnosia, left-right confusion, acalculia and agraphia and is localised to the dominant side angular gyrus. In dominant parietal lobe lesions, patients will be able to copy a diagram but will not be able to orient the parts correctly due to visuo-spatial skills being impaired.

Lesions of the *deep parietal lobe white matter* result in an incongruous contralateral homonymous hemianopia with the inferior visual fields being more affected. The component of optokinetic nystagmus that occurs on ipsilateral movement of the target will be abolished. There may be deficits related to guiding the arm to the object that is presented in the visual field with bilateral lesions (optic ataxia).

In the absence of motor and sensory dysfunction, dominant parietal lobe lesions can result in ideational and ideomotor apraxia with resultant inability to perform learned motor skills.

The clinical manifestations of parietal lobe disease have been summarized in Table 3.

Table 3. Clinical effects of parietal lobe disease

Unilateral (either side)	**Side specific**	**Bilateral**
Contralateral cortical sensory impairment Contralateral mild hemiparesis with marked atrophy (parietal wasting)	*Dominant* – Alexia, Gerstmann syndrome, Tactile agnosia, Bilateral ideational and ideomotor apraxia	Visuospatial imperception Optic ataxia Spatial disorientation
Incongruous contralateral homonymous hemianopia	*Nondominant* – Contralateral hemineglect Topographic memory	

	impairment, Dressing apraxia, Constructional apraxia	
Abolition of opticokinetic nystagmus with target moving towards the side of lesion		

Occipital lobe

The most common finding with an occipital lobe lesion is contralateral congruous homonymous hemianopia. Cortical blindness is a condition resulting from *bilateral lesions of area 17* (primary visual cortex) in which there is loss of sight and reflex closure of eyelids to bright light or a threat in the visual field. The pupillary light reflexes are however preserved. In Anton syndrome or visual anosognosia, there is denial of blindness by the patient and the lesion extends beyond the striate cortex to the visual association areas.

Elementary visual hallucinations such as flashes of light can occur with occipital cortex lesions while formed visual hallucinations (like images of persons) occur with lesions in the temporal lobe. These visual aura may be part of a seizure aiding in localizing a lesion to the occipital lobe. Visual illusions are of numerous types and have poor localizing value.

In *dominant occipital lobe lesions*, there is a failure to name and indicate the use of a seen object by spoken or written word or gesture – termed as visual object agnosia. Simultagnosia is an inability to perceive simultaneously all the elements of a scene (such as a picture presented to the patient) and to properly interpret the scene. This is usually seen with lesions of the inferolateral part of the dominant occipital lobe or with bilateral lesions. Lesions of *bilateral ventromesial occipitotemporal* regions result in defects in identifying familiar faces by looking at the person or a picture. This is termed prosopagnosia.

In Balint syndrome, seen in *bilateral lesions of the parieto occipital regions* (involving areas 19 and 17) there is an inability to look voluntarily into and to scan the peripheral field although the extraocular movements are normal in range. This is

termed as psychic paralysis of fixation of gaze. There is a failure to precisely grasp or touch an object under visual guidance. There is associated visual inattention.

In colour agnosis, there is disturbance in distinguishing between hues of different colours of wool presented to them. This is usually seen in *bilateral lesions of the inferomesial occipital lobes*. In colour anomia, there is an inability to name colours although they can match colours. This is seen with left sided mesial occipital lesions close to the junction with the temporal lobes. The clinical manifestations of parietal lobe disease have been summarized in Table 4.

Table 4. Clinical effects of occipital lobe disease

Unilateral (Either side)	Side specific	Bilateral
Contralateral congruent homonymous hemianopia	*Left*-Alexia and colour naming defect Visual object agnosia	Anton syndrome Cortical blindness Balint syndrome Loss of colour perception Prosopagnosia Simultagnosia
Elementary visual hallucinations	*Right*- Visual illusions and hallucinations, Loss of visual orientation	

Suggested Reading

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EXAMINATION OF UNCONSCIOUS PATIENT

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Consciousness has two components, namely arousal and content. Level of consciousness can be assessed by the Glasgow Coma Scale[1] (GCS) shown in table 1. GCS ≤ 8 is a generally accepted operational definition of coma.

Anatomy of consciousness:

Reticular activating system (RAS) is the vital complex pathway responsible for maintenance of consciousness. The RAS is composed of several neuronal circuits connecting the brainstem to the cortex. These pathways originate in the upper brainstem reticular core and project through synaptic relays in the rostral intralaminar and thalamic nuclei to the cerebral cortex. As a result, individuals with bilateral lesions of thalamic intralaminar nuclei are lethargic or somnolent. Several areas traditionally included in the RAS are:

- Midbrain Reticular Formation
- Mesencephalic Nucleus (mesencephalon)
- Thalamic Intralaminar nucleus (centromedian nucleus)
- Dorsal Hypothalamus
- Tegmentum

Table 1[1]: Glasgow coma scale for age >4 yr [Total 15; min 3 and max 15]

Eye opening	Best verbal response	Best motor response

Open spontaneously -4 Open only to verbal stimuli - 3 Only to pain -2 Never open -1	Oriented and converses -5 Converses, but disoriented, confused -4 Uses inappropriate words -3 Makes incomprehensible sounds -2 No verbal response - 1	Obeys commands - 6 Localizes pain - 5 Exhibits flexion withdrawal - 4 Decorticate rigidity -3 Decerebrate rigidity - 2 No motor response -1

How to approach an unconscious patient?

Approach to the unconscious patient should be in a step-by-step manner (illustrated in the underlying flowchart) in order to reveal all the possible etiologies.

1) Cardiovascular evaluation: Assess airway, breathing & circulation [CPR if necessary]
2) Stat Investigations: S. glucose, S. electrolyte, arterial blood gas analysis CBC, S.creatinine, S.ammonia, S.calcium and antiepileptic drug serum levels if patient is on antiepileptic drugs,
3) **Core neurological examination:**
   - **A) Respiratory rate & pattern:**
     1) Cheyne-Stokes respiration :

        Pattern: Crescendo - decrescendo - expiratory pause - crescendo repetition of cycle

        Etiology: It results from diencephalic or bilateral cerebral hemisphere lesion due to raised ICP or metabolic abnormality leading to increased response to CO_2.

     2) Hyperventilation :

It occurs due to dysfunction within pons and is seen usually in response to hypoxia, metabolic acidosis, aspiration or pulmonary edema.

3) Cluster breathing :

Consists of periods of rapid irregular breathing separated by apnoeic spells due to upper medullary or lower pontine lesion.

4) Apneustic breathing:

Consists of a pause at full inspiration due to pontine lesions.

5) Ataxic breathing :

No fixed rate or pattern of breathing, occurs due to medullary lesion.

B) Pupils:

1) Equal and reactive pupils : Toxic or metabolic cause
   - Only metabolic cause of fixed and dilated pupil: glutethemide toxicity, anoxic encephalopathy, anticholinergic drugs, botulinum toxin, etc.
   - Narcotic agents cause small pupils with a small range of constriction and sluggish reaction to light.

2) Unequal pupil:- {Afferent pupil does not produce anisocoria}
   - Fixed and dilated pupil: 3rd nerve palsy
   - Horner syndrome : Carotid occlusion/dissection

3) Bilateral pupil abnormality:
   - Pinpoint pupil:- Pontine lesion
   - Bilateral fixed and dilated (7-9mm): Subtotal damage to medulla, postanoxic state and hypothermia (temp<90$^{0)}$.

- Midposition and fixed (4-6mm): Extensive midbrain lesion [involvement of both sympathetic and parasympathetic].

C) Extraocular muscle function:

1) Gaze deviation at rest:

A) Bilateral gaze deviation:

1) Frontal lobe lesion: Deviation occurs to the ipsilateral side to the destructive lesion and contra lateral side to the irritative lesion like seizure focus.

2) Pontine lesion: Deviation towards the contra lateral side of lesion and ipsilateral to hemiparetic side.

3) Thalamic lesion: known as 'wrong way gaze'. Gaze deviated towards the contra lateral side of lesion and ipsilateral to hemi paretic side.

4) Tectum of midbrain: Parinaud's syndrome.

B) Unilateral gaze deviation toward the side of larger pupil: 3^{rd} nerve palsy.

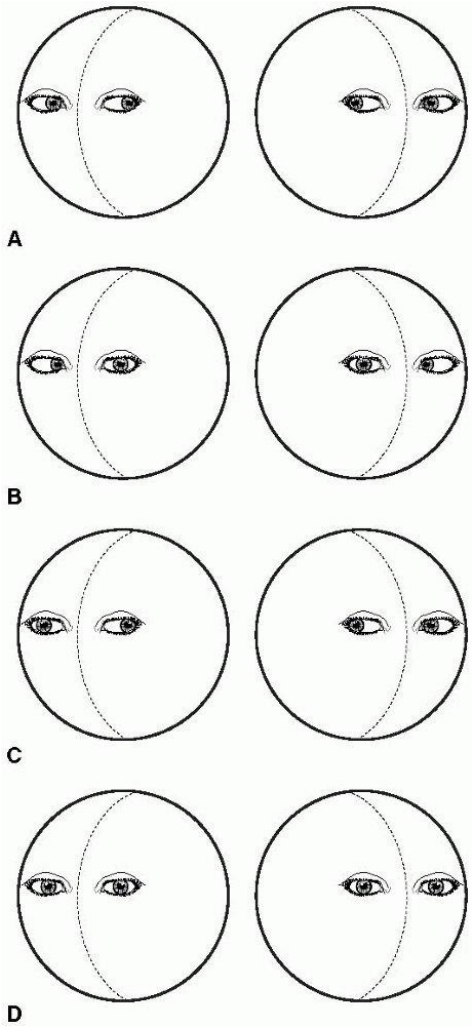
C) Unilateral inward deviation: VIth nerve palsy

D) Skew deviation:

- 3^{rd} or 4^{th} nerve / nucleus lesion

- Infratentorial lesion (dorsal midbrain)

FIGURE: Examples of oculocephalic responses that may be seen in comatose patients. When the brainstem is intact, the eyes move in the opposite direction from head rotation. **A.** Normal response, the usual response in a patient with metabolic encephalopathy. **B.** Bilateral sixth nerve palsies. **C.** Right third nerve palsy or internuclear ophthalmoplegia. **D.** Absent response, seen when the reflex pathways are impaired.



2) Spontaneous eye movements:

A) "Windsheild wiper eyes": Random roving conjugate eye

Movement.

B) Periodic alternate gaze: Bilateral cerebral dysfunction

C) Ocular bobbing: Repetitive rapid downward deviation followed by neutral position of the gaze. Seen in pontine lesion.

3) Internuclear ophthalmoplegia (INO):

Occur due to lesion in medial longitudinal fasciculus (MLF). Fibre crossing the contralateral to 3rd nerve nucleus is interrupted. On spontaneous or reflex eye movement, eye of the ipsilateral side of lesion fail to adduct.

4) Reflex eye movements:

a) Oculovestibular reflex (caloric test)

b) Oculocephalic reflex (doll's eye)

D) Motor:

Muscle tone and reflexes, response to pain and babinski (note asymmetry)

1) Asymmetrical: supratentorial lesion

2) Inconsistent/ variable: seizures, psychiatric

3) Symmetric: metabolic, asterexix, tremor, myoclonus in metabolic coma

4) hyporeflexia: myxoedema coma

5) Patterns:

-Decorticate rigidity: arms flex, legs extended; cortical or subcortical lesion

-Decerebrate rigidity: arms and leg extended; injury below lower midbrain

- Arm flexed and leg flaccid: pontine tegmentum - arm flaccid and leg appropriate: ("man in barrel syndrome") anoxic injury

E) Ciliospinal reflex:

- Pupil dilatation after noxious cutaneous stimuli.

- It test the integrity of sympathetic pathway

- If present bilateral, suggestive of metabolic cause

- If present unilateral, suggestive of 3rd nerve lesion or Horner's Syndrome.

- If bilateral absent, inconsistent.

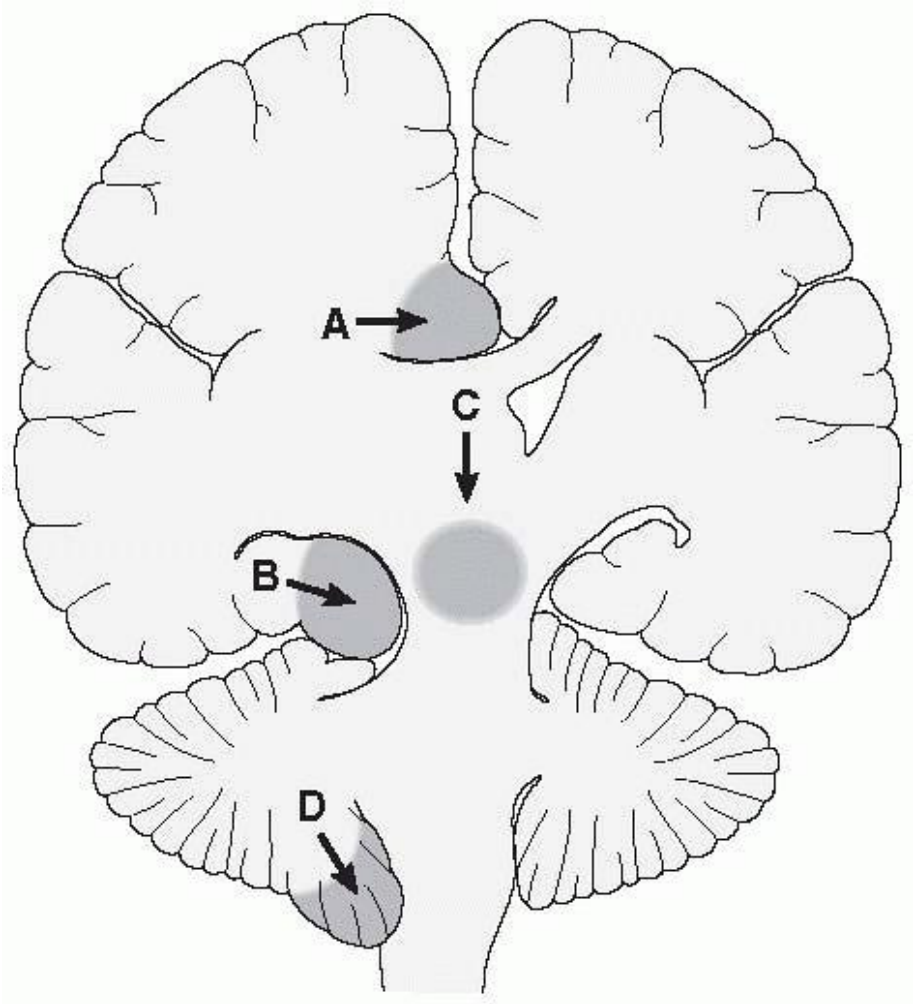


FIGURE 2[4] • Patterns of brain herniation. **A.** Herniation of the cingulate gyrus under the falx cerebri. **B.** Uncal (lateral transtentorial) herniation. **C.** Central transtentorial herniation. **D.** Herniation of the cerebellar tonsils through the foramen magnum.

- **Clinical manifestations of common herniation syndromes**

1) Central transtentorial herniation:

Impaired consciousness, abnormal respirations, symmetric small or midposition fixed or minimally reactive pupils,
decorticate evolving to Decerebrate posturing, rostrocaudal deterioration

2) Lateral transtentorial (uncal) herniation:

Impaired consciousness, abnormal respirations, third nerve palsy (unilaterally dilated pupil), hemiparesis (may be false localizing), rostrocaudal deterioration

3) Cerebellar tonsillar (foramen magnum) herniation:

Impaired consciousness, neck rigidity, opisthotonos, decerebrate rigidity, vomiting, irregular respirations, apnea, bradycardia

4) Upward herniation:

Prominent brainstem signs, downward gaze deviation, up gaze palsy, decerebrate posturing (usually due to a cerebellar mass lesion).

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1) Teasdale G, Jennett B: Assessment of coma and impaired consciousness. A practical scale. Lancet 2: 81-4, 1974.

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