Nystagmus
NYSTAGMUS

• Nystagmus is characterized by rhythmic oscillations of the eyes in which each phase is of equal amplitude.

• Nystagmoid or saccadic intrusions: oscillatory, but not rhythmic.

• Pendular nystagmus: movements with equal speed in both directions.

• Jerk nystagmus: slow deviation on one side (slow phase) followed by a rapid movement in the opposite direction (rapid phase).
Important features to consider when describing a nystagmus:

1) **direction**
   (in the jerk nystagmus it is that of the rapid phase),

2) **Plane of oscillation**
   (horizontal, vertical, rotational),

3) **amplitude** (fine, medium, large)

4) **frequency** (low, high).
# Nystagmi

## Physiologic

1) End point nystagmus
2) Optokinetic nystagmus
3) Vestibular nystagmus

## Congenital

1) Congenital nystagmus
2) Latent nystagmus
3) Spasmus mutans

## Acquired

1) Peripheral vestibular nystagmus
2) Central vestibular nystagmus
3) Gaze-evoked nystagmus
4) Periodic alternating nystagmus
5) Rebound nystagmus
6) Pendular nystagmus
7) See-saw nystagmus
8) Sensitive nystagmus
9) Convergence retraction nystagmus
10) Dissociated nystagmus
1) End point nystagmus

2) Opticokinetic nystagmus

3) Vestibular nystagmus
1) End-point nystagmus

- Jerk nystagmus commonly found in extreme positions of gaze, with rapid phase in the direction of gaze;

  It may present:
  - after prolonged (>10-15 sec) deviations of the eye in extreme positions
  - immediately in extreme positions, but transitory, ending after a few seconds,
  - permanently in extreme positions but may regress if the gaze returns 15-20° towards the primary position.
  - Anomalies of ocular motility are not present in any of the three forms.
2) **Opticokinetic nystagmus**

- Jerk nystagmus caused by the repetitive movement of an image in the visual field.

- The slow phase is a pursuit in which the eye follows the object while the rapid phase is a saccade which brings the eye in the primary position in order to fixate the subsequent object.

- The opticokinetic nystagmus is useful to demonstrate the non organic nature of a visual defect.
3) Vestibular nystagmus

• Due to the movement of endolymph towards the ampulla of one side, induced by both passive movements of the body or head, and the irrigation of the ear with warm water,

• This causes a slow movement of the eyes to the opposite side (the vestibule projects towards the contralateral horizontale gaze centers) followed by a rapid compensating phase in the opposite direction.

• The nystagmus “beats” towards the stimulated side (in the direction of head movements or towards the ear irrigated with warm water).

• Cold water irrigation causes, instead, the opposite phenomenon, with inhibition of the utricle and a nystagmus which “beats” towards the opposite direction.
Caloric testing
1) Congenital nystagmus
2) Latent nystagmus
3) Spasmus mutans
CONGENITAL NYSTAGMUS

1) Congenital nystagmus

- Present at birth or, more frequently, in the first months of life.

- Binocular, of similar amplitude in both eyes, it may be pendular or jerky in the primary position, while in the lateral position it is generally jerky.
It is characterized by:

- the movement occurs in a single plane, generally the horizontal plane, and remains the same in all gaze positions;
- the oscillation is independent from the position of the head, increases with fixation and is reduced by the convergence or closing of the eyes.
- it is often identifiable in a particular position of the eyes, known as the «cancellation position», in which it is inhibited or changes its direction.
- An anomalous position of the head is often present to allow the cancellation area to coincide with the primary position of gaze.
- Another characteristic is the inverison of the opticokinetic nystagmus (the nystagmus occurs to the opposite side with respect to normal conditions).
2) *Latent nystagmus*

- Latent nystagmus is a type of jerk nystagmus only present in monocular vision: when an eye is closed, a binocular and conjugated oscillation occurs, with rapid phase directed towards the viewing side.

  - this nystagmus is always associated to strabismus (generally esotropia) and amblyopia is frequently present.

- Latent nystagmus follows *Alexander’s rule*, that is it increases in amplitude when the gaze is in the direction of the rapid phase.

- The origin of this nystagmus is unknown and therapy must be directed towards the prevention of amblyopia.
3) *Spasmus mutans*

- This syndrome is characterized by the association of nystagmus, oscillations and anomalous positions of the head (stiff neck).

- It generally presents in the first year of life, and spontaneously regresses 1-2 years after onset.

- The nystagmus is irregular, often horizontal, with high frequency and reduced amplitude.

- The amplitude is typically different in the two eyes and monocular forms are also present.
• **Acquired spasmus mutans** may be an early sign of tumors of the visual pathways, in particular of the optic chiasm, or brainstem.

• The diagnosis must therefore be of exclusion, and neuroradiologic examinations are necessary (CT and/or MRI) in the presence of any signs of alterations of the optic nerve or chiasmatic function.
Acquired nystagmus

1) Peripheral vestibular nystagmus
2) Central vestibular nystagmus
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4) Periodic alternating nystagmus
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**1) Peripheral vestibular nystagmus**

- It is caused by a dysfunction of the labyrinth or vestibular nerve causing an asymmetry in the signal from the right and left semicircular canals.

- There are violent vestibular dysfunctions with vertigo, and low hearing. The nystagmus is one-directional with rapid phase directed towards the healthy side. The amplitude of the oscillation increases when the gaze is in the direction of the rapid phase (*Alexander’s rule*) while the trajectory is generally mixed, horizontal-torsional, never purely vertical or torsional.

- Fixation reduces the nystagmus as the movements due to the visual stimuli are normal and balance the oscillation caused by the vestibular dysfunction. Oscillopsia and positive Romberg’s test are common, with the direction of the fall depending on the position of the head.

- The most common causes include vestibulitis, neuronitis, ischaemic lesions, traumas, and drug toxicity.
2) Central vestibular nystagmus

- It is caused by a lesion of the central vestibular connections (the vestibular nuclei and their projections, including the vestibulocerebellum).

- Vestibular symptoms are generally less accented and longer lasting with respect to the peripheral form.

- Fixation does not inhibit the nystagmus and the position of the head does not influence Romberg’s test. Vestibulo-ocular movements and pursuit are frequently altered.

- Neuroradiologic examinations are always indicated.
Types of central and peripheral vestibular nystagmus:

• **Vertical nystagmus, downwards.** Cerebellar lesions, lesions of the cranial base-spinal cord junction (*Arnold-Chiari malformations*) or of regional vessels. Nystagmus is present in the primary position and in the lateral gaze and increases in the downwards gaze.

• **Vertical nystagmus, upwards.** Intrinsic lesions of the brainstem or cerebellum, due to ischaemia, demyelination or tumors. Nystagmus is present in the primary position, and has maximum amplitude in the upwards gaze, but differently from the preceding nystagmus, does not increase in the lateral gaze.

• **Torsional nystagmus.** A rare form, suggestive of lesions of the brainstem such as syringobulbia, syringomyelia, Wallenberger’s s. and vascular alterations.

• **Horizontal nystagmus.** Due to peripheral vestibular dysfunctions, it may also present as a consequence of central lesions. The oscillation is not reduced by fixation and is associated to homolateral pursuit anomalies.
3) Gaze-evoked nystagmus

- A rare form of nystagmus with low frequency jerks similar to end-point nystagmus, except for the fact that it presents in less extreme gaze positions and has greater amplitude.

- It is due to alterations of the neuronal integrator in the control of saccadic pulse-step. The eyes cannot maintain an eccentric position and return slightly backwards towards the primary position. The rapid phase brings the gaze back to the desired direction.

- Smooth pursuits and fixation are also frequently altered.

- It is associated to cerebellar pathologies but may present following hemispheric or brainstem lesions which affect conjugated gaze mechanisms (gaze-paretic nystagmus).
- The most common cause of this nystagmus is the use of sedatives and antiepileptic drugs.
4) **Periodic alternating nystagmus**

- It is a horizontal jerk nystagmus present in the primary position characterized by periodic changes in direction (approx. every 2 minutes)
- It may be congenital or secondary to cerebellar or brainstem pathologies, or due to visual deprivation. *Smooth pursuit* and opticokinetic nystagmus are also often altered.
- Badofen, myorelaxing and antispastic drug, is able to control the nystagmus in the acquired form.

5) **Rebound nystagmus**

- It consists in a horizontal jerk nystagmus with rapid phase initially directed towards the direction of gaze but which changes direction after few seconds of eccentric position.
- It primarily occurs following alterations of the cerebellar parenchyma but also following lesions of the posterior cranial fossa.
6) **Pendular nystagmus**

- A nystagmus frequently encountered after vascular or demyelinating lesions of the brainstem or following monolateral visual defects.
- It is characterized by pendular movements with mixed horizontal, vertical and torsional trajectory; these characteristics differentiate it from the congenital form of pendular nystagmus in which the oscillation is purely horizontal and becomes jerky in the lateral gaze.
- Internuclear ophthalmoplegia, skew deviation and nystagmus with upwards jerks are frequently associated anomalies.

7) **See-saw nystagmus**

- A rare form of pendular nystagmus characterized in the first half of the cycle by the elevation and intorsion of an eye with synchronous depression and extratorsion of the other eye; in the other half of the cycle the movement is inverted.
- Frequently associated to tumours of the chiasmatic region.
8) Sensitive nystagmus

- It presents following lesions of the afferent visual system with loss of central vision. In this situation, the capacity of maintaining stable foveation is lost due to micromovements of the fixation system.

- The severity of the nystagmus depends on the severity of the visual defect. In the case of bilateral visual reduction, the nystagmus is conjugated and prevalently pendular, is reduced by convergence and often accompanied by oscillations of the head if the visual defect presents in the early months of life.

- When the visual defect is monolateral, fixation instability with prevalently vertical oscillations, more evident on the affected side, are present.
9) **Convergence retraction nystagmus**

- This anomalous movement is frequently found in the posterior mesencephalus syndrome (*Parinaud’s s.*).

- It is characterized by rapid phases which cause the convergence and retraction of the eyes in the upward gaze. In fact, this is not a real nystagmus, but rather an alteration of the saccadic system with asynchrononous, opposite, adduction movements.

10) **Dissociated nystagmus**

- It is characterized by oscillations which are asymmetric between the two eyes for amplitude and/or trajectory
Convergence retraction nystagmus
Convergence retraction nystagmus

Convergence retraction

convergence

downward rotating drum

Convergence retraction nystagmus
Pupillary light reflex: near dissociation in Parinaud's syndrome
OTHER OCULAR OSCILLATIONS

1) Square-wave jerk
2) Flutter and opsoclonus
1) *Square-wave jerk*

- These are saccadic intrusions which interrupt the fixation and which, after a normal intersaccadic interval of 200 msec, are followed by the return to the initial position.

- They represent an exaggeration of normal microsaccadic movements associated to fixation, and with small amplitude, may be considered normal, especially in the elderly; if ample (1-5°) and frequent, they are generally secondary to cerebellar pathology or to progressive supranuclear paralysis.

- If movement amplitude is greater than 10° they are known as *macro square-wave jerks*.
2) Flutter and opsoclonus

Ocular oscillations without intersaccadic interval.

- **Flutter**: oscillations on the horizontal plan in the form of pulses of 3-4 microoscillations which interrupt fixation in the primary position.

- **Opsoclonus**: involuntary, chaotic, multivectorial (horizontal, vertical, diagonal) movements. This anomaly, also known as *saccadomania*, disappears during sleep.

- *Flutter and opsoclonus* are frequent in the course of cerebellar pathologies and following postviral encephalitis.
  - They may also be remote (paraneoplastic) manifestations of neuroblastoma in the child and visceral carcinoma in the adult.