NYSTAGMUS

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NYSTAGMUS

Nystagmus is the rhythmic oscillation of the eyes that can be physiological or pathological. It is not a diagnosis but rather a manifestation of an underlying disease usually due to vestibular or cerebellar disease.
The rhythmic oscillation may be symmetrical. If the speed to one side is the same as the speed to the opposite side, it is a **Pendular Nystagmus.**

Rhythmic oscillation may be asymmetrical. If the speed to one side is slower than the speed to the opposite side, then it is a **Jerk Nystagmus.** There is a slow drift and a fast recovery movement.
The slow drift is the pathological component that moves away from the object of regard. The fast phase is the corrective component.

However by convention, a nystagmus is described by the direction of the fast phase. It can be in primary gaze or secondary positions as in gaze-evoked nystagmus.

JERK NYSTAGMUS

The direction is defined by the direction of the fast phase. The slow phase is first and pathologic and the fast phase is second and corrective.

Spontaneous Jerk nystagmus in primary gaze is classified according to direction:

- Horizontal
- Vertical
- Upbeat or downbeat
- Torsional
- Oblique
- Mixed

JERK NYSTAGMUS

Jerk nystagmus that occurs only in an eccentric position is called **gaze-evoked nystagmus.** Gaze-evoked nystagmus is due to a defect of the central neural integrators. Gaze-paretic nystagmus is due to weakness of extra-ocular muscles.

PHYSIOLOGIC NYSTAGMUS

Physiological nystagmus is also known as end-point nystagmus. This occurs when a person looks laterally beyond 40° for an extended period (Figure 5.1). It results in a low amplitude horizontal nystagmus. There is an initial slow phase which is due to the elastic pull of the eye muscles. The corrective phase is back to the eccentric position, is faster and is due to the brain's gaze-holding integrator.

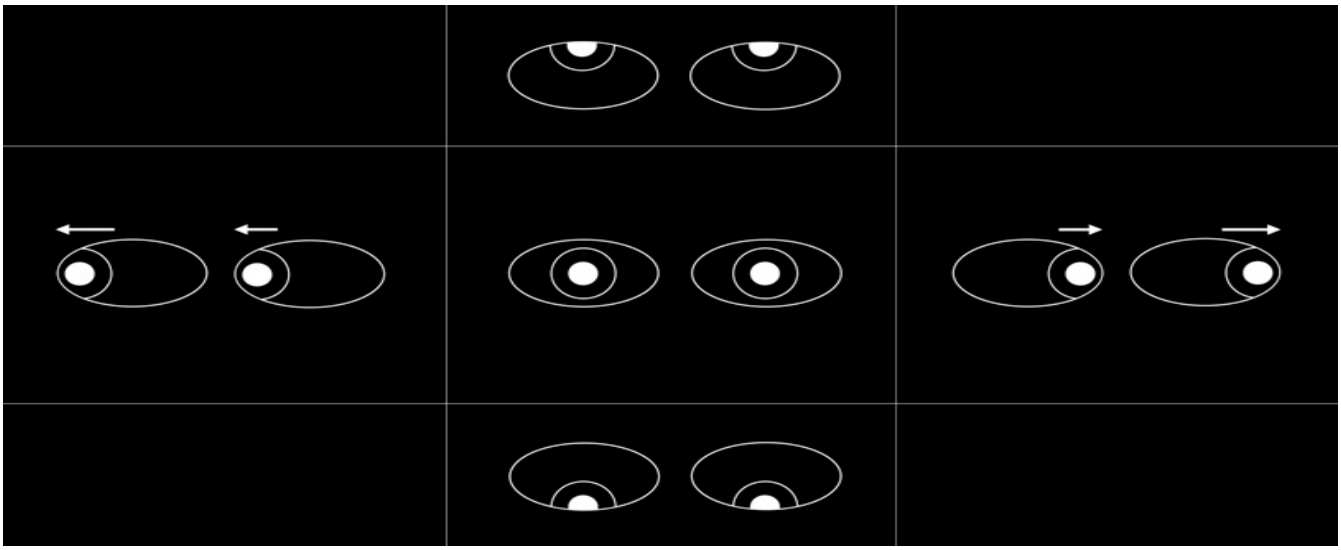

Figure 5.1: *Physiologic nystagmus.*

PATHOLOGICAL NYSTAGMUS

Pathological nystagmus is a slow drift with rapid correction. It is rarely the main clinical sign. It is usually associated with vestibular or cerebellar signs: slurred speech, staccato speech, and unsteady gait, vertigo, incoordination. Isolated nystagmus presents with blurred vision and oscillopsia (jumping images in the vertical or horizontal directions).

Ophthalmoscopy can detect subtle nystagmus. The direction of nystagmus seen on the disc is opposite to movement of the fundus. The common causes are drugs such as tegretol and benzodiazapines and alcohol. With cerebellar disease, the nystagmus is ipsilateral, coarse and slow. With peripheral disease, the nystagmus is contralateral, fine, fast and mixed torsional/horizontal.

APPROACH TO NYSTAGMUS

- Binocular or monocular?
- Binocular symmetric or dissociated? Dissociated = one eye affected more than the other
- Conjugate or disconjugate? Conjugate = in same direction, disconjugate = opposite directions
- Spontaneous in primary position?
- Gaze-evoked in eccentric position?
- In all positions of gaze? Null point? Vary in different gaze positions? Null point = point of least nystagmus and best visual acuity as in congenital nystagmus
- Fast and slow phase – Jerk nystagmus?
- Equal speed in both directions – Pendular nystagmus?

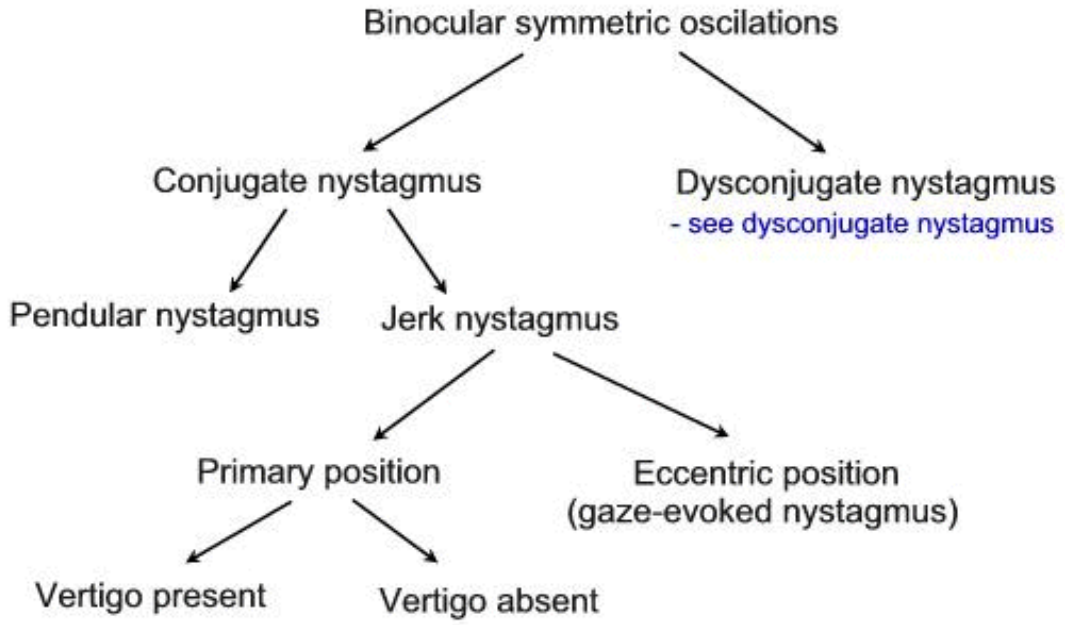

Figure 5.2: *Binocular symmetric oscillations*

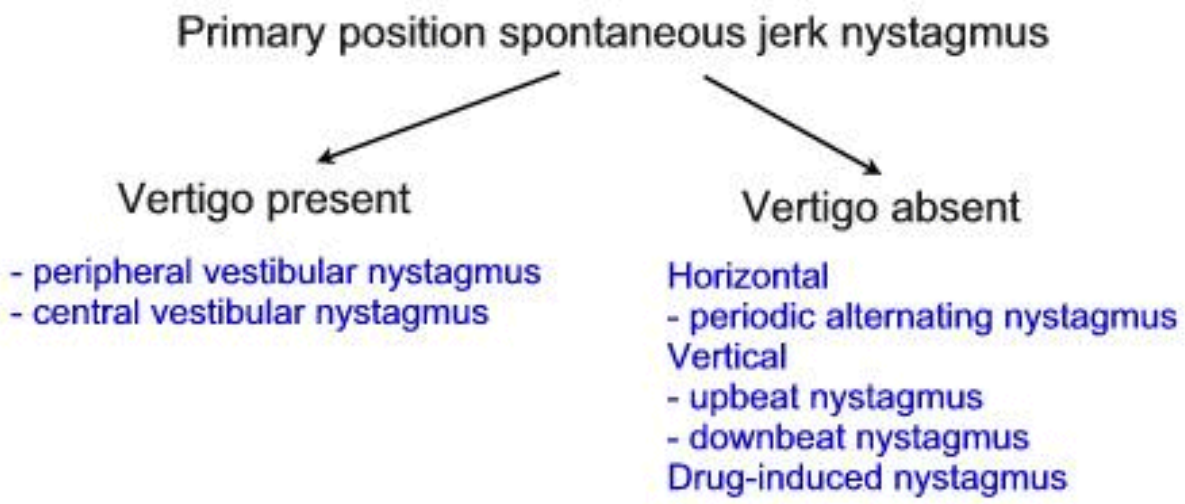

Figure 5.3*: Jerk nystagmus*

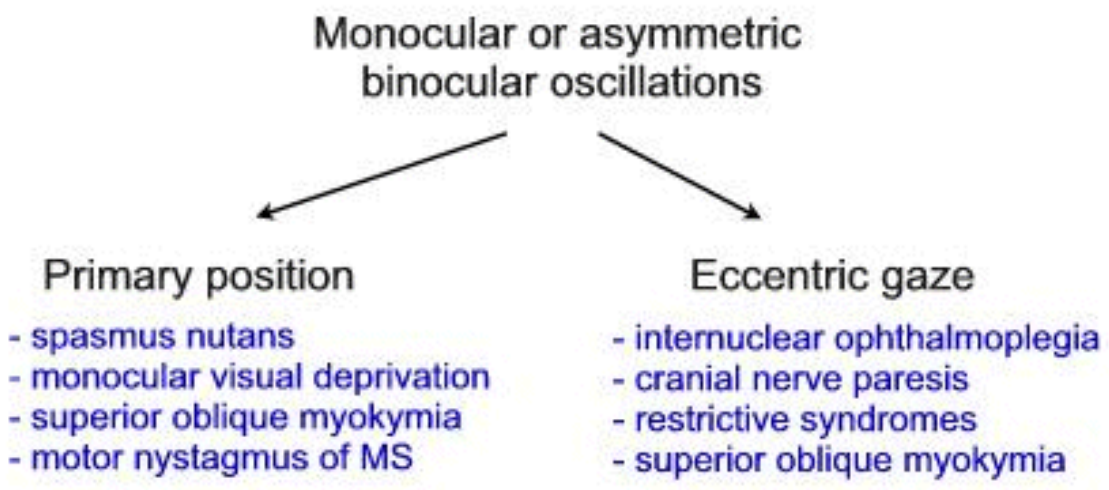

Figure 5.4: *Monocular or asymmetric binocular oscillations*

DISCONJUGATE NYSTAGMUS

- Seesaw nystagmus
- Convergence retraction nystagmus
- Convergence nystagmus (Whipple's Disease)

EYE MOVEMENT DISORDERS

- Ocular Dysmetria
- Ocular Flutter
- Opsoclonus
- Ocular Bobbing
- Ocular Dipping/Inverse bobbing
- Reverse ocular bobbing
- Converse bobbing/reverse ocular dipping
- Ping Pong Gaze
- Square wave jerks
- Macro square wave jerks
- Oculomasticatory Myorhythmia

OCULAR DYSMETRIA

Ocular dysmetria is a side-to-side oscillation of the eyes seen as they return to the primary position from lateral gaze. It lasts for a few moments before stabilisation. It is usually accompanied by extremity, gait, or speech ataxia. It reflects cerebellar system dysfunction. Eye movements do not cause visual symptoms. Brain imaging may show lesions of brainstem or cerebellum. It is a symptom of several neurological conditions including multiple sclerosis. It is a condition that can cause symptoms similar to sea sickness.

Differential Diagnosis:

- Voluntary nystagmus
- Congenital nystagmus
- Ocular flutter or opsoclonus

OCULAR FLUTTER

Ocular flutter is characterized as having brief bursts of oscillation of the eyes in the neutral position (Figure 5.5). This is an involuntary rapid occurrence when attempting to fixate on an object. In ocular flutter, the trajectory is strictly in the horizontal plane. It is caused by dysfunction of pontine pause cells that normally prevent involuntary conjugate eye movements.

It is associated with:

- brief muscle contractions (myoclonus) at joints
- head and trunk oscillations (titubation)
- extremity tremor
- ataxia

Common causes in children include: postviral autoimmune disorder or paraneoplastic disorder associated with metastatic neuroblastoma.

Common causes in adults include: postviral or paraneoplastic encephalitic manifestation of metastatic lung, breast, or ovarian cancer. Treatment of the underlying cancer may improve neurological symptoms.

Figure 5.5: *Ocular flutter. Involuntary rapid saccades move the eyes conjugately off target. As the movements are limited to the horizontal plane, this is called ocular flutter.*

OPSOCLONUS

The eyes move conjugately, spontaneously and continuously oscillate in a variety of directions in opsoclonus (Figure 5.6). This is an involuntary movement of the eyes that persists during sleep. Opsoclonus indicates brainstem, mesencephalic or pontine, or cerebellar disease. It may occur as a paraneoplastic disorder in children who have neuroblastoma and in adults who have occult malignancy. It may also occur as a postinfectious condition accompanied by ataxia and myoclonus.

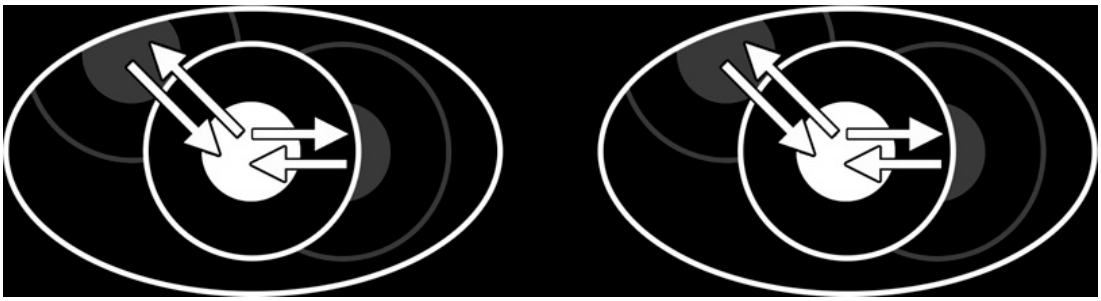

Figure 5.6: *Opsoclonus. The conjugate saccadic eye movements occur in horizontal and vertical planes.*

OCULAR BOBBING

Ocular bobbing is a rapid downward eye movement with a slow return to the primary position (Figure 5.7). It is associated with pontine and other structural, metabolic or toxic disorders.

Spontaneous eye movements seen in comatose patients include the following:

TYPICAL OCULAR BOBBING

- fast downward movement followed by slow upward drift to primary position
- horizontal eye movements are absent

OCULAR DIPPING

- slow downward movement followed by fast upward movement
- horizontal eye movements are often intact
- It may occur in cases of hypoxic ischaemic insult, metabolic disorders

REVERSE BOBBING

- There is a fast upward movement followed by slow downward drift to primary position
- The horizontal eye movements often intact
- It may occur in patients with metabolic encephalopathy, viral encephalitis, or pontine haemorrhage.

REVERSE DIPPING

- There is a slow upward drift followed by a fast downward movement
- The horizontal eye movements are often intact
- It is associated with pontine infarction and with metabolic or viral encephalopathy and AIDS

Occasionally ocular bobbing, ocular dipping, and reverse bobbing may occur at different times in the same patient.

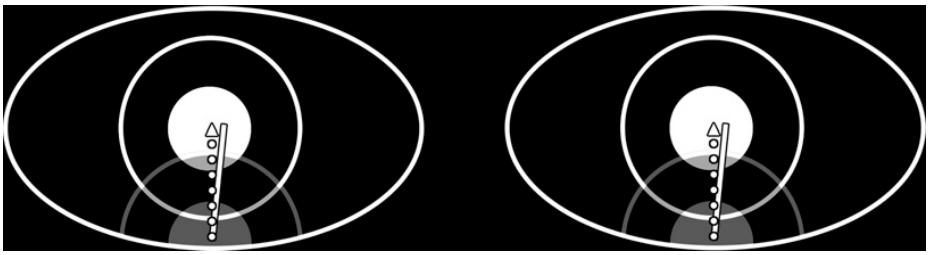

Figure 5.7: *Ocular bobbing. The eyes involuntarily move downward rapidly (solid lines) and drift slowly up to primary position (dotted lines).*

Inspired by: Duker JS, Macsai MS, Trobe JD. Rapid Diagnoses in Ophthalmology: Neuro-ophthalmology. 2008

PING PONG GAZE

The ping pong gaze is characterized by rapid horizontal conjugate deviation of the eyes alternating every few seconds. This is usually seen in patients with bilateral cerebral hemisphere dysfunction.

SQUARE WAVE JERKS

These are small saccades away from fixation (0.5 – 5 ^{0}) and back with a 200 msec interval (Figure 5.8). It is often mistaken for nystagmus, but this eye movement abnormality is actually a saccadic intrusion on fixation. It is a non-specific sign of brainstem dysfunction. It produces no symptoms. They may also occur with lithium or tobacco use. Square wave jerks may occur in patients with cerebellar disease or progressive supranuclear palsy (PSP).

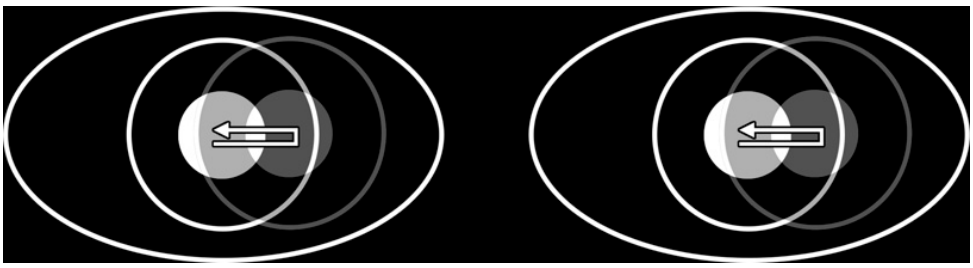

Figure 5.8*: Square wave jerks.*

Inspired by: Duker JS, Macsai MS, Trobe JD. Rapid Diagnoses in Ophthalmology: Neuro-ophthalmology. 2008

MACRO SQUARE WAVE JERKS

This is characterized by saccadic intrusions (5 – 15 ^{0}) and return within 70-150 ms. They are occasionally present in the vertical plane. They are usually conjugate, horizontal, and symmetric in both directions of gaze, but may occur in torsional or vertical planes. They may occur in patients with multiple sclerosis and olivopontocerebellar atrophy (OPCA).

OCULOMASTICATORY MYORHYTHMIA:

This refers to acquired pendular vergence oscillations associated with concurrent contraction of the masticatory muscles. There is a smooth, rhythmic eye convergence, followed by divergence back to the primary position. If nonfacial skeletal muscles are involved, it is called oculofacial-skeletal myorhythmia. Rhythmic elevation and depression of the mandible is synchronous with the ocular oscillations that persist in sleep and are unaltered by stimuli. Patients with oculomasticatory myorhythmia may also have paralysis of vertical gaze, progressive somnolence, and intellectual deterioration. This distinct movement disorder has been recognized only in Whipple's disease (class III–IV, level B).