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Ear Click

- see PALATAL MYOCLONUS; TINNITUS

Eastchester Clapping Sign

The Eastchester clapping sign is advocated as an early and sensitive sign of hemispatial ego-centric neglect. Patients are asked to clap: those with neglect perform one handed motions which stop at the midline. Hemiplegic patients without neglect reach across the midline and clap against their plegic hand.

Reference

Ostrow LW, Llinas RH. Eastchester clapping sign: a novel test of parietal neglect. *Ann Neurol*. 2009; **66**: 114–7.

Cross Reference

Neglect

Echolalia

Echolalia is the involuntary repetition of an interviewer's speech utterances (as opposed to the voluntary mimicry-taking which characterises an irritating game typical of childhood, but sometimes indulged in by adults). This may be observed in a variety of clinical situations:

- Transcortical sensory aphasia:
In the context of a fluent aphasia with repetition often well or normally preserved, usually as a result of a vascular lesion of the left hemisphere although an analogous situation may be encountered in Alzheimer's disease; "incorporational echolalia", when the patient uses the examiner's question to help form an answer, may be observed as a feature of "dynamic aphasia" which bears resemblance to transcortical motor aphasia, but may result from a frontal lesion; this has also been reported in progressive supranuclear palsy.
- Transcortical motor aphasia:
"Effortful echolalia" has been reported in the context of infarction of the left medial frontal lobe, including the supplementary motor area, showing that neither the ability to repeat nor fluent speech is required for echolalia.
- Tourette syndrome:
As a complex vocal tic, along with coprolalia.
- Alzheimer's disease, frontotemporal lobar degeneration, Creutzfeldt-Jakob disease:
As a symptom of cognitive impairment/dementia.
- Epilepsy:
From a left frontal lobe (supplementary motor area) focus.
- Schizophrenia:
As a catatonic symptom.
- Early infantile autism, mental retardation:
As a reflection of pathological mental development.

- Frontal lobe lesions:
As a feature of imitation behaviour.
- Normal children:
At a particular stage of language acquisition.

References

Hadano K, Nakamura H, Hamanaka T. Effortful echolalia. *Cortex*. 1998; **34**: 67–82.
Mendez MF. Prominent echolalia from isolation of the speech area. *J Neuropsychiatry Clin Neurosci*. 2002; **14**: 356–7.

Cross References

Aphasia; Coprolalia; Dynamic aphasia; Imitation behaviour; Jargon aphasia; Logorrhoea; Palilalia; Transcortical aphasias

Echolocation

More usually associated with bats and dolphins, humans can also echolocate, either passively (e.g. by listening for echoes bouncing off furniture) or actively by making tongue clicks and listening for echoes, a skill which allows some blind individuals to undertake activities such as mountain biking. Compensatory enhancement (cross-modal plasticity) underlies this faculty.

Reference

Thaler L, Arnott SR, Goodale MA. Neural correlates of natural human echolocation in early and late blind echolocation experts. *PLoS One*. 2011; **6**: e20162.

Echophenomena

A number of echophenomena are described in the neurological literature: echolalia, echopraxia, echolocation. The term echophenomena has sometimes been used interchangeably with imitation behaviour.

Reference

Larner AJ. Neurological signs: echo phenomena. *Adv Clin Neurosci Rehabil*. 2015; **15**(3): 16.

Cross Reference

Imitation behaviour

Echopraxia

Echopraxia is the involuntary, automatic, imitation of an interviewer's movements. This may be observed as a feature of apraxic syndromes such as corticobasal degeneration, as a complex motor tic in Tourette syndrome, and in frontal lobe disorders (imitation behaviour), and rarely as an ictal phenomenon.

Reference

Pridmore S, Brüne M, Ahmadi J, Dale J. Echopraxia in schizophrenia: possible mechanisms. *Aust N Z J Psychiatry*. 2008; **42**: 565–71.

Cross References

Copropraxia; Imitation behaviour; Tic

Écriture En Double Miroir

- see MIRROR WRITING

Ectropion

- see LID RETRACTION

Eidetic Memory

Eidetic, or “photographic”, memory is an enhancement of memory function to prodigious capacity, beyond hypermnnesia. Synaesthesia may be linked to eidetic memory, synaesthesia being used as a mnemonic aid.

Reference

Luria AR. The mind of a mnemonist. New York: Basic Books; 1968.

Cross Reference

Synaesthesia

Eight-And-A-Half Syndrome

The combination of a facial (VII) nerve palsy with a one-and-a-half syndrome due to a pontine lesion has been labelled the eight-and-a-half syndrome. Recognised causes include infarction and inflammation (e.g. multiple sclerosis). Patients may develop oculopalatal myoclonus months to years after the onset of the ocular motility problem.

References

Eggenberger EJ. Eight-and-a-half syndrome: One-and-a-half syndrome plus cranial nerve VII palsy. *J Neuroophthalmol.* 1998; **18**: 114–6.

Nandhagopal R, Krishnamoorthy SG. Eight-and-a-half syndrome. *J Neurol Neurosurg Psychiatry.* 2006; **77**: 463.

Wolin MJ, Trent RG, Lavin PJM, Cornblath WT. Oculopalatal myoclonus after the one-and-a-half syndrome with facial nerve palsy. *Ophthalmol.* 1996; **103**: 177–80.

Cross References

Facial paresis, Facial weakness; Myoclonus; One-and-a-half syndrome; Palatal myoclonus

Ekbom's Syndrome

Ekbom's syndrome or delusional parasitosis (not to be confused with Willis-Ekbom syndrome or disease, better known as restless legs syndrome) is a condition in which patients believe with absolute certainty that insects, maggots, lice or other vermin infest their skin or other parts of the body. Sometimes other psychiatric features may be present, particularly if the delusions are part of a psychotic illness such as schizophrenia or depressive psychosis. Females are said to be more commonly affected. Clinical examination may sometimes show evidence of skin picking, scratching, or dermatitis caused by repeated use of antiseptics. The patient may produce skin fragments or other debris as "evidence" of infestation. Treatment should be aimed at the underlying condition if appropriate; if the delusion is isolated, anti-psychotics such as pimozide may be tried.

References

Enoch MD, Ball HN. Uncommon psychiatric syndromes. 4th ed. London: Arnold; 2001. p. 209–23.

Karroum E, Konofal E, Arnulf I, Karl-Axel Ekbom (1907-1977). *J Neurol.* 2009; **256**: 683–4.

Lombardi C, Belli D, Passalacqua G. When allergology meets psychiatry: delusional parasitosis (Ekbom's syndrome). *Eur Ann Allergy Clin Immunol.* 2011; **43**: 89–91.

Cross Reference

Delusion

Emotionalism, Emotional Lability

Emotionalism, or emotional lability, or emotional incontinence, implies both frequent and unpredictable changes in emotional expression, for example tearfulness followed shortly by elation, and an inappropriate expression of emotion, for example uncontrollable ("uninhibited" or disinhibited) laughter or crying. A distinction may be drawn between the occurrence of these phenomena spontaneously or without motivation, or in situations which although funny or sad are not particularly so. Also, a distinction may be made between such phenomena when there is congruence of mood and affect, sometimes labelled with terms such as *moria* or *witzelsucht* (e.g. laughing when feeling happy or elated), and when there is no such congruence (e.g. laughing when not feeling happy or elated), sometimes labelled as pathological, forced, or inappropriate laughter and crying.

The neurobehavioural state of emotional lability reflects frontal lobe (especially orbito-frontal) lesions, often vascular in origin, and may coexist with disinhibited behaviour. It is more common in vascular dementia than Alzheimer's disease. It may also be seen in delirium

and in psychiatric disorders (mania). Pathological laughter and crying may occur as one component of pseudobulbar palsy (“pseudobulbar affect”).

Reference

Heilman KM, Blonder LX, Bowers D, Valenstein E. Emotional disorders associated with neurological diseases. In: Heilman KM, Valenstein E, editors. *Clinical neuropsychology*. 4th ed. Oxford: Oxford University Press; 2003. p. 447–78.

Cross References

Delirium; Disinhibition; Frontal lobe syndromes; Moria; Pathological crying, Pathological laughter; Pseudobulbar palsy; *Witzelsucht*

Emprosthotonos

Emprosthotonos is an abnormal posture consisting of flexion of the head on the trunk and the trunk on the knees, sometimes with flexion of the limbs (*cf.* opisthotonos). Such attacks of “bowing” may be seen in infantile epilepsy syndromes such as West’s syndrome, sometimes called salaam seizures or jack-knife spasms. Describing the tonic spasms of tetanus, Gowers noted that “Opisthotonic spasm is the rule, to which the exceptions are few. Rarely the trunk is bent forwards, from predominant cramp in the abdominal muscles and other flexors of the spine – “emprosthotonos”. Still more rarely there is slight lateral flexion, “pleurothotonos”, or the trunk and neck are rigid in a straight line, “orthotonos”.”

Reference

Gowers WR. *Manual of diseases of the nervous system*, vol 2, 2nd ed. London: J&A Churchill; 1893. p. 682.

Cross References

Opisthotonos; Seizures; Spasm

Encephalopathy

Encephalopathy is a general term referring to any acute or chronic diffuse disturbance of brain function (literally “sick brain”). Characteristically it is used to describe an altered level of consciousness, which may range from drowsiness to a failure of selective attention, to hypervigilance; with or without: disordered perception, memory (*i.e.* cognitive deficits); epileptic seizures; headache; abnormal movements such as tremor, myoclonus, or asterixis; and focal neurological deficits (less common). Clearly these features overlap with those of delirium, sometimes itself denoted as “toxic-metabolic encephalopathy”.

As with terms such as coma and stupor, it is probably better to give a description of the patient’s clinical state rather than use a term that is open to variable interpretation.

Although the term encephalopathy is sometimes reserved for metabolic causes of diffuse brain dysfunction, this usage is not universal. Conditions which may be described as an encephalopathy include:

- Metabolic disorders: hypoxia/ischaemia, hypoglycaemia; organ failure, electrolyte disturbances, hypertension.
- Drug/toxin ingestion.
- Brain inflammation/infection (*e.g.* encephalitis).
- Miscellaneous conditions, *e.g.* Alzheimer’s disease, Creutzfeldt-Jakob disease.

Cross References

Asterixis; Coma; Delirium; Myoclonus; Stupor; Tremor

En Garde Position

- see FENCER’S POSTURE, FENCING POSTURE

Enhanced Ptosis

Enhanced ptosis describes the increased drooping (*i.e.* worsening) of a ptotic eyelid when the contralateral eyelid is manually lifted. Enhanced ptosis may be explained by the action of Hering’s law of equal innervation to paired yoked muscles (*i.e.* the eyelids). Recognised causes

of enhanced ptosis include myasthenia gravis, Miller Fisher syndrome, botulism, and Lambert Eaton myasthenic syndrome

Reference

Gorelick PB, Rosenberg M, Pagano RJ. Enhanced ptosis in myasthenia gravis. *Arch Neurol.* 1981; **38**: 531.

Cross References

Curtainng; Ptosis

Enophthalmos

Enophthalmos is an inward displacement of the eyeball (sinking or withdrawal) into the eye socket (*cf.* exophthalmos). It is classically described as one of the cardinal features of Horner's syndrome (along with miosis, ptosis, and anhidrosis) but is seldom actually measured. Enophthalmos may also occur in dehydration (probably the most common cause), orbital trauma (*e.g.* orbital floor fracture), senile orbital fat atrophy, hemifacial atrophy (Parry-Romberg syndrome), and orbital tumour causing tethering and posterior traction on the eyeball.

Cross References

Anhidrosis; Exophthalmos; Horner's syndrome; Miosis; Parry-Romberg syndrome; Ptosis

Entomopia

Entomopia (literally "insect eye") is the name given to a grid-like pattern of multiple copies of the same visual image; hence, this is a type of polyopia. This phenomenon has been reported in migraine; its pathogenesis is uncertain.

References

Larner AJ. Entomopia. *Adv Clin Neurosci Rehabil.* 2006; **6**(4): 30.

Lopez JR, Adornato BT, Hoyt WF. "Entomopia": a remarkable case of cerebral polyopia. *Neurology.* 1993; **43**: 2145–6.

Cross Reference

Polyopia

Entrainment Test

An entrainment test has been advocated for the identification of psychogenic tremor, in which the examiner asks the patient to tap another limb at a different frequency to the tremor. Entrainment of tapping and tremor suggests that the latter is psychogenic.

Reference

Roper LS, Saifee TA, Parees I, Rickards H, Edwards MJ. How to use the entrainment test in the diagnosis of functional tremor. *Pract Neurol.* 2013; **13**: 396–8.

Cross Reference

Tremor

Environmental Dependency Syndrome

- see IMITATION BEHAVIOUR; UTILIZATION BEHAVIOUR

Environmental Tilt

Environmental tilt, also known as tortopia, is the sensation that visual space is tilted on its side or even upside down ("floor-on-ceiling" phenomenon, "upside-down" reversal of vision, *verkehrtsehen*). This may last seconds to minutes. The temptation to dismiss such bizarre symptoms as functional should be resisted, since environmental tilt is presumed to reflect damage to connections between cerebellar and central vestibular-otolith pathways causing cortical mismatch of visual and vestibular three-dimensional coordinate maps. It has been reported in the following situations:

- Posterior circulation ischaemia: lateral medullary syndrome of Wallenberg, transient ischaemic attacks in basilar artery territory.
- Demyelinating disease.

- Head injury.
- Encephalitis.
- Following third ventriculostomy for hydrocephalus.

Reference

Sierra-Hildago F, de Pablo-Fernandez E, Herrero-San Martin A et al. Clinical and imaging features of the room tilt illusion. *J Neurol.* 2012; **259**: 2555–64.

Cross References

Lateral medullary syndrome; Vertigo; Vestibulo-ocular reflexes

Epiphora

Epiphora is overflow of tears down the cheek. This may be due to a blocked nasolacrimal duct, or irritation to the cornea causing increased lacrimation, but it may also be neurological in origin, *e.g.* due to the sagging of the lower eyelid (ectropion) in a peripheral facial (VII) nerve (Bell's) palsy, or the "crocodile tears" following aberrant facial nerve regeneration. Lacrimation is also a feature of trigeminal autonomic cephalalgias such as cluster headache.

Cross References

Bell's palsy; Crocodile tears

Epley Manoeuvre

- see HALLPIKE MANEOUVRE, HALLPIKE TEST; VERTIGO

Erythropsia

This name has been given to a temporary distortion of colour vision in which objects take on an abnormal reddish hue. This has been characterized as a visual illusion. There are various causes, including drug use, visual diseases, and pseudophakia.

Cross References

Illusion; "Monochromatopsia"; Phantom chromatopsia

Esophoria

Esophoria is a variety of heterophoria in which there is a tendency for the visual axes to deviate inward (latent convergent strabismus). Clinically this may be observed using the cover-uncover test as an outward movement of the covered eye as it is uncovered. Esophoria may occur in individuals with hyperopia (long-sightedness).

Cross References

Cover tests; Exophoria; Heterophoria

Esotropia

Esotropia is a variety of heterotropia in which there is manifest inward turning of the visual axis of one eye; the term is synonymous with convergent strabismus. It may be demonstrated using the cover test as an outward movement of the eye which is forced to assume fixation by occlusion of the other eye.

Esotropia may be associated with congenital latent nystagmus (*i.e.* nystagmus appearing when one eye is covered) in the presence of amblyopia; the slow phase in the viewing eye is towards the nose.

With lateral rectus muscle paralysis, the eyes are esotropic or crossed on attempted lateral gaze towards the paralysed side, but the images are uncrossed. Acute esotropia has been described following contralateral thalamic infarction.

Cross References

Amblyopia; Cover tests; Diplopia; Exotropia; Heterotropia; Nystagmus

Eutonia

Kinnier Wilson used this term to describe an emotional lack of concern associated with the dementia of multiple sclerosis. It may perhaps reflect the cognitive anosognosia of a dementia syndrome.

Ewart Phenomenon

This is the elevation of ptotic eyelid on swallowing, a synkinetic movement. The mechanism is said to be aberrant regeneration of fibres from the facial (VII) nerve to the oculomotor (III) nerve innervating the levator palpebrae superioris muscle.

Cross References

Ptosis; Synkinesia, Synkinesis

Exophoria

Exophoria is a variety of heterophoria in which there is a tendency for the visual axes to deviate outward (latent divergent strabismus). Clinically this may be observed in the cover-uncover test as an inward movement as the covered eye is uncovered. Exophoria may occur in individuals with myopia, and may be physiological in many subjects because of the alignment of the orbits.

Cross References

Cover tests; Esophoria; Heterophoria

Exophthalmos

Exophthalmos is forward displacement of the eyeball. The definition and the causes overlap with proptosis. The most common cause is dysthyroid eye disease (Graves' disease).

Cross References

Lid retraction; Proptosis

Exosomaesthesia

The sensory disturbance associated with parietal lobe lesions may occasionally lead the patient to refer the source of a stimulus to some point outside the body, exosomaesthesia. A possible example occurs in Charles Dickens's novel *Hard Times* (1854) in which Mrs Gradgrind locates her pain as "somewhere in the room".

Exotropia

Exotropia is a variety of heterotropia in which there is manifest outward turning of the visual axis of an eye; the term is synonymous with divergent strabismus. It may be demonstrated using the cover test as an inward movement of the eye which is forced to assume fixation by occlusion of the other eye.

When the medial rectus muscle is paralysed, the eyes are exotropic (wall-eyed) on attempted lateral gaze towards the paralysed side, and the images are crossed.

Cross References

Cover tests; Esotropia; Heterotropia

Extensor Posturing

- see DECEREBRATE RIGIDITY

External Malleolar Sign

- see CHADDOCK'S SIGN

External Ophthalmoplegia

- see OPHTHALMOPARESIS, OPHTHALMOPLEGIA

Extinction

The failure to sense a stimulus on one side when stimulation is simultaneously presented on both sides was described by Morris Bender as extinction to simultaneous stimulation. Extinction is the failure to respond to a novel or meaningful sensory stimulus on one side when a homologous stimulus is given simultaneously to the contralateral side (*i.e.* double simultaneous stimulation); it is sometimes called "suppression". The stimuli may be visual, auditory, or tactile, *e.g.* asking the patient to say which hand is touched when the eyes are shut. It is important to show that the patient responds appropriately to each hand being

touched individually, but then neglects one side when both are touched simultaneously. With repeated testing the phenomenon may break down: extinguishing of extinction.

More subtle defects may be tested using simultaneous bilateral heterologous (asymmetrical) stimuli, although it has been shown that some normal individuals may show extinction in this situation.

A motor form of extinction has been postulated, manifesting as increased limb akinesia when the contralateral limb is used simultaneously (hemiakinesia).

The presence of extinction is one of the behavioural manifestations of neglect, and most usually follows non-dominant (right) hemisphere (parietal lobe) lesions. There is evidence for physiological interhemispheric rivalry or competition in detecting visual stimuli from both hemifields, which may account for the emergence of extinction following brain injury.

Reference

Fink GR, Driver J, Rorden C, Baldeweg T, Dolan RJ. Neural consequences of competing stimuli in both visual hemifields: a physiological basis for visual extinction. *Ann Neurol*. 2000; 47: 440–6.

Cross References

Akinesia; Hemiakinesia; Neglect; Visual extinction

Extrapyramidal Signs

- see PARKINSONISM

Eyelid Apraxia

Eyelid apraxia is an inability to open the eyelids at will, although they may open spontaneously at other times (*i.e.* there is voluntary-automatic dissociation). Eyelids may be opened manually or by a backwards head thrust. The term has been criticised on the grounds that this may not always be a true “apraxia”, in which case the term “levator inhibition” may be preferred since the open eyelid position is normally maintained by tonic activity of the levator palpebrae superioris. Clinically there is no visible contraction of orbicularis oculi, which distinguishes eyelid apraxia from blepharospasm (however, perhaps paradoxically, the majority of cases of eyelid apraxia occur in association with blepharospasm). Neurophysiological studies do in fact show abnormal muscle contraction in the pre-tarsal portion of orbicularis oculi, which has prompted the suggestion that “focal eyelid dystonia” may be a more appropriate term. Although the phenomenon may occur in isolation, associations have been reported with:

- Progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome).
- Parkinson's disease.
- Huntington's disease.
- Multiple system atrophy.
- MPTP intoxication.
- Motor neurone disease.
- Acute phase of nondominant hemisphere cerebrovascular event.
- Wilson's disease.
- Neuroacanthocytosis.

The precise neuroanatomical substrate is unknown but the association with basal ganglia disorders points to involvement of this region. The underlying mechanisms may be heterogeneous, including involuntary inhibition of levator palpebrae superioris. Botulinum toxin injections may be helpful in some patients.

References

Kanazawa M, Shimohata T, Sato M, Onodera O, Tanaka K, Nishizawa M. Botulinum toxin A injections improve apraxia of eyelid opening without overt blepharospasm associated with neurodegenerative diseases. *Mov Disord.* 2008; **22**: 597–8.

Kerty E, Eidal K. Apraxia of eyelid opening: clinical features and therapy. *Eur J Ophthalmol.* 2006; **16**: 204–8.

Cross References

Apraxia; Blepharospasm; Dystonia

Eyelid Retraction

- see LID RETRACTION