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## **Saccades**

Saccades are rapid, ballistic, yoked movements of the eyes which bring the gaze to a new location in visual space. These movements may be performed voluntarily (tested clinically by asking the patient to “Look to your left, keeping your head still,” *etc.*) or reflexively, *i.e.*, in response to an object of potential interest within the visual field (tested clinically by asking the patient to shift gaze from one of examiner’s hands to another). Internuclear ophthalmoplegia may be revealed when testing saccadic eye movements.

A number of parameters may be observed, including latency of saccade onset, saccadic amplitude, and saccadic velocity. An antisaccadic task (*i.e.*, suppression of saccades to a novel visual stimulus) may be used to assess ease of saccade suppression. Of these, saccadic velocity is the most important in terms of localization value, since it depends on burst neurones in the brainstem (paramedian pontine reticular formation for horizontal saccades, rostral interstitial nucleus of the medial longitudinal fasciculus for vertical saccades). Latency involves cortical and basal ganglia circuits; antisaccades involve frontal lobe structures; and amplitude involves basal ganglia and cerebellar circuits (saccadic hypometria, with a subsequent correctional saccade, may be seen in extrapyramidal disorders, such as Parkinson’s disease; saccadic hypermetria or overshoot may be seen in cerebellar disorders). Difficulty in initiating saccades may be described as ocular (motor) apraxia. Antisaccades may be poorly suppressed in Huntington’s disease. In Alzheimer’s disease, patients may make reflex saccades toward a target in an antisaccadic task (visual grasp reflex).

Assessment of saccadic velocity may be of particular diagnostic use in parkinsonian syndromes. In progressive supranuclear palsy slowing of vertical saccades is an early sign (suggesting brainstem involvement; horizontal saccades may be affected later), whereas vertical saccades are affected late (if at all) in corticobasal degeneration, in which condition increased saccade latency is the more typical finding, perhaps reflective of cortical involvement.

## **References**

Carpenter R. The saccadic system: a neurological microcosm. *Advances in Clinical Neuroscience & Rehabilitation* 2004; **4(1)**: 6-8  
Leigh RJ, Riley DE. Eye movements in parkinsonism: it’s saccadic speed that counts. *Neurology* 2000; **54**: 1018-1019

## **Cross References**

Internuclear ophthalmoplegia; Ocular apraxia; Ocular flutter; Opsoclonus; Parkinsonism; Saccadic intrusion, Saccadic pursuit; Square-wave jerks

**Saccadic Intrusion, Saccadic Pursuit**

Saccadic intrusions are inappropriate saccades which interfere with visual fixation (static, or during motor pursuit: saccadic pursuit). Several types of saccadic intrusion are described, including ocular flutter, opsoclonus, and square-wave jerks. Saccadic (cogwheel) pursuit is normal in infants and may be a nonspecific finding in adults; however, it may be seen in Huntington's disease.

**Cross References**

Ocular flutter; Opsoclonus; Saccades; Square-wave jerks

**Saccadomania**

- see OPSOCLONUS

**Sacral Sparing**

Sacral sparing is the preservation of pain and temperature sensation in sacral dermatomes. This is a late, unusual, but diagnostic feature of an intrinsic (intramedullary) spinal cord lesion. Spastic paraparesis below the level of the lesion due to corticospinal tract involvement is invariably present by this stage.

Sacral sparing is explained by the lamination of fibers within the spinothalamic tract: ventrolateral fibers (of sacral origin), the most external fibers, are involved later than the dorsomedial fibers (of cervical and thoracic origin) by an expanding central intramedullary lesion (*e.g.*, glioma, ependymoma, syringomyelia).

Although sacral sparing is rare, sacral sensation should always be checked in any patient with a spastic paraparesis.

**Cross References**

Dissociated sensory loss; Myelopathy; Paraparesis

**Saddle Anesthesia**

- see ANESTHESIA; CAUDA EQUINA SYNDROME

**Saturday Night Palsy**

- see WRIST DROP

**Scanning Speech**

Scanning speech is a motor speech disorder (*i.e.*, a dysarthria) comprising slow, deliberate, dysprosodic, monotonic verbal output. It may be confused with nonfluent aphasia (Broca's aphasia).

Scanning speech was originally considered a feature of cerebellar disease in multiple sclerosis (after Charcot), and the term is often used with this implication. However, cerebellar disease typically produces an ataxic dysarthria (variable intonation, interruption between syllables, "explosive" speech) which is somewhat different to scanning speech. Scanning speech correlates with midbrain lesions, often after recovery from prolonged coma.

**Cross References**

Asynergia; Aphasia; Broca's aphasia; Cerebellar syndromes; Dysarthria

**Scapula Alata**

- see WINGING OF THE SCAPULA

**Schizophasia**

This term has been used to describe the language disorder in schizophrenia, which may be characterized by paraphasias and neologisms, loose connections between thoughts, tangential thinking, and delusional intrusions. The resulting output may be unintelligible and may resemble Wernicke's aphasia.

**Cross References**

Delusion; Neologism; Paraphasia; Wernicke's aphasia

**Schwabach Test**

A vibrating tuning fork is held against the mastoid process, as in Rinne's test, until it is no longer audible to the patient. The examiner then places the tuning fork over his/her own mastoid, hence comparing bone conduction with that of the patient. If still audible to the examiner (presumed to have normal hearing), a sensorineural hearing loss is suspected, whereas in conductive hearing loss the test is normal.

**Cross References**

Rinne's test

**Scoliosis**

- see KYPHOSCOLIOSIS

**Scotoma**

A scotoma is a localized area of impaired vision within an otherwise normal visual field. Mapping of the defect may be manual, by confrontation testing, or automated. In addition to the peripheral field, the central field should also be tested, with the target object moved around the fixation point. A central scotoma may be picked up in this way, or a more complex defect, such as a centrocecal scotoma in which both the macula and the blind spot are involved. Infarction of the occipital pole will produce a central visual loss, as will optic nerve inflammation. Scotomata may be absolute (no perception of form or light) or relative (preservation of form, loss of color).

A scotoma may be physiological, as in the blind spot or angioscotoma, or pathological, reflecting disease anywhere along the visual pathway from retina and choroid to visual cortex.

Various types of scotoma may be detected (see individual entries for more details):

Central scotoma

Cecocentral or centrocecal scotoma

Arcuate scotoma

Annular or ring scotoma

Junctional scotoma

Junctional scotoma of Traquair

Peripapillary scotoma (enlarged blind spot)

**Cross References**

Altitudinal field defect; Angioscotoma; Blind spot; Central scotoma, Centrocecal scotoma; Hemianopia; Junctional scotoma, Junctional scotoma of traquair; Maculopathy; Papilledema; Quadrantanopia; Retinitis pigmentosa; Retinopathy; Visual field defects

**“Scratch Test”**

The “scratch test,” or “direction of scratch” test, examines perception of the direction (up or down) of a scratch applied to the anterior shin (for example, with the sharp margin of a paper clip). It has been claimed as a reliable test of posterior column function of the spinal cord. Errors in this test correlate with central conduction times and vibration perception threshold.

**References**

Hankey GJ, Edis R. The utility of testing tactile perception of direction of scratch as a sensitive clinical sign of posterior column dysfunction in spinal cord disorders. *Journal of Neurology, Neurosurgery and Psychiatry* 1989; **52**: 395-398

Motoi Y, Matsumoto H, Kaneshige Y, Chiba S. A reappraisal of “direction of scratch” test: using somatosensory evoked potentials and vibration perception. *Journal of Neurology, Neurosurgery and Psychiatry* 1992; **55**: 509-510

**Cross References**

Proprioception; Vibration

**Seborrhea**

Seborrhea is a greasiness of the skin which may occur in extrapyramidal disorders, particularly Parkinson’s disease.

**Cross References**

Parkinsonism

**Seelenblindheit**

- see VISUAL AGNOSIA

**Seizures**

Seizures are sudden, paroxysmal episodes of neurological dysfunction with or without impairment of consciousness, which may be epileptic (*i.e.*, due to abnormal synchronous electrical activity within the brain, either focally or generally) or nonepileptic in origin (“pseudoseizures,” nonepileptic attack disorder). The two varieties may coexist. Seizure morphology may be helpful in establishing etiology and/or focus of onset.

- *Epileptic:*

Idiopathic generalized: tonic-clonic (“grand mal”); absence attack (“petit mal”); myoclonic epilepsy

Partial: simple (no impairment of consciousness), for example jerking of one arm, which may spread sequentially to other body parts (jacksonian march); or complex, in which

there is impairment or loss of consciousness: may be associated with specific aura (olfactory, *déjà vu*, *jamais vu*) and/or automatisms (motor, *e.g.*, cursive; or emotional, *e.g.*, gelastic, dacrystic); limb posturing (salutatory, fencing posture) and pelvic thrusting may be seen in frontal lobe epilepsy. Secondary generalization of seizures of partial onset may occur.

Investigation of partial seizures to exclude a symptomatic cause is recommended (MR imaging, EEG). Some are amenable to surgical intervention. Otherwise, as for idiopathic generalized epilepsies, various antiepileptic medications are available. Partial seizures may prove more resistant to treatment than generalized seizures.

- **Nonepileptic:**

Often long lasting, thrashing, pelvic thrusting, carpet burns, may have incontinence; past history of physical or sexual abuse. Best treated with psychological approaches, or drug treatment of underlying affective disorders; antiepileptic medications are best avoided.

The differentiation of epileptic from nonepileptic seizures may be difficult; it is sometimes helpful to see a video recording of the attacks, or to undertake in-patient video-telemetry.

### References

Duncan JS, Shorvon SD, Fish DR. *Clinical epilepsy*. New York: Churchill Livingstone 1995: 25-101

Lüders H, Acharya J, Baumgartner C *et al*. Semiological seizure classification. *Epilepsia* 1998; **39**: 1006-1013

Panayiotopoulos CP. *A clinical guide to epileptic syndromes and their treatment: based on the new ILAE diagnostic scheme*. Chipping Norton: Bladon, 2002

### Cross References

Absence; Aura; Automatism; Déjà vu; Fencer's posture, Fencing posture; Incontinence; Jacksonian march; Jamais vu; Pelvic thrusting

### Self-Mutilation

Self-injury to the point of mutilation, especially around the mouth, may be seen in certain neurological conditions, such as Lesch-Nyhan syndrome, Gilles de la Tourette syndrome, and neuroacanthocytosis.

### Sensory Ataxia

- see ATAXIA; ROMBERGISM, ROMBERG'S SIGN

### Sequential Paresis

Sequential, or "round the clock," paresis or weakness refers to the sequential development of weakness in one arm, the ipsilateral leg, the contralateral leg, and contralateral arm (*i.e.*, hemiparesis, triparesis, tetra- or quadriparesis). This pattern is highly suggestive of a foramen magnum lesion, usually a tumor but sometimes demyelination or other

intrinsic inflammatory disorder, sequentially affecting the lamination of corticospinal fibers in the medullary pyramids.

**Cross References**

Hemiparesis; Paresis; Quadriparesis, Quadriplegia

**Setting Sun Sign**

The setting sun sign, or sunset sign, consists of tonic downward deviation of the eyes with retraction of the upper eyelids exposing the sclera. There may be downbeating nystagmus. Setting sun sign is a sign of dorsal midbrain compression in children with untreated hydrocephalus.

A similar appearance may also be observed in progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome; Stellwag's sign) and in Parinaud's syndrome, but without the tonic downward deviation.

**Cross References**

Lid retraction; Parinaud's syndrome; Stellwag's sign

**Shadowing**

A neurobehavioral disorder, occasionally seen in patients with dementia, in which the patient follows the spouse or caregiver around like a shadow.

**Cross References**

Dementia

**Shin-Tapping**

A modification of the heel-knee-shin test or heel-shin test in which the heel is tapped repetitively on the shin before sliding it down to the foot, claimed to be a better test of motor coordination.

**References**

Fisher CM. An improved test of motor coordination in the lower limbs. *Neurology* 1961; **11**: 335-336

**Cross References**

Ataxia; Cerebellar syndromes; Heel-knee-shin test, Heel-shin test

**Sialorrhea**

Sialorrhea (drooling) is excessive salivation, possibly due to excess flow of saliva but more likely secondary to a reduced frequency of swallowing (e.g., in parkinsonian syndromes) or difficulty swallowing (e.g., motor neurone disease, developmental perisylvian syndrome).

Metallic poisonings (mercury, bismuth, lead) may also produce marked salivation (ptyalism).

If troublesome, treatment of sialorrhea with anticholinergic agents may be tried (atropine, hyoscine), although they may cause confusion in Parkinson's disease. In extreme cases, irradiation of the salivary glands has been used. Recently, the use of intraparotid injections of botulinum toxin has been found useful.

**References**

Bhatia KP, Münchau A, Brown P. Botulinum toxin is a useful treatment in excessive drooling of saliva. *Journal of Neurology, Neurosurgery and Psychiatry* 1999; **67**: 697

**Cross References**

Bulbar palsy; Parkinsonism

**Sighing**

Occasional deep involuntary sighs may occur in multiple system atrophy. Sighing is also a feature, along with yawning, of the early (diencephalic) stage of central herniation of the brainstem with an otherwise normal respiratory pattern. Sudden inspiratory or expiratory sighs are said to be a feature of the hyperkinetic choreiform dysarthria characteristically seen in choreiform disorders, such as Huntington's disease.

**References**

Quinn N. Multiple system atrophy. In: Marsden CD, Fahn S (eds.). *Movement disorders 3*. Boston: Butterworth, 1994: 262-281

**Cross References**

Blinking; Dysarthria; Yawning

***Signe de l'Eventail (Fan Sign)***

- see BABINSKI'S SIGN (1)

***Signe de Rideau***

*Signe de rideau*, or curtain sign, refers to the motion of the posterior pharyngeal wall toward the intact side, resembling the drawing of a curtain, in unilateral paresis of the superior pharyngeal constrictor muscle, as seen in unilateral vagus (X) nerve palsy.

***Signe du Journal***

- see FROMENT'S SIGN

**Simian Hand**

Simian hand or ape hand has been used to describe the atrophy of the thenar eminence with recession of the metacarpal bones of the thumb to the plane of the other metacarpal bones seen in median nerve lesions in the axilla or upper arm.

**Cross References**

Benediction hand

**Simian Posture**

- see PARKINSONISM

**Simultanagnosia**

Simultanagnosia is impaired perception of multi-element or multipart visual displays, such that pictures are described in a piecemeal manner. Recognition of single objects is preserved; this is likened to having a fragment or island of clear vision which may shift from region to region.

Two types of simultanagnosia are described:

- *Dorsal:*  
An attentional limitation preventing more than one object being seen at a time; although superficially similar to apperceptive visual agnosia, with which it has sometimes been classified, patients with dorsal simultanagnosia can recognize objects quickly and accurately, but unattended objects are not seen. There may be inability to localize stimuli even when they are seen, manifest as visual disorientation. Reading is severely impaired. Patients may grope, as though blind. Dorsal simultanagnosia is associated with bilateral posterior parieto-occipital lesions, and is one feature of Balint's syndrome.
- *Ventral:*  
A limitation in the number of objects which can be recognized in unit time, *i.e.*, there is no primary recognition problem in that individual shapes can be recognized. Ventral simultanagnosia is most evident during reading which is severely impaired and empirically this may be the same impairment as seen in pure alexia; otherwise deficits may not be evident, unlike dorsal simultanagnosia. Ventral simultanagnosia may be a form of associative visual agnosia. It is associated with left inferior temporo-occipital cortical lesions.

### References

Coslett HB, Saffran E. Simultanagnosia: to see but not two see. *Brain* 1991; **114**: 1523-1545

Farah MJ. *Visual agnosia: disorders of object recognition and what they tell us about normal vision*. Cambridge: MIT Press, 1995

### Cross References

Agnosia; Alexia; Balint's syndrome; Visual agnosia; Visual disorientation

### Singultus

- see HICCUPS

### “Sink Sign”

- see ROMBERG'S SIGN, ROMBERGISM

### Skew Deviation

Skew deviation, or the Hertwig-Magendie sign, is a supranuclear vertical misalignment of the visual axes; the final common efferent pathway for eye movements is spared (*cf.* hypertropia, hypotropia, due to ocular motor nerve palsies or extraocular muscle disease). This is thought to reflect damage to otolith-ocular pathways or vestibulo-ocular pathways. There may be concurrent ocular tilt reaction. Bielschowsky's head tilt test is usually negative (*cf.* ocular motor nerve palsies).

Skew deviation has been associated with posterior fossa lesions, from midbrain to medulla. Ipsiversive skew deviation (ipsilateral eye lowermost) has been associated with caudal pontomedullary lesions,



whereas contraversive skew (contralateral eye lowermost) occurs with rostral pontomesencephalic lesions, indicating that skew type has localizing value.

#### References

Brandt Th, Dieterich M. Skew deviation with ocular torsion: a vestibular brainstem sign of topographic diagnostic value. *Annals of Neurology* 1993; **33**: 528-534

#### Cross References

Bielschowsky's sign, Bielschowsky's test; Hypertropia; Hypotropia; Ocular tilt reaction; Tullio phenomenon

### Smile-Wink Phenomenon

This name has been given to narrowing of the palpebral fissure aggravated by smiling following a contralateral lenticulocapsular infarction. Dysarthria, facial paresis, hemiparesis with or without hemihypoesthesia, and excessive laughing with or without crying, were common accompanying features.

#### References

Kim JS. Smile-wink phenomenon: aggravated narrowing of palpebral fissure by smiling after lenticulocapsular stroke. *Journal of Neurology* 2001; **248**: 389-393

#### Cross References

Dysarthria; Facial paresis; Hemiparesis; Hypoesthesia

### Snarling Facies

- see "MYASTHENIC SNARL"

### Sneezing

Loss of the ability to sneeze has been recorded following lateral medullary syndrome. Sneezing (or ptarmus) as a herald of lateral medullary syndrome may reflect a cause of vertebral artery dissection rather than a primary irritative lesion.

#### References

Hersch M. Loss of ability to sneeze in lateral medullary syndrome. *Neurology* 2000; **54**: 520-521

Bernat JL, Suranyi L. Loss of ability to sneeze in lateral medullary syndrome. *Neurology* 2000; **55**: 604

#### Cross References

Lateral medullary syndrome

### Snoring

Reduced muscle tone in the upper airway during sleep leads to increased resistance to the flow of air, and partial obstruction often results in loud snoring. This symptom may be associated with the obstructive sleep apnea-hypopnea syndrome (OSAHS), which may be associated with a variety of neurological symptoms including excessive daytime somnolence, episodic loss of consciousness, headache (especially morning), cognitive decline, and

increased risk of stroke (snoring may be an independent risk factor for stroke).

### References

Huang L, Quinn SJ, Ellis PDM, Ffowes Williams JE. Biomechanics of snoring. *Endeavour* 1995; **19**: 96-100

Larner AJ. Obstructive sleep apnea syndrome presenting in a neurology outpatient clinic. *International Journal of Clinical Practice* 2003; **57**: 150-152

Spriggs DA, French JM, Murdy JM, Curless RH, Bates D, James OFW. Snoring increases the risk of stroke and adversely affects prognosis. *Quarterly Journal of Medicine* 1992; **84**: 555-562

### Cross References

Hypersomnolence

### Snout Reflex

Sometimes used interchangeably with pout reflex, this term should probably be reserved for the puckering or pouting of the lips induced by constant pressure over the philtrum, rather than the phasic response to a tap over the muscle with finger or tendon hammer.

### Cross References

Frontal release signs; Pout reflex; Primitive reflexes

### Somatoparaphrenia

Ascription of hemiplegic limb(s) to another person (*e.g.*, the examiner, a family member); possibly a confabulation.

### Cross References

Anosognosia; Autotopagnosia; Confabulation

### Somatotopagnosia

- see AUTOTOPAGNOSIA

### Spasm

The word spasm implies a sudden, involuntary, muscle contraction, which may be painful (cramp). For example, flexor spasms in patients paraplegic due to upper motor neurone lesions are sudden contractions of the flexor musculature, particularly of the legs, either spontaneous or triggered by light touch. Hemifacial spasm is an involuntary contraction of facial musculature.

Spasm may also refer to a tetanic muscle contraction (tetany), as seen in hypocalcemic states (*e.g.*, *main d'accoucheur*), tetanus (*e.g.*, risus sardonicus), or tonic spasms of various muscles (*e.g.*, jaw musculature, trismus) which may be dystonic or spastic in origin. Involuntary movements, such as tics, may be known as spasms or habit spasms.

Patients may use the word spasm differently, *e.g.*, to denote paroxysmal sensory phenomena, or even seizures. Infantile seizures consisting of brief flexion of the trunk and limbs (emprostotonos, salaam or jack-knife seizures) may be known as spasms.

**References**

Rowland LP. Cramps, spasms, and muscle stiffness. *Revue Neurologique (Paris)* 1985; **141**; 261-273

**Cross References**

Contracture; Dystonia; Hemifacial spasm; Main d'accoucheur; Paraplegia; Risus sardonicus; Seizures; Tic; Trismus

**Spasmus Nutans**

Spasmus nutans is the clinical triad of head nodding, anomalous head postures, and nystagmoid eye movements seen in children aged between 1 and 8 years. This is usually a benign condition, but the diagnosis should prompt consideration of an optic pathway tumor.

**Cross References**

Nystagmus

**Spastic Catch**

- see SPASTICITY

**Spasticity**

Spasticity is an increased resistance to the passive movement of a joint due to abnormally high muscle tone (hypertonus) which varies with the amplitude and speed of displacement of a joint (*cf.* rigidity). The excessive resistance evident at the extremes of joint displacement may suddenly give way, a phenomenon known as clasp-knife (or, confusingly, clasp-knife rigidity). Spasticity may vary in degree from mild, (*e.g.*, a spastic catch on supination/pronation of the forearm), to extreme (*e.g.*, immobile limbs in fixed flexion with secondary contractures and painful spasms: paraplegia in flexion). Spasticity may need to be differentiated clinically from rigidity and stiffness.

The amount and pattern of spasticity depends on the location of the lesion and tends to be greater with spinal cord than cortical lesions. Scales to quantitate spasticity are available (Ashworth, modified Ashworth, Wartenberg pendulum test) but have shortcomings. Spasticity may also vary in distribution: for lesions above the spinal cord it typically affects the arm flexors and the leg extensors to a greater extent (hemiparetic posture).

Spasticity is a clinical feature of the upper motor neurone syndrome, and may be accompanied by both positive (clonus, hyperreflexia, Babinski's sign, flexor or extensor spasms) and negative phenomena (weakness in a pyramidal distribution, motor underactivity): the latter may be more significant determinants of disability. Slow, labored speech, with slow voluntary tongue movements, may be referred to as spastic dysarthria, which may occur in the context of a pseudobulbar palsy.

The pathogenesis of spasticity has traditionally been ascribed to damage to the corticospinal and/or corticobulbar pathways at any level from cerebral cortex to spinal cord. However, various lines of evidence (*e.g.*, the failure of pyramidotomy to produce spasticity in animals, rare human cases of isolated pyramid infarction causing

hyperreflexia and weakness without spasticity) has led to the implication of other motor tracts in the genesis of spasticity, viz.:

- The dorsal reticulospinal tract, which lies in the lateral funiculus of the spinal cord and hence is often damaged concurrently with the adjacent lateral corticospinal tract (e.g., in MS, which seems to have a predilection for the lateral funiculus); this descending pathway has an inhibitory effect on stretch reflexes which is under cortical control;
- The medial reticulospinal tract and vestibulospinal tracts which are not under cortical control and whose excitatory effects on extensor tone may remain unopposed.

Physiologically, spasticity has been characterized as an exaggeration of the muscle stretch reflexes, with reduced threshold (hyperexcitable  $\alpha$ -motor neurones) and abnormal reflex transmission (increased gain). The role of neurotransmitters (glutamate, glycine, catecholamines, serotonin) in the pathogenesis of spasticity is unclear, but the efficacy of baclofen (a GABA<sub>B</sub> agonist) and benzodiazepines suggest impaired GABAergic transmission may contribute, perhaps through a loss of presynaptic inhibition mediated by interneurons or the inhibition of glutamate release.

Treatment of severe spasticity, for example in multiple sclerosis, often requires a multidisciplinary approach. Urinary infection, constipation, skin ulceration and pain can all exacerbate spasticity, as may inappropriate posture; appropriate management of these features may ameliorate spasticity. Drugs which may be useful include baclofen, dantrolene (a blocker of muscle excitation-contraction coupling), and tizanidine ( $\alpha_2$ -adrenoreceptor agonist). Intrathecal baclofen given via a pump may also be of benefit in selected cases, and for focal spasticity injections of botulinum toxin may be appropriate. For painful immobile spastic legs with reflex spasms and double incontinence, irreversible nerve injury with intrathecal phenol or alcohol may be advocated to relieve symptoms. The place of cannabinoids has yet to be fully determined.

### References

Shakespeare DT, Boggild M, Young C. Anti-spasticity agents for multiple sclerosis. *Cochrane Database of Systematic Reviews* 2003; **4**: CD001332  
 Sheean G. Neurophysiology of spasticity. In: Barnes MP, Johnson GR (eds.). *Upper motor neurone syndrome and spasticity. Clinical management and neurophysiology*. Cambridge: CUP, 2001: 12-78

### Cross References

Babinski's sign (1); Clasp-knife phenomenon; Clonus; Contracture; Dysarthria; Hyperreflexia; Hypertonus; Paraplegia; Pseudobulbar palsy; Reflexes; Spasm; Upper motor neurone (UMN) syndrome; Weakness

### Speech Apraxia

Speech apraxia is one of the labels applied to a disorder of communication characterized by slow speech tempo ("groping for words"), impaired articulation, and dysprosody, with relatively intact language function and no dysgraphia. More errors occur with increasing articu-

latory complexity (consonant clusters *vs.* single consonants). Automatic or reactive speech (*e.g.*, expletives, clichés) is without error. This, or a very similar, constellation of features has also been known as cortical dysarthria, aphemia, or phonetic disintegration. There may be associated orofacial apraxia.

Speech apraxia has been associated with inferior frontal dominant (left) hemisphere damage in the region of the lower motor cortex or frontal operculum; it has been claimed that involvement of the anterior insula is specific for speech apraxia.

The exact nosological status of this entity remains in some doubt. The syndrome is thought to reflect disturbances of planning articulatory and phonatory functions, but is most often encountered as part of a nonfluent aphasia.

#### References

Dronkers NF. A new brain region for coordinating speech articulation. *Nature* 1996; **384**: 159-161

#### Cross References

Aphasia; Aphemia; Apraxia

#### Spinal Mass Reflex

The spinal mass reflex is involuntary flexion of the trunk in a comatose patient, such that they appear to be attempting to sit up.

#### Cross References

Coma

#### Spurling's Sign

This is the name given to increase in arm pain (brachialgia) associated with compressive cervical radiculopathy following neck rotation and flexion to the side of the pain. A variant of this foraminal compression test involves rotation, side bend and slight extension of the neck with the application of axial pressure to the head.

#### Cross References

Radiculopathy

#### Square-Wave Jerks

Square-wave jerks are small saccades which interrupt fixation, moving the eye away from the primary position and then returning. This instability of ocular fixation is a disorder of saccadic eye movements in which there is a saccadic interval (of about 200 ms; *cf.* ocular flutter, opsoclonus). Very frequent square-wave jerks may be termed square-wave oscillations. Very obvious square-wave jerks (amplitude > 7°) are termed macro-square-wave jerks.

Square-wave jerks are often best appreciated on ophthalmoscopy. Their name derives from the appearance they produce on electro-oculographic recordings.

Although square-wave jerks may be normal in elderly individuals, they may be indicative of disease of the cerebellum or brainstem, *e.g.*, Huntington's disease, Parkinson's disease, progressive supranuclear palsy, cerebellar degeneration.

**Cross References**

Nystagmus; Ocular flutter; Opsoclonus; Saccadic intrusion, Saccadic pursuit

**Square-Wave Oscillations**

- see SQUARE-WAVE JERKS

**Squint**

- see HETEROTROPIA

**Stapedius Reflex**

- see HYPERACUSIS

**Stellwag's Sign**

Stellwag's sign is a widening of the palpebral fissure due to upper eyelid retraction. Along with a reduced blink rate, this creates a very typical staring, "astonished," facies. The clinical phenomena of Stellwag's sign overlap with those labeled as the sunset sign.

Stellwag's sign is seen in progressive supranuclear palsy, and in dysthyroid eye disease.

**Cross References**

Blinking; Lid lag; Lid retraction; Setting sun sign

**Steppage, Stepping Gait**

Steppage or stepping gait occurs with a lower motor neurone type of foot drop ("floppy" foot drop), *e.g.*, due to a common peroneal nerve palsy, peripheral neuropathies. Because of the weakness of foot dorsiflexion (weak tibialis anterior) there is compensatory overaction of hip and knee flexors during the swing phase of walking to ensure the foot clears the ground. In the strike phase, there is a characteristic slapping down of the foot, again a consequence of weak ankle dorsiflexion. Proprioceptive loss, as in dorsal column spinal disease, may also lead to a gait characterized by high lifting of the feet, and also stomping (stamping with a heavily accented rhythm) or slapping of the foot onto the floor in the strike phase.

The pattern of gait with upper motor neurone foot drop ("stiff" foot drop), *e.g.*, due to a corticospinal tract lesion, is quite different, with the foot being dragged, sometimes with circumduction of the leg. This may lead to falls as a consequence of tripping over the foot, especially on up-hill gradients, and a characteristic pattern of wear on the point of the shoe.

**Cross References**

Foot drop; Lower motor neurone (LMN) syndrome; Proprioception; Rombergism, Romberg's sign; Upper motor neurone (UMN) syndrome

**Stereoanesthesia**

- see ASTEREOGNOSIS

**Stereohypesthesia**

- see ASTEREOGNOSIS

**Stereotypy**

Stereotypies may be defined as regular repeated movements, which are voluntary but not apparently goal-directed, and which may be carried out in a uniform pattern for long periods of time (*cf.* tic). Whole areas of the body may be involved by stereotypies and hence this movement is more complex than a tic.

Stereotypies are common in patients with learning disability and schizophrenia. Very characteristic manual stereotypies (washing, rubbing movements: “hand washing”) may be seen in Rett’s disease. The term has also been used to describe movements associated with chronic neuroleptic use; indeed adult-onset stereotypy is highly suggestive of prior exposure to dopamine receptor blocking drugs.

Verbal stereotypies are reiterated words or syllables produced by patients with profound nonfluent aphasia (*e.g.*, Broca’s original case, Leborgne, who could only repeat “tan, tan, tan,” by which name he was known).

**References**

Jankovic J. Stereotypies. In: Marsden CD, Fahn S (eds.). *Movement disorders 3*. Boston: Butterworth, 1994: 503-517

Lees AJ. *Tics and related disorders*. Edinburgh: Churchill Livingstone, 1985

Stacy M, Cardoso F, Jankovic J. Tardive stereotypy and other movement disorders in tardive dyskinesia. *Neurology* 1993; **43**: 937-941

**Cross References**

Aphasia; Broca’s aphasia; Tic

**Sternocleidomastoid Test**

It has been reported that apparent weakness of the sternocleidomastoid muscle is common (80%) in functional hemiparesis, usually ipsilateral to the hemiparesis, whereas it is rare in vascular hemiparesis (11%), presumably because of the bilateral innervation of the muscle.

**References**

Diukova GM, Stolajrova AV, Vein AM. Sternocleidomastoid (SCM) muscle test in patients with hysterical and organic paresis. *Journal of the Neurological Sciences* 2001; **187** (suppl1): S108

**Cross References**

Functional weakness and sensory disturbance; Hemiparesis

**Stethoscope Loudness Imbalance Test**

- see HYPERACUSIS

**Stewart-Holmes Sign**

- see REBOUND PHENOMENON

**Stiffness**

Stiffness of muscles occurs as a feature of all pyramidal and extrapyramidal disorders (as spasticity and rigidity, respectively), but the term stiffness is usually reserved for disorders in which stiffness is the principal symptom due to continuous motor unit activity within muscles. There may be associated muscle pain (cramp). Stiffness may be primarily of muscular origin (myotonia) or of neural origin (myokymia, neuromyotonia). Accompanying signs may prove helpful in diagnosis, such as slow muscle relaxation (myotonia), percussion irritability of muscle (myoedema), and spontaneous and exertional muscle spasms. Hyperlordotic posture is typical of stiff man/stiff person syndrome. Stiffness must be differentiated from both rigidity and spasticity.

Recognized causes of stiffness include:

Stiff man/stiff person syndrome

Stiff limb syndrome

Progressive encephalomyelitis with rigidity +/-myoclonus

Neuromyotonia (Isaac's syndrome; armadillo syndrome)

Schwartz-Jampel syndrome (chondrodystrophic myotonia)

Tetanus

Strychnine poisoning

The stiff man/stiff person syndrome is probably of autoimmune pathogenesis since it is strongly associated with insulin-dependent diabetes mellitus and the presence of antibodies to glutamic acid decarboxylase (anti-GAD antibodies), the enzyme in the synthetic pathway of GABA. Intravenous immunoglobulin therapy may be of symptomatic benefit.

**References**

Barker R, Revesz T, Thom M, Marsden CD, Brown P. Review of 23 patients affected by the stiff man syndrome: clinical subdivision into stiff trunk (man) syndrome, stiff limb syndrome, and progressive encephalomyelitis with rigidity. *Journal of Neurology, Neurosurgery and Psychiatry* 1998; **65**: 633-640

Scolding NJ. Stiff man syndrome. In: Scolding NJ (ed.). *Immunological and inflammatory disorders of the central nervous system*. Oxford: Butterworth Heinemann 1999: 139-146

Thompson PD. Stiff people. In: Marsden CD, Fahn S (eds.). *Movement disorders 3*. Boston: Butterworth, 1994: 373-405

**Cross References**

Myokymia; Myotonia; Neuromyotonia; Paramyotonia; Rigidity; Spasticity

**“Stork Legs”**

A name given to describe the disproportionate wasting of the lower legs, a pattern characteristic of hereditary motor and sensory neuropathies (Charcot-Marie-Tooth diseases), which may be evident even before the development of gait disorder with foot drop and steppage gait.

**Cross References**

Foot drop; Steppage, Stepping gait; Wasting



**Strabismus**

- see HETEROPHORIA; HETEROTROPIA

**Straight Leg Raising**

- see LASÈGUE'S SIGN

**“Straight Thumb Sign”**

Median nerve lesions in the forearm cause weakness of flexor pollicis longus, which normally flexes the distal phalanx of the thumb. Hence the thumb remains straight when the patient attempts to grasp something or make a fist. The “pinch sign” may also be present.

**References**

Cherington M. Anterior interosseous nerve syndrome straight thumb sign. *Neurology* 1977; **27**: 800-801

**Cross References**

“Pinch sign”

**Striatal Toe**

Striatal toe refers to the tonic extension of the hallux which is seen in dystonic syndromes, and as a feature of extrapyramidal disorders.

Striatal toe may be confused with Babinski's sign (extensor plantar response), the principal difference being that the latter is elicited by stimulation whereas the former is a tonic response.

**References**

Winkler AS, Reuter I, Harwood G, Chaudhuri KR. The frequency and significance of “striatal toe” in parkinsonism. *Parkinsonism and Related Disorders* 2002; **9**: 97-101

**Cross References**

Babinski's sign (1); Parkinsonism

**Stupor**

Stupor is a state of altered consciousness characterized by deep sleep or unresponsiveness, susceptible to arousal only by vigorous and/or repeated stimuli, with lapse back into unresponsiveness when the stimulus stops. Stupor is a less severe impairment of conscious level than coma, but worse than obtundation (torpor). It is suggestive of diffuse cerebral dysfunction, *e.g.*, drug-induced.

**References**

Plum F, Posner JB. *The diagnosis of stupor and coma* (3rd edition). Philadelphia: FA Davis, 1980

**Cross References**

Coma; Delirium; Encephalopathy; Obtundation

**Stutter**

Stutter, one of the reiterative speech disorders, is usually a developmental problem, but may be acquired in aphasia with unilateral or bilateral hemisphere lesions (*e.g.*, vascular damage, trauma, Alzheimer's disease, Parkinson's disease, progressive supranuclear palsy). Unlike developmental stutter, acquired stutter may be evident

throughout sentences, rather than just at the beginning. Furthermore, developmental stutter tends to occur more with plosives (phonemes where the flow of air is temporarily blocked and suddenly released, as in 'p', 'b'), whereas acquired stutter is said to affect all speech sounds fairly equally. Cessation of developmental stutter following bilateral thalamic infarction in adult life has been reported.

#### References

Fleet WS, Heilman KM. Acquired stuttering from a right hemisphere lesion in a right-hander. *Neurology* 1985; **35**: 1343-1346

Muroi A, Hirayama K, Tanno Y, Shimizu S, Watanabe T, Yamamoto T. Cessation of stuttering after bilateral thalamic infarction. *Neurology* 1999; **53**: 890-891

#### Cross References

Aphasia; Echolalia; Palilalia

### Sucking Reflex

Contact of an object with the lips will evoke sucking movements in an infant. The reflex may re-emerge in dementia.

#### Cross References

Akinetic mutism; Dementia; Frontal release signs

### Summerskill's Sign

- see LID RETRACTION

### "Sundowning"

"Sundowning," or sundown syndrome, is increased confusion, agitation or disorientation in the late afternoon, evening, and night-time, which may be seen in patients with delirium, and sometimes in dementia. In dementia, there may be complete reversal of sleep schedule with daytime somnolence and nocturnal wakefulness. Although this syndrome may relate to worsening of visual cues with increasing darkness, it may also occur in well-lit environments. A disorder of circadian rhythms is a possible physiological correlate of "sundowning": EEG recordings in delirious patients may suggest this. Suggested management for dementia patients with sundowning includes use of structured activities at the relevant times, and increased staffing or availability of family members. Sedative medications are probably best avoided.

#### References

Evans LK. Sundown syndrome in institutionalized elderly. *Journal of the American Geriatrics Society* 1987; **35**: 101-108

Volicer L, Harper DG, Manning BC, Goldstein R, Satlin A. Sundowning and circadian rhythms in Alzheimer's disease. *American Journal of Psychiatry* 2001; **158**: 704-711

#### Cross References

Delirium; Dementia

### Sunset Sign

- see SETTING SUN SIGN

**Suppression**

- see EXTINCTION

**Supranuclear Gaze Palsy**

A supranuclear gaze palsy results from pathology located above the nuclei of the nerves supplying the extraocular muscles. Voluntary gaze is impaired while the integrity of the oculomotor nuclei and infranuclear connections may be demonstrated by the preservation of:

Vestibulo-ocular reflexes (VOR): overcoming the ophthalmoplegia, at least in the early stages (*e.g.*, the supranuclear gaze palsy in the vertical plane in progressive supranuclear palsy);  
Oculocephalic reflex (doll's head, doll's eye maneuver);  
Bell's phenomenon.

Supranuclear gaze palsies may be:

- *Horizontal:*
  - Hemisphere (frontal) lesion: eyes deviated to the side of the lesion, or in the case of an irritative (*e.g.*, epileptic) focus away from the side of the lesion
  - Paramedian pontine reticular formation: eyes deviated to contralateral side.
- *Vertical:*
  - Brainstem compression/distortion
  - Dorsal upper midbrain (*e.g.*, rostral interstitial nucleus of the median longitudinal fasciculus; pineal lesion causing Parinaud's syndrome)
  - Skew deviation

Recognized causes of supranuclear gaze palsy include:

Progressive supranuclear palsy (PSP; Steele-Richardson-Olszewski syndrome)  
Creutzfeldt-Jakob disease  
Corticobasal degeneration  
Progressive subcortical gliosis of Neumann  
Adult-onset Niemann-Pick disease  
Gaucher's disease.

**References**

Lees AJ. The Steele-Richardson-Olszewski syndrome (progressive supranuclear palsy). In: Marsden CD, Fahn S (eds.). *Movement Disorders 2*. London: Butterworth, 1987: 272-287

**Cross References**

Gaze palsy; Parinaud's syndrome; Parkinsonism; Prevost's sign; Skew deviation; Vestibulo-ocular reflexes

**Suspended Sensory Loss**

Sensory loss or impairment involving the trunk and proximal limbs may be described as suspended, or in a "cape-like," "bathing suit," "vest-like," or cuirasse distribution. This may reflect intrinsic or

intramedullary spinal cord pathology (in which case other signs of myelopathy may be present, including dissociated sensory loss), but can also occur in peripheral neuropathic disease, such as acute porphyria.

#### **Cross References**

Dissociated sensory loss; Myelopathy

#### **“Swan Neck”**

This term has been applied to thinning of the neck musculature, as in myotonic dystrophy for example.

#### **Swinging Flashlight Sign**

The swinging flashlight sign or test, originally described by Levitan in 1959, compares the direct and consensual pupillary light reflexes in one eye; the speed of swing is found by trial and error. Normally the responses are equal but in the presence of an afferent conduction defect an inequality is manifest as pupillary dilatation. The test is known to be unreliable in the presence of bilateral afferent defects of light conduction. Subjective appreciation of light intensity, or light brightness comparison, is a subjective version of this test.

#### **References**

Thompson HS, Corbett JJ. Swinging flashlight test. *Neurology* 1989; **39**: 154-156

#### **Cross References**

Marcus gunn pupil, Marcus gunn sign; Relative afferent pupillary defect (RAPD)

#### **Synesthesia**

Synesthesia is a perceptual experience in one sensory modality following stimulation of another sensory modality. The most commonly encountered example is color-word synesthesia (“colored hearing” or chromesthesia), experiencing a visual color sensation on hearing a particular word. Synesthesia occurs in a small percentage of the normal population. Known synesthetes include the composers Messiaen and Scriabin, the artist Kandinsky, and the author Nabokov. There may be concurrent excellent memory (hypermnnesia), sometimes of a photographic nature (eidetic memory). Symptomatic synesthesia is rare but has been described with epileptic seizures of temporal lobe origin and with drug use (LSD).

Neuropsychologically this phenomenon has been conceptualized as a break down of modularity. Functional imaging studies of color-word synesthetes show activation of visual associative areas of cortex (but not primary visual cortex), as well as perisylvian language areas, when listening to words which evoke the experience of color.

#### **References**

Baron-Cohen S, Harrison JE (eds.). *Synaesthesia: classic and contemporary readings*. Oxford: Blackwell, 1997

Cytowic RE. *The man who tasted shapes*. Cambridge: MIT Press, 2003

Paulesu E, Harrison J, Baron-Cohen S, *et al.* The physiology of colored hearing: a PET activation study of color-word synaesthesia. *Brain* 1995; **118**: 661-676

#### **Cross References**

Auditory-visual synesthesia; Phosphene

#### **Synkinesia, Synkinesis**

The term synkinesis may be used in different ways. It may refer to involuntary movements which accompany or are associated with certain voluntary movements (*mitbewegungen*, motor overflow). These may be physiological, for example the swinging of the arms when walking. Alternatively, such associated phenomena may be pathological, *e.g.*, the involuntary contraction of orbicularis oculi when opening the mouth (the Marin-Amat syndrome: inverse Marcus Gunn phenomenon), acquired after lower motor neurone facial (VII) nerve palsy and presumed to reflect aberrant reinnervation. Aberrant nerve regeneration is common to a number of synkinetic phenomena, such as elevation of a ptotic eyelid on swallowing (Ewart phenomenon) and upper eyelid elevation or retraction on attempted downgaze (pseudo-von Graefe's sign). Crocodile tears, lacrimation when salivating, due to reinnervation following a lower motor neurone facial nerve palsy, may also fall under this rubric, although there is no movement *per se* (autonomic synkinesis).

Abnormal synkinesis may be useful in assessing whether weakness is organic or functional (*cf.* Hoover's sign).

Synkinesis may also refer to the aggravation of limb rigidity detected when performing movements in the opposite limb (*e.g.*, clenching and relaxing the fist), also known as activated rigidity or Froment's sign.

#### **Cross References**

Crocodile tears; Ewart phenomenon; Froment's sign; Hoover's sign; Jaw winking; Pseudo-von Graefe's sign; Rigidity