A

Abadie's Sign

Abadie's sign is the absence or diminution of subjective pain sensation when exerting deep pressure on the Achilles tendon by squeezing. This is a frequent finding in the tabes dorsalis variant of neurosyphilis, *i.e*. with dorsal column disease, but now is more usually encountered in the context of diabetes mellitus as a consequence of intratendinous changes which might predispose to tendon rupture. The sign has also been reported in adrenomyeloneuropathy.

References

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Ohtomo R, Matsukawa T, Tsuji S, Iwata A. Abadie's sign in adrenomyeloneuropathy. *J Neurol Sci*. 2014; **340**: 245–6.

Cross Reference

Argyll Robertson pupil

Abdominal Paradox

- see PARADOXICAL BREATHING

Abdominal Reflexes

Both superficial and deep abdominal reflexes are described, of which the superficial (cutaneous) reflexes are the more commonly tested in clinical practice. A wooden stick or pin is used to scratch the abdominal wall, from the flank to the midline, parallel to the line of the dermatomal strips, in upper (supraumbilical), middle (umbilical), and lower (infraumbilical) areas. The manoeuvre is best performed at the end of expiration when the abdominal muscles are relaxed, since the reflexes may be lost with muscle tensing; to avoid this, patients should lie supine with their arms by their sides. Superficial abdominal reflexes are lost in a number of circumstances:

- Normal ageing.
- Obesity.
- Following abdominal surgery.
- Following multiple pregnancies.
- In acute abdominal disorders (Rosenbach's sign).

However, absence of all superficial abdominal reflexes may be of localising value for corticospinal pathway damage (upper motor neurone lesions) above T6. Lesions at or below T10 lead to selective loss of the lower reflexes with the upper and middle reflexes intact, in which case Beevor's sign may also be present. All abdominal reflexes are preserved with lesions below T12.

Abdominal reflexes are said to be lost early in multiple sclerosis, but late in motor neurone disease, an observation of possible clinical use, particularly when differentiating the progressive lateral sclerosis variant of motor neurone disease from multiple sclerosis. However, no prospective study of abdominal reflexes in multiple sclerosis has been reported.

Reference

Dick JPR. The deep tendon and the abdominal reflexes. *J Neurol Neurosurg Psychiatry*. 2003; **74**: 150–3.

A.J. Larner, *A Dictionary of Neurological Signs*,
DOI 10.1007/978-3-319-29821-4_1

Cross References
Beevor's sign; Upper motor neurone (UMN) syndrome

Abducens (VI) Nerve Palsy

Abducens, abducent, or sixth, cranial nerve palsy causes a selective weakness of the lateral rectus muscle resulting in impaired abduction of the affected eye, manifest clinically as diplopia on lateral gaze, or on shifting gaze from a near to a distant object.

Abducens nerve palsy may occur anywhere along its nuclear, fascicular, subarachnoid, petrous apex, cavernous sinus, and orbital course. It may be an isolated finding or be associated with accompanying neurological features which may assist with topographical diagnosis, for example with ipsilateral postganglionic Horner syndrome (posterior cavernous sinus: Parkinson syndrome) or twelfth nerve palsy (clivus).

Many causes of abducens nerve palsy have been described, but the most common include:

- Microinfarction in the nerve, due to hypertension, diabetes mellitus.
- Raised intracranial pressure: a "false-localising sign", possibly caused by stretching of the nerve in its long intracranial course over the ridge of the petrous temporal bone.
- Nuclear pontine lesions: congenital, *e.g.* Duane retraction syndrome, Möbius syndrome.
- Mass lesions anywhere along the course.

Bilateral abducens palsy is more often seen with tumours, subarachnoid haemorrhage, meningitis, Wernicke's encephalopathy, and raised intracranial pressure.

Isolated weakness of the lateral rectus muscle causing impaired abduction may also occur in myasthenia gravis. In order not to overlook this fact, and miss a potentially treatable condition, it is probably better to label isolated abduction failure of the eye initially as "lateral rectus palsy", rather than abducens nerve palsy, until the aetiological diagnosis is established.

Excessive or sustained convergence associated with a midbrain lesion (at the diencephalic-mesencephalic junction) may also result in slow or restricted abduction, so called pseudo-abducens palsy or "midbrain pseudo-sixth".

Reference
Leigh RJ, Zee DS. The neurology of eye movements. 4th ed. Oxford: Oxford University Press; 2006. p. 416–23.

Cross References
Diplopia; "False-localising signs"

Abductor Sign

The abductor sign is tested by asking the patient to abduct each leg whilst the examiner opposes movement with hands placed on the lateral surfaces of the patient's legs: the leg contralateral to the abducted leg shows opposite actions dependent upon whether paresis is organic or non-organic. Abduction of a paretic leg is associated with the sound leg remaining fixed in organic paresis, but in non-organic paresis there is hyperadduction. Hence the abductor sign is suggested to be useful to detect non-organic paresis.

Reference
Sonoo M. Abductor sign: a reliable new sign to detect unilateral non-organic paresis of the lower limb. *J Neurol Neurosurg Psychiatry*. 2004; **75**: 121–5.

Cross Reference
Functional weakness and sensory disturbance

Absence

An absence, or absence attack, is a brief interruption of awareness of epileptic origin. This may be a barely noticeable suspension of speech or attentiveness, without postictal confusion or any awareness that an attack has occurred, as in idiopathic generalized epilepsy of absence type (absence epilepsy; petit mal), a disorder exclusive to childhood and associated with 3 Hz spike and slow wave EEG abnormalities.

Absence epilepsy may be confused with a more obvious distancing, staring, "trance-like" state, or "glazing over", unresponsive to question or command, possibly with associated automatisms such as lip smacking, due to a complex partial seizure of temporal lobe origin ("atypical absence").

Ethosuximide and/or sodium valproate are the treatments of choice for idiopathic generalized absence epilepsy, whereas carbamazepine, sodium valproate, or lamotrigine are first-line agents for localisation-related complex partial seizures.

Cross References

Automatism; Seizures

Abulia

Abulia (or aboulia) is a "syndrome of hypofunction", characterized by lack of initiative, spontaneity and drive (aspontaneity), apathy, slowness of thought (bradyphrenia), and blunting of emotional responses and response to external stimuli. It may be confused with the psychomotor retardation of depression and is sometimes labelled as "pseudodepression". More plausibly, abulia has been thought of as a minor or partial form of akinetic mutism. A distinction may be drawn between abulia major (= akinetic mutism) and abulia minor, a lesser degree of abulia associated particularly with bilateral caudate nucleus stroke and thalamic infarcts in the territory of the polar artery and infratentorial stroke. There may also be some clinical overlap with catatonia and athymhormia.

Abulia may result from frontal lobe damage, most particularly that involving the frontal convexity, and has also been reported with focal lesions of the caudate nucleus, thalamus and midbrain. As with akinetic mutism, it is likely that lesions anywhere in the "centromedial core" of brain frontal-subcortical circuitry, from frontal lobes to brainstem, may produce this picture.

Pathologically, abulia may be observed in:

- Infarcts in anterior cerebral artery territory and ruptured anterior communicating artery aneurysms, causing basal forebrain damage.
- Closed head injury.
- Parkinson's disease; sometimes as a forerunner of a frontal type dementia.
- Other causes of frontal lobe disease: tumour, abscess.
- Metabolic, electrolyte disorders: hypoxia, hypoglycaemia, hepatic encephalopathy.

Treatment is of the underlying cause where possible. There is anecdotal evidence that dopaminergic therapy such as bromocriptine and levodopa may sometimes help.

References

Fisher CM. Abulia. In: Bogousslavsky J, Caplan L, editors. Stroke syndromes. Cambridge: Cambridge University Press; 1995. p. 182–7.

Ghoshal S, Gokhale S, Rebovich G, Caplan LR. The neurology of decreased activity: abulia. *Rev Neurol Dis*. 2011; **8**: e55–67.

Vijayaraghavan L, Krishnamoorthy ES, Brown RG, Trimble MR. Abulia: a Delphi survey of British neurologists and psychiatrists. *Mov Disord*. 2002; **17**: 1052–7.

Cross References

Akinetic mutism; Apathy; Athymhormia; Bradyphrenia; Catatonia; Frontal lobe syndromes; Psychomotor retardation

Acalculia

First named and described by Henschen in 1919, acalculia or dyscalculia is difficulty or inability in performing simple mental arithmetic. This depends on two processes: number processing and calculation. A deficit confined to the latter process is termed anarithmetia. Acalculia may be classified as:

- *Primary*:
  A specific deficit in arithmetical tasks, more severe than any other co-existing cognitive dysfunction.

- *Secondary*:
  In the context of other cognitive impairments, for example of language (aphasia, alexia or agraphia for numbers), attention, memory, or space perception (*e.g.* neglect). Acalculia may occur in association with alexia, agraphia, finger agnosia, right-left disorientation and difficulty spelling words as one part of the Gerstmann syndrome associated with lesions of the dominant parietal lobe.

Secondary acalculia is the more common variety.

Isolated acalculia may be seen with lesions of:

- Dominant (left) parietal/temporal/occipital cortex, especially involving the angular gyrus (Brodmann areas 39 and 40).
- Medial frontal lobe (perhaps as a consequence of impaired problem solving ability).
- Subcortical structures (caudate nucleus, putamen, internal capsule).

Impairments may be remarkably focal, for example one operation (*e.g.* subtraction) may be preserved whilst all others are impaired.

In patients with mild to moderate Alzheimer's disease with acalculia but no attentional or language impairments, cerebral glucose metabolism was found to be impaired in the left inferior parietal lobule and inferior temporal gyrus. Preservation of calculation skills in the face of total language dissolution (production and comprehension) has been reported with focal left temporal lobe atrophy.

References

Boller F, Grafman J. Acalculia: historical development and current significance. *Brain Cogn*. 1983; **2**: 205–23.

Denburg N, Tranel D. Acalculia and disturbances of body schema. In: Heilman KM, Valenstein E, editors. Clinical neuropsychology. 4th ed. Oxford: Oxford University Press; 2003. p. 161–84.

Gitelman DR. Acalculia: a disorder of numerical cognition. In: D'Esposito M, editor. Neurological foundations of cognitive neuroscience. Cambridge: MIT Press; 2003. p. 129–63.

Cross References

Agraphia; Alexia; Aphasia; Gerstmann syndrome; Neglect

Accommodation Reflex

- see PUPILLARY REFLEXES

Achilles Reflex

- see ANKLE JERK, ANKLE REFLEX

Achromatopsia

Achromatopsia, or dyschromatopsia, is an inability or impaired ability to perceive colours. This may be ophthalmological or neurological in origin, and congenital or acquired; only in the latter case does the patient complain of impaired colour vision.

Achromatopsia is most conveniently tested for clinically by using pseudoisochromatic figures (*e.g.* Ishihara plates), although these were specifically designed for detecting congenital colour blindness and test the red-green channel more than blue-yellow. Sorting colours according to hue, for example with the Farnsworth-Munsell 100 Hue test, is more quantitative, but more time consuming. Difficulty performing these tests does not always reflect achromatopsia (see Pseudoachromatopsia).

Probably the most common cause of achromatopsia is inherited "colour blindness", of which several types are recognized: in monochromats only one of the three cone photoreceptor classes is affected, in dichromats two; anomalous sensitivity to specific wavelengths of light may also occur (anomalous trichromat). These inherited dyschromatopsias are binocular, symmetrical, and do not change with time. There are also a number of retinal dystrophies

characterised by achromatopsia, photophobia, nystagmus, and severely reduced visual acuity for which a number of mutant genes have been characterised (CNGA3, CNBG3, GNAT2, PDE6C, PDE6H, ATF6).

Acquired achromatopsia may result from damage to the optic nerve or the cerebral cortex. Unlike inherited conditions, these deficits are noticeable (patients describe the world as looking "grey" or "washed out"; *grisaille* is the technical, artistic, term) and may be confined to only part of the visual field (*e.g.* hemiachromatopsia). Optic neuritis typically impairs colour vision (red-green > blue-yellow) and this defect may persist even when other features of the acute inflammation (impaired visual acuity, central scotoma) remit.

Cerebral achromatopsia results from cortical damage to the inferior occipitotemporal area, most usually as a result of infarction. Area V4 of the visual cortex, which is devoted to colour processing, is located in the occipitotemporal (fusiform) and lingual gyri. There is also a loss of colour imagery. Unilateral lesions may produce a homonymous hemiachromatopsia. Lesions in this region may also produce prosopagnosia, alexia, and visual field defects, either a peripheral scotoma which is always in the upper visual field, or a superior quadrantanopia, reflecting damage to the inferior limb of the calcarine sulcus in addition to the adjacent fusiform gyrus. Transient achromatopsia in the context of vertebrobasilar ischaemia has been reported.

The differential diagnosis of achromatopsia encompasses colour agnosia, a loss of colour knowledge despite intact perception; and colour anomia, an inability to name colours despite intact perception.

References

Bartolomeo P, Bachoud-Lévi AC, Thiebault de Schotten M. The anatomy of cerebral achromatopsia: a reappraisal and comparison of two case reports. *Cortex*. 2014; **56**: 138–44.

Zeki S. A century of cerebral achromatopsia. *Brain*. 1990; **113**: 1721–77.

Cross References

Agnosia; Alexia; Anomia; Prosopagnosia; Pseudoachromatopsia; Quadrantanopia; Scotoma; Xanthopsia

Acoasm

- see HALLUCINATION

Acousticopalpebral Reflex

- see BLINK REFLEX

Acroparaesthesia

- see PARAESTHESIA

Action Dystonia

- see DYSTONIA

Action Myoclonus

- see MYOCLONUS

Adiadochokinesia

- see DYSDIADOCHOKINESIA

Adie's Syndrome, Adie's Tonic Pupil

- see HOLMES-ADIE PUPIL, HOLMES-ADIE SYNDROME

Adson's Test

Adson's test may be helpful in the diagnosis of vascular thoracic outlet syndrome, along with Roos test. The arm is extended at the elbow, abducted, then rotated posteriorly; following deep inspiration, the patient's head is turned from one side to the other. Loss of the radial pulse may occur in normals but a bruit over the brachial artery is thought to suggest the

presence of entrapment. A Doppler Adson's test over the subclavian artery may predict successful outcome from thoracic outlet decompression surgery.

Reference

Lee AD, Agarwal S, Sadhu D. Doppler Adson's test: predictor of outcome of surgery in non-specific thoracic outlet syndrome. *World J Surg*. 2006; **30**: 291–2.

Cross Reference

Roos test

Adventitious Movements

- see STEREOTYPY

Affective Agnosia

- see AGNOSIA; APROSODIA, APROSODY

Afferent Pupillary Defect (APD)

- see RELATIVE AFFERENT PUPILLARY DEFECT (RAPD)

Age-Related Signs

A number of neurological signs are reported to be more prevalent with increasing age, and related to ageing *per se* rather than any underlying age-related disease. Hence these signs are not necessarily of pathological significance when assessing the neurological status of older individuals, although there are methodological difficulties in reaching such conclusions.

A brief topographical overview of age-related signs includes:

- *Cognitive function*:

  Loss of processing speed, cognitive flexibility, efficiency of working memory (sustained attention).

  Preservation of vocabulary, remotely learned information including semantic networks, and well-encoded new information.
- *Cranial nerves*:

  I: olfactory sense diminished.

  II, III, IV, VI: presbyopia; reduced visual acuity, depth perception, contrast sensitivity, motion perception; "senile miosis"; restricted upward conjugate gaze.

  VIII: presbycusis; impaired vestibulospinal reflexes.
- *Motor system*:

  Appearance: loss of muscle bulk; "senile" tremor.

  Tone: rigidity; *gegenhalten*/paratonia.

  Power: decline in muscle strength.

  Co-ordination: impaired speed of movement (bradykinesia).

  Reflexes:

    Phasic muscle stretch reflexes: depressed or absent, especially ankle (Achilles tendon) jerk; jaw jerk.

    Cutaneous (superficial) reflexes: abdominal reflexes may be depressed with ageing.

    Primitive/developmental reflexes: glabellar, snout, palmomental, grasp reflexes may be more common with ageing.

  Impairments of gait; parkinsonism.
- *Sensory system*:

  Decreased sensitivity to vibratory perception; ± pain, temperature, proprioception, two-point discrimination.

Neuroanatomical correlates of some of these signs have been defined. There does seem to be an age-related loss of distal sensory axons and of spinal cord ventral horn motor neurones accounting for sensory loss, loss of muscle bulk and strength, and reflex diminution.

References

Franssen EH. Neurologic signs in ageing and dementia. In: Burns A, editor. Ageing and dementia: a methodological approach. London: Edward Arnold; 1993. p. 144–74.

Larner AJ. Neurological signs of ageing. In: Sinclair A, Morley JE, Vellas B, editors. Pathy's principles and practice of geriatric medicine. 5th ed. Chichester: Wiley; 2012. p. 609–16.

McGeer PL, McGeer EG, Suzuki JS. Aging and extrapyramidal function. *Arch Neurol*. 1977; **34**: 33–5.

Vrancken AFJE, Kalmijn S, Brugman F, Rinkel GJE, Notermans NC. The meaning of distal sensory loss and absent ankle reflexes in relation to age. A meta-analysis. *J Neurol*. 2006; **253**: 578–89.

Cross References

Frontal release signs; Parkinsonism; Reflexes

Ageusia

Ageusia or hypogeusia is a loss or impairment of the sense of taste (gustation). This may be tested by application to each half of the protruded tongue the four classical tastes (sweet, sour, bitter, salt).

Isolated ageusia is most commonly encountered as a transient feature associated with coryzal illnesses of the upper respiratory tract, as with anosmia. Indeed, many complaints of loss of taste are in fact due to anosmia, since olfactory sense is responsible for the discrimination of many flavours. Ageusia as an adverse drug effect is described (*e.g*. with clopidogrel).

Neurological disorders may also account for ageusia. Afferent taste fibres run in the facial (VII) and glossopharyngeal (IX) cranial nerves, from taste buds in the anterior two-thirds and posterior one-third of the tongue respectively. Central neuronal processes run in the solitary tract in the brainstem and terminate in its nucleus (nucleus tractus solitarius), the rostral part of which is sometimes called the gustatory nucleus. Fibres then run to the ventral posterior nucleus of the thalamus, hence to the cortical area for taste adjacent to the general sensory area for the tongue (insular region).

Lesions of the facial nerve proximal to the departure of the chorda tympani branch in the mastoid (vertical) segment of the nerve (*i.e*. proximal to the emergence of the facial nerve from the stylomastoid foramen), can lead to ipsilateral impairment of taste sensation over the anterior two-thirds of the tongue, along with ipsilateral lower motor neurone facial weakness (*e.g*. in Bell's palsy), with or without hyperacusis.

Lesions of the glossopharyngeal nerve causing impaired taste over the posterior one-third of the tongue usually occur in association with ipsilateral lesions of the other lower cranial nerves (X, XI, XII; jugular foramen syndrome) and hence may be associated with dysphonia, dysphagia, depressed gag reflex, vocal cord paresis, anaesthesia of the soft palate, uvula, pharynx and larynx, and weakness of trapezius and sternocleidomastoid.

Ageusia as an isolated symptom of neurological disease is extremely rare, but has been described with focal central nervous system lesions (infarct, tumour, demyelination) affecting the nucleus of the tractus solitarius (gustatory nucleus) and/or thalamus, and with bilateral insular lesions. Anosmia and dysgeusia have also been reported following acute zinc loss.

Reference

Finelli PF, Mair RG. Disturbances of smell and taste. In: Bradley WG, Daroff RB, Fenichel GM, Jankovic J, editors. Neurology in clinical practice. 5th ed. Philadelphia: Butterworth Heinemann Elsevier; 2008. p. 255–62.

Cross References

Anosmia; Bell's palsy; Cacogeusia; Dysgeusia; Facial paresis, Facial weakness; Hyperacusis; Jugular foramen syndrome

Agnosopsia

This term has been suggested to describe the retention of accurate visual perceptual judgements despite lacking conscious visual perception, in other words knowing without seeing. Anopsognosia has also been used to describe this phenomenon. Unlike blindight, patients have some residual awareness of the presentation of stimuli.

Reference

Carota A, Calabrese P. The achromatic "philosophical zombie", a syndrome of cerebral achromatopsia with color anopsognosia. *Case Rep Neurol*. 2013; **5**: 98–103.

Cross Reference

Blindsight

Agnosia

Agnosia is a deficit of higher sensory (most often visual) processing causing impaired recognition. The term, coined by Freud in 1891, means literally "absence of knowledge", but its precise clinical definition continues to be a subject of debate. Lissauer (1890) originally conceived of two kinds of agnosia:

- *Apperceptive*:

  in which there is a defect of complex (higher order) perceptual processes.

- *Associative*:

  in which perception is thought to be intact but there is a defect in giving meaning to the percept by linking its content with previously encoded percepts (the semantic system); this has been described as "a normal percept that has somehow been stripped of its meaning", or "perception without knowledge".

These deficits should not be explicable by a concurrent intellectual impairment, disorder of attention, or by an inability to name or describe the stimulus verbally (anomia). As a corollary of this last point, some argue that there should be no language disorder (aphasia) to permit a diagnosis of agnosia.

Intact perception is sometimes used as a *sine qua non* for the diagnosis of agnosia, in which case it may be questioned whether apperceptive agnosia is truly agnosia. However, others retain this category, not least because the supposition that perception is normal in associative visual agnosia is probably not true. Moreover, the possibility that some agnosias are in fact higher order perceptual deficits remains: examples include some types of visual and tactile recognition of form or shape (*e.g.* agraphognosia; astereognosis; dysmorphopsia); some authorities label these phenomena "pseudoagnosias". The difficulty with definition perhaps reflects the continuing problem of defining perception at the physiological level. Other terms which might replace agnosia have been suggested, such as non-committal terms like "disorder of perception" or "perceptual defect", or as suggested by Hughlings Jackson "imperception".

Theoretically, agnosias can occur in any sensory modality, but some authorities believe that the only unequivocal examples are in the visual and auditory domains (*e.g.* prosopagnosia, and pure word deafness, respectively). Nonetheless, many other "agnosias" have been described, although their clinical definition may lie outwith some operational criteria for agnosia. With the passage of time, acquired agnosic defects merge into anterograde amnesia (failure to learn new information).

Anatomically, agnosias generally reflect dysfunction at the level of the association cortex, although they can on occasion result from thalamic pathology. Some may be of localizing value. The neuropsychological mechanisms underpinning these phenomena are often ill understood.

References

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Critchley M. The citadel of the senses and other essays. New York: Raven Press; 1986. p. 239.

Farah MJ. Visual agnosia: disorders of object recognition and what they tell us about normal vision. Cambridge: MIT Press; 1995.
Ghadiali E. Agnosia. *Adv Clin Neurosci Rehabil*. 2004; **4**(5): 18–20.
Cross References
Agraphognosia; Alexia; Amnesia; Anosognosia; Aprosodia, Aprosody; Asomatognosia; Astereognosis; Auditory agnosia; Autotopagnosia; Dysmorphopsia; Finger agnosia; Phonagnosia; Prosopagnosia; Pure word deafness; Simultanagnosia; Tactile agnosia; Topographagnosia; Visual agnosia; Visual form agnosia

Agrammatism
Agrammatism is a reduction in, or loss of, the production or comprehension of the syntactic elements of language, for example articles, prepositions, conjunctions, verb endings (*i.e*. the non-substantive components of language), whereas nouns and verbs are relatively spared. Despite this impoverishment of language, or "telegraphic speech", meaning is often still conveyed to auditors because of the high information content of verbs and nouns.

Agrammatism is encountered in Broca's type of non-fluent aphasia, associated with lesions of the posterior inferior part of the frontal lobe of the dominant hemisphere (Broca's area), and also in progressive non-fluent aphasia (the agrammatic variant of primary progressive aphasia) in which there may also be speech apraxia. Agrammatic speech may also be dysprosodic.
Cross References
Aphasia; Aprosodia, Aprosody; Speech apraxia

Agraphaesthesia
Agraphaesthesia, dysgraphaesthesia, or graphanaesthesia, is a loss or impairment of the ability to recognize letters or numbers written or traced on the skin, *i.e*. of graphaesthesia. This ability was first described by Henry Head in 1920. Whether this is a perceptual deficit or a tactile agnosia ("agraphognosia") has been debated.

It has been observed with defects at all levels of the nervous system from periphery, to spinal cord, to parietal cortex, and is usually if not invariably associated with other sensory deficits (*e.g*. two-point discrimination). Hence it would seem to have little value as a localizing sign. By contrast, stereognosis and Braille reading are complex motion-dependent derived functions, so their concurrence with agraphaesthesia would point to a cortical lesion (*e.g*. as seen in corticobasal degeneration).
Reference
Bender MB, Stacy C, Cohen J. Agraphesthesia. A disorder of directional cutaneous kinesthesia or a disorientation in cutaneous space. *J Neurol Sci*. 1982; **53**: 531–55.
Cross References
Agnosia; Astereognosis; Tactile agnosia; Two-point discrimination

Agraphia
Agraphia, or dysgraphia, is a loss or disturbance of the ability to write or spell. Since writing depends not only on language function but also on motor, visuospatial and kinaesthetic function, many factors may lead to dysfunction. Agraphias may be classified as follows:

- *Central*, *aphasic*, *or linguistic dysgraphias*:

  These are usually associated with aphasia and alexia, and the deficits mirror those seen in the Broca/anterior/motor and Wernicke/posterior/sensory types of aphasia. Oral spelling is impaired. From the linguistic viewpoint, two types of paragraphia may be distinguished, *viz*.:
  Surface/lexical/semantic dysgraphia: misspelling of irregular words, producing phonologically plausible errors (*e.g*. simtums for symptoms); this is seen with left temporoparietal lesions, *e.g*. Alzheimer's disease, Pick's disease;
  Deep/phonological dysgraphia: inability to spell unfamiliar words and non-words; semantic errors; seen with extensive left hemisphere damage.

- *Mechanical agraphia*:

  Impaired motor control, due to paresis (as in dominant parietal damage), apraxia (may be accompanied by ideomotor limb apraxia), dyskinesia (hypokinetic or hyperkinetic), or dystonia; oral spelling may be spared.

- *Neglect* (*spatial*) *dysgraphia*:

  Associated with other neglect phenomena consequent upon a non-dominant hemisphere lesion; there may be missing out or misspelling of the left side of words (paragraphia); oral spelling may be spared.

- *Pure agraphia*:

  A rare syndrome in which oral language, reading and praxis are normal.

A syndrome of agraphia, alexia, acalculia, finger agnosia, right-left disorientation and difficulty spelling words (Gerstmann syndrome) may be seen with dominant parietal lobe pathologies.

Writing disturbance due to abnormal mechanics of writing is the most sensitive language abnormality in delirium, possibly because of its dependence on multiple functions.

References

Benson DF, Ardila A. Aphasia: a clinical perspective. New York: Oxford University Press; 1996. p. 212–34

Roeltgen DP. Agraphia. In: Heilman KM, Valenstein E, editors. Clinical neuropsychology. 4th ed. Oxford: Oxford University Press; 2003. p. 126–45.

Cross References

Alexia; Allographia; Aphasia; Apraxia; Broca's aphasia; Fast micrographia; Gerstmann syndrome; Hypergraphia; Macrographia; Micrographia; Neglect; Wernicke's aphasia

Agraphognosia

- see AGRAPHAESTHESIA

Agrypnia (Excitata)

Agrypnia (from the Greek, to chase sleep), or agrypnia excitata, is characterised by severe, total insomnia of long duration, sometimes with persistent motor and autonomic hyperactivation (hence agrypnia excitata).

Recognised causes of agrypnia include trauma to the brainstem and/or thalamus, von Economo's disease, and trypanosomiasis.

Agrypnia excitata (AE) has been used to describe three particular disorders: fatal familial insomnia, a prion disease associated with thalamic degeneration; Morvan's syndrome, an autoimmune encephalitis often associated with autoantibodies directed against voltage-gated potassium channels; and delirium tremens, an alcohol withdrawal syndrome. The pathophysiology of AE in these various conditions is thought to be loss of cortico-limbic inhibitory control of the hypothalamus and ascending brainstem reticular formation.

Reference

Provini F. Agrypnia excitata. *Curr Neurol Neurosci Rep*. 2013; **13**: 341.

Akathisia

Akathisia is a feeling of inner restlessness, often associated with restless movements of a continuous and often purposeless nature, such as rocking to and fro, repeatedly crossing and uncrossing the legs, standing up and sitting down, pacing up and down (forced walking, tasikinesia). Moaning, humming, and groaning may also be features. Voluntary suppression of the movements may exacerbate inner tension or anxiety. The Barnes Akathisia Rating Scale is the standard assessment scale.

Recognized associations of akathisia include Parkinson's disease and neuroleptic medication use (acute or tardive side effect), suggesting that dopamine depletion may contribute to the pathophysiology. Dopamine depleting agents (*e.g.* tetrabenazine, reserpine) may also cause akathisia. Acute akathisia following pontine infarction is reported.

Treatment of akathisia by reduction or cessation of neuroleptic therapy may help, but may exacerbate coexistent psychosis. Centrally acting β-blockers such as propranolol may also be helpful, as may anticholinergic agents, amantadine, clonazepam, and clonidine.

References

Barnes TR. A rating scale for drug-induced akathisia. *Br J Psychiatry*. 1989; **154**: 672–6.
Sachdev P. Akathisia and restless legs. Cambridge: Cambridge University Press; 1995.

Cross References

Parkinsonism; Tasikinesia; Tic

Akinesia

Akinesia is a lack of, or an inability to initiate, voluntary movements. More usually in clinical practice there is a difficulty (reduction, delay), rather than complete inability, in the initiation of voluntary movement, perhaps better termed bradykinesia, or reduced amplitude of movement or hypokinesia. These difficulties cannot be attributed to motor unit or pyramidal system dysfunction. Reflexive motor activity may be preserved (*kinesis paradoxica*). There may be concurrent slowness of movement, also termed bradykinesia.

Akinesia may co-exist with any of the other clinical features of extrapyramidal system disease, particularly rigidity, but the presence of akinesia is regarded as an absolute requirement for the diagnosis of parkinsonism.

Hemiakinesia may be a feature of motor neglect of one side of the body (possibly a motor equivalent of sensory extinction). Bilateral akinesia with mutism (akinetic mutism) may occur if pathology is bilateral. Pure akinesia, without rigidity or tremor, may occur: if levodopa-responsive, this is usually due to Parkinson's disease; if levodopa-unresponsive, it may be the harbinger of progressive supranuclear palsy. A few patients with PSP have "pure akinesia" without other features until late in the disease course; freezing of gait may also be a feature.

Neuroanatomically, akinesia is a feature of disorders affecting:

- Frontal-subcortical structures, *e.g*. the medial convexity subtype of frontal lobe syndrome.
- Basal ganglia.
- Ventral thalamus.
- Limbic system (anterior cingulate gyrus).

Neurophysiologically, akinesia is associated with loss of dopamine projections from the substantia nigra to the putamen.

Pathological processes underpinning akinesia include:

- Neurodegeneration, *e.g*. Parkinson's disease, progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome), multiple system atrophy (striatonigral degeneration); akinesia may occur in some frontotemporal lobar degeneration syndromes, Alzheimer's disease, and some prion diseases.
- Hydrocephalus.
- Neoplasia, *e.g*. butterfly glioma of the frontal lobes.
- Cerebrovascular disease.

Akinesia resulting from nigrostriatal dopamine depletion (*i.e*. idiopathic Parkinson's disease) may respond to treatment with levodopa or dopamine agonists. However, many parkinsonian/akinetic-rigid syndromes show no or only partial response to these agents.

References

Imai H. Clinicophysiological features of akinesia. *Eur Neurol*. 1996; **36**(Suppl 1): 9–12.
Riley DE, Fogt N, Leigh RJ. The syndrome of "pure akinesia" and its relationship to PSP. *Neurology*. 1994; **44**: 1025–9.

Cross References
Akinetic mutism; Bradykinesia; Extinction; Freezing of gait; Frontal lobe syndromes; Hemiakinesia; Hypokinesia; Hypometria; *Kinesis paradoxica*; Neglect; Parkinsonism

Akinetic Mutism

Akinetic mutism is a "syndrome of negatives", characterized by lack of voluntary movement (akinesia), absence of speech (mutism), lack of response to question and command, but with normal alertness and sleep-wake cycles (*cf*. coma). Blinking (spontaneous and to threat) is preserved. Frontal release signs, such as grasping and sucking, may be present, as may double incontinence, but there is a relative paucity of upper motor neurone signs affecting either side of the body, suggesting relatively preserved descending pathways. Akinetic mutism represents an extreme form of abulia, hence sometimes referred to as abulia major.

Pathologically, akinetic mutism is associated with bilateral lesions of the "centromedial core" of the brain interrupting reticular-cortical or limbic-cortical pathways but which spare corticospinal pathways; this may occur at any point from frontal lobes to brainstem.

Different forms of akinetic mutism are sometimes distinguished, *e.g*. according to lesion location:

- Fronto-diencephalic: associated with bilateral occlusion of the anterior cerebral arteries or with haemorrhage and vasospasm from anterior communicating artery aneurysms; damage to the cingulate gyri appears crucial but not sufficient for this syndrome.
- Akinetic mutism with disturbances of vertical eye movements and hypersomnia: associated with paramedian thalamic and thalamo-mesencephalic strokes.

Or according to lesion location and clinical phenotype:

- Mesencephalic-diencephalic region, also called apathetic akinetic mutism or somnolent mutism;
- Anterior cingulate gyrus and adjacent frontal lobes, also called hyperpathic akinetic mutism, a more severe presentation.

Other structures (*e.g*. globus pallidus) have sometimes been implicated. Pathology may be vascular, neoplastic, or structural (subacute communicating hydrocephalus), and evident on structural brain imaging. Akinetic mutism may be the final state common to the end-stages of a number of neurodegenerative pathologies. EEG may show slowing with lack of desynchronization following external stimuli.

Occasionally, treatment of the cause may improve akinetic mutism (*e.g*. relieving hydrocephalus). Agents such as dopamine agonists (*e.g*. bromocriptine), ephedrine and methylphenidate have also been tried.

References
Cairns H. Disturbances of consciousness with lesions of the brain stem and diencephalon. *Brain*. 1952; **75**: 109–46.
Nagaratnam N, Nagaratnam K, Ng K, Diu P. Akinetic mutism following stroke. *J Clin Neurosci*. 2004; **11**: 25–30.
Ross ED, Stewart RM. Akinetic mutism from hypothalamic damage: successful treatment with dopamine agonists. *Neurology*. 1981; **31**: 1435–9.
Shetty AC, Morris J, O'Mahony P. Akinetic mutism – not coma. *Age Ageing*. 2009; **38**: 350–1.
Cross References
Abulia; Akinesia; Athymhormia; Blink reflex; Catatonia; Coma; Frontal lobe syndromes; Frontal release signs; Grasp reflex; Locked-in syndrome; Mutism

Akinetic Rigid Syndrome

- see PARKINSONISM

Akinetopsia

Akinetopsia is a specific inability to see objects in motion, the perception of other visual attributes such as colour, form, and depth, remaining intact. This statokinetic dissociation may be known as Riddoch's phenomenon; the syndrome may also be called cerebral visual motion blindness. Such cases, although exceptionally rare, suggest a distinct neuroanatomical substrate for movement vision, as do cases in which motion vision is selectively spared in a scotomatous area (Riddoch's syndrome).

Akinetopsia reflects a lesion selective to area V5 of the visual cortex. Clinically there may be associated acalculia and aphasia.

References

Zeki S. Cerebral akinetopsia (cerebral visual motion blindness). *Brain*. 1991; **114**: 811–24.

Zihl J, Von Cramon D, Mai N. Selective disturbance of movement vision after bilateral brain damage. *Brain*. 1983; **106**: 313–40.

Cross References

Acalculia; Aphasia; Riddoch's phenomenon; *Zeitraffer* phenomenon

Alalia

Alalia is now an obsolete term, once used to describe a disorder of the material transformation of ideas into sounds. Lordat used it to describe the aphasia following a stroke.

Reference

Bogousslavsky J, Assal G. Stendhal's aphasic spells: the first report of transient ischemic attacks followed by stroke. In: Bogousslavsky J, Hennerici MG, Bäzner H, Bassetti C, editors. Neurological disorders in famous artists – part 3. Basel: Karger; 2010. p. 130–42. [at 139].

Cross References

Aphasia; Aphemia

Alexia

Alexia is an acquired disorder of reading. The word dyslexia, though in some ways equivalent, is often used to denote a range of disorders in people who fail to develop normal reading skills in childhood. Alexia, in contrast, may be described as an acquired dyslexia. Alexia may be categorised as:

- *Peripheral*:

  A defect of perception or decoding the visual stimulus (written script); other language functions are often intact.
- *Central*:

  A breakdown in deriving meaning; other language functions are often also affected.

Peripheral alexias include:

- *Alexia without agraphia*:

  Also known as pure alexia or pure word blindness. This is the archetypal peripheral alexia. Patients lose the ability to recognise written words quickly and easily; they seem unable to process all the elements of a written word in parallel. They can still access meaning but adopt a laborious letter-by-letter strategy for reading, with a marked word-length effect (*i.e.* greater difficulty reading longer words). Patients with pure alexia may be able to identify and name individual letters, but some cannot manage even this ("global alexia"). Tracing letters with a finger may speed up recognition ("Wilbrand's sign"). Strikingly, the patient can write at normal speed (*i.e.* no agraphia) but is then unable to read what they have just written. Alexia without agraphia often coexists with a right homonymous hemianopia, and colour anomia or impaired colour perception (achromatopsia); this latter may be restricted

to one hemifield, classically right-sided (hemiachromatopsia). Pure alexia has been characterized by some authors as a limited form of associative visual agnosia or ventral simultanagnosia. The term word blindness was first used by Henry Charlton Bastian in 1869; Sir William Broadbent (1872) and James Hinshelwood (1895) were also early writers on the subject.

- *Hemianopic alexia*:

  This occurs when a right homonymous hemianopia encroaches into central vision. Patients tend to be slower with text than single words as they cannot plan rightward reading saccades.

- *Neglect alexia*:

  Or hemiparalexia, results from failure to read either the beginning or end of a word (more commonly the former) in the absence of a hemianopia, due to hemispatial neglect.

The various forms of peripheral alexia may coexist; following a stroke, patients may present with global alexia which evolves to a pure alexia over the following weeks.

Pure alexia is caused by damage to the left occipito-temporal junction or its afferent inputs from early mesial visual areas or its efferent outputs to the medial temporal lobe. Global alexia usually occurs when there is additional damage to the splenium or white matter above the occipital horn of the lateral ventricle. Hemianopic alexia is usually associated with infarction in the territory of the posterior cerebral artery damaging geniculostriate fibres or area V1 itself, but can be caused by any lesion outside the occipital lobe that causes a macular splitting homonymous field defect. Neglect alexia is usually caused by occipito-parietal lesions, right-sided lesions causing left neglect alexia.

Central (linguistic) alexias include:

- *Alexia with aphasia*:

  Patients with aphasia often have coexistent difficulties with reading (reading aloud and/or comprehending written text) and writing (alexia with agraphia, such patients may have a complete or partial Gerstmann syndrome, the so-called "third alexia" of Benson). The reading problem parallels the language problem; thus in Broca's aphasia reading is laboured with particular problems reading function words (of, at) and verb inflections (-ing, -ed); in Wernicke's aphasia numerous paraphasic errors are made.

From the linguistic viewpoint, different types of paralexia (substitution in reading) may be distinguished:

- *Surface dyslexia*:

  Reading by sound: there are regularization errors with exception words (*e.g.* pint pronounced to rhyme with mint), but non-words can be read; this may be seen with left medial ± lateral temporal lobe pathology, *e.g.* infarction, semantic dementia, late Alzheimer's disease.

- *Phonological dyslexia*:

  Reading by sight: difficulties with suffixes, unable to read non-words; left temporo-parietal lobe pathology.

- *Deep dyslexia*:

  The inability to translate orthography to phonology, manifesting as an inability to read plausible non-words (as in phonological dyslexia), plus semantic errors related to word meaning rather than sound (*e.g.* sister read as uncle); visual errors are also common (*e.g.* sacred read as scared). Deep dyslexia is seen with extensive left hemisphere temporo-parietal damage.

The term transcortical alexia has been used to describe patients with Alzheimer's disease with severe comprehension deficits who nonetheless are able to read aloud virtually without error all regular and exception words.

References
Coslett HB. Acquired dyslexia. In: D'Esposito M, editor. Neurological foundations of cognitive neuroscience. Cambridge: MIT Press; 2003. p. 109–27.
Farah MJ. Visual agnosia: disorders of object recognition and what they tell us about normal vision. Cambridge: MIT Press; 1995.
Leff A, Starrfelt R. Alexia, Diagnosis, treatment and theory. London: Springer; 2014.
Cross References
Acalculia; Achromatopsia; Agnosia; Agraphia; Aphasia; Broca's aphasia; Gerstmann syndrome; Hemianopia; Macula sparing, Macula splitting; Neglect; Prosopagnosia; Saccades; Simultanagnosia; Visual agnosia; Visual field defects; Wernicke's aphasia; Wilbrand's sign

Alexithymia

Alexithymia is a reduced ability to identify and express ones feelings. This may contribute to various physical and behavioural disorders. It may be measured using the Toronto Alexithymia Score. There is evidence from functional imaging studies that alexithymics process facial expressions differently from normals, leading to the suggestion that this contributes to disordered affect regulation. Alexithymia is a common finding in split-brain patients, perhaps resulting from disconnection of the hemispheres.

References
Kano M, Fukudo S, Gyoba J, et al. Specific brain processing of facial expressions in people with alexithymia: an $H_2{}^{15}O$-PET study. *Brain*. 2003; **126**: 1474–84.
TenHouten WD, Hoppe KD, Bogen JE, Walter DO. Alexithymia: an experimental study of cerebral commissurotomy patients and normal control subjects. *Am J Psychiatry*. 1986; **143**: 312–6.

"Alice in Wonderland" Syndrome

The name "Alice in Wonderland" syndrome was coined by Todd in 1955 to describe the phenomena of micro- or macrosomatognosia, altered perceptions of body image, although these had first been described by Lippman in the context of migraine some years earlier. It has subsequently been suggested that Charles Lutwidge Dodgson's own experience of migraine, recorded in his diaries, may have given rise to Lewis Carroll's descriptions of Alice's changes in body form, graphically illustrated in *Alice's Adventures in Wonderland* (1865) by Sir John Tenniel. Some authors have subsequently interpreted these as somesthetic migrainous auras whereas others challenge this on chronological grounds, finding no evidence in Dodgson's diaries for the onset of migraine until after he had written the Alice books. Moreover, migraine with somesthetic auras is rare, and Dodgson's diaries have no report of migraine-associated body image hallucinations.

Other conditions may also give rise to the phenomena of micro- or macrosomatognosia, including epilepsy, encephalitis, cerebral mass lesions, schizophrenia, and drug intoxication.

References
Fine EJ. The Alice in Wonderland syndrome. *Prog Brain Res*. 2013; **206**: 143–56.
Larner AJ. The neurology of "Alice". *Adv Clin Neurosci Rehabil*. 2005; **4**(6): 35–6.
Todd J. The syndrome of Alice in Wonderland. *CMAJ*. 1955; **73**: 701–4.
Cross References
Aura; Metamorphopsia

Alien Grasp Reflex

The term alien grasp reflex has been used to describe a grasp reflex occurring in full consciousness, which the patient could anticipate but perceived as alien (*i.e.* not modified by will), occurring in the absence of other abnormal movements. These phenomena were associated with an intrinsic tumour of the right (non-dominant) frontal lobe. It was suggested that the grasp reflex and alien hand syndromes are not separate entities but part of the spectrum of frontal lobe dysfunction, the term "alien grasp reflex" attempting to emphasize the overlap.

Reference
Silva MT, Howard RS, Kartsounis LD, Ross Russell RW. The alien grasp reflex. *Eur Neurol*. 1996; **36**: 55–6.

Cross References
Alien hand, Alien limb; Grasp reflex

Alien Hand, Alien Limb

An alien limb, most usually the arm but occasionally the leg, is one which manifests slow, involuntary, wandering (levitating), quasi-purposive movements. An arm so affected may show apraxic difficulties in performing even the simplest tasks, and may be described by the patient as uncooperative or "having a mind of its own" (hence alternative names such as anarchic hand sign, *le main étranger*, and "Dr Strangelove syndrome"). These phenomena may be associated with a prominent grasp reflex, forced groping, intermanual conflict, magnetic movements of the hand, and levitation of the limb.

Different types of alien hand/limb have been described, reflecting the differing anatomical locations of underlying lesions:

- Anterior or motor types:
  Callosal type: characterized primarily by intermanual conflict.
  Frontal type: shows features of environmental dependency, such as forced grasping and groping, and utilization behaviour.
- Sensory or posterior variant:
  Resulting from a combination of cerebellar, optic, and sensory ataxia; rare.

A paroxysmal alien hand has been described, probably related to seizures of frontomedial origin.

Recognized pathological associations of alien limb include:

- Corticobasal (ganglionic) degeneration.
- Corpus callosum tumours, haemorrhage.
- Medial frontal cortex infarction (territory of the anterior cerebral artery).
- Trauma and haemorrhage affecting both corpus callosum and medial frontal area.
- Alzheimer's disease, familial Creutzfeldt-Jakob disease (very rare).
- Posterior cerebral artery occlusion (sensory variant).
- Following commissurotomy (corpus callosotomy alone insufficient).

Disconnection of parietal cortex (especially right side) from other cortical areas may underpin alien limb phenomenon.

References
Brion S, Jedynak CP. Troubles du transfer interhemisphérique. A propos de trois observations de tumeurs du corps calleux. Le signe de la main étrangère. *Rev Neurol (Paris)*. 1972; **126**: 257–66.
Fisher CM. Alien hand phenomena: a review with the addition of six personal cases. *Can J Neurol Sci*. 2000; **27**: 192–203.
Graff-Radford J, Rubin MN, Jones DT, et al. The alien limb phenomenon. *J Neurol*. 2013; **260**: 1880–8.

Cross References
Alien grasp reflex; Apraxia; Ataxia; "Compulsive grasping hand"; Forced groping; Grasp reflex; Intermanual conflict; Levitation; Magnetic movements; Utilization behaviour

Alienation Du Mot

This has been used to describe a loss of the feeling of familiarity with a word, part of the comprehension deficit seen in semantic dementia.

Reference
Poeck K, Luzzatti C. Slowly progressive aphasia in three patients: the problem of accompanying neuropsychological deficit. *Brain*. 1988; **111**: 151–68.

Alloacousia

Alloacousia describes a form of auditory neglect seen in patients with unilateral spatial neglect, characterised by spontaneous ignoring of people addressing the patient from the contralesional side, failing to respond to questions, or answering as if the speaker were on the ipsilesional side.

Reference
Heilman K, Valenstein E. Auditory neglect in man. *Arch Neurol*. 1972; **26**: 32–5.

Cross Reference
Neglect

Alloaesthesia

Alloaesthesia (allesthesia, alloesthesia) is the condition in which a sensory stimulus given to one side of the body is perceived at the corresponding area on the other side of the body after a delay of about half a second. The trunk and proximal limbs are affected more often than the face or distal limbs. Early reports were those of Allen (1928) and Bender et al. (1949); in the former, when an object was placed in the patient's left hand following the removal of a large meningioma from the posterior Rolandic area of the left cortex, another similar object was felt spontaneously in the right hand; this disappeared 1 week after operation.

Tactile alloaesthesia may be seen in the acute stage of right putaminal haemorrhage (but seldom in right thalamic haemorrhage) and occasionally with anterolateral spinal cord lesions. The author has seen a patient report sensation below the stump of an amputated leg following stimulation of the contralateral remaining leg, a phenomenon which might be termed "phantom alloaesthesia". "Mirror pain", which has been reported after percutaneous cordotomy interrupting spinothalamic tracts to alleviate refractory pain syndromes (ML Sharma, personal communication), may share a similar neurobiological substrate. The mechanism of alloaesthesia is uncertain: some consider it a disturbance within sensory pathways, others that it is a sensory response to neglect.

Visual alloaesthesia, the illusory transposition of an object seen in one visual field to the contralateral visual field, is also described, for example in "top of the basilar" syndrome or with occipital lobe tumours.

References
Allison RS. The senile brain. A clinical study. London: Edward Arnold; 1962. p. 24,57,65.
Kasten E, Poggel DA. A mirror in the mind: a case of visual allaesthesia in homonymous hemianopia. *Neurocase*. 2006; **12**: 98–106.
Kawamura M, Hirayama K, Shinohara Y, Watanabe Y, Sugishita M. Alloaesthesia. *Brain*. 1987; **110**: 225–36.

Cross References
Allochiria; Allokinesia, Allokinesis; Neglect

Allochiria

Allochiria is the mislocation of sensory stimuli to the corresponding half of the body or space, a term coined by Obersteiner in 1882. There is overlap with alloaesthesia, a term originally used by Stewart (1894) to describe stimuli displaced to a different point on the same extremity.

Transposition of objects may occur in patients with neglect, *e.g*. from the neglected side (usually left) to the opposite side (usually right): for example in a patient with left visuospatial neglect from a right frontoparietal haemorrhage, a figure was copied with objects from the left side transposed to the right.

Allochiria, understood to mean "right-left confusion", is reported in synaesthetes.

References
Halligan PW, Marshall JC, Wade DT. Left on the right: allochiria in a case of left visuospatial neglect. *J Neurol Neurosurg Psychiatry*. 1992; **55**: 717–9.
Meador KJ, Allen ME, Adams RJ, Loring DW. Allochiria vs allesthesia. Is there a misperception? *Arch Neurol*. 1991; **48**: 546–9.
Walsh RD, Floyd JP, Eidelman BH, Barrett KM. Balint syndrome and visual allochiria in a patient with reversible cerebral vasoconstriction syndrome. *J Neuroophthalmol*. 2012; **32**: 302–6.
Cross References
Alloaesthesia; Allokinesia, Allokinesis; Neglect; Right-left disorientation; Synaesthesia

Allodynia

Allodynia is the elicitation of pain by light mechanical stimuli (such as touch or light pressure) which do not normally provoke pain (*cf*. hyperalgesia), *i.e*. this is a positive sensory phenomenon. Examples of allodynia include the trigger points of trigeminal neuralgia, the affected skin in areas of causalgia, and some peripheral neuropathies; it may also be provoked, paradoxically, by prolonged morphine use.

Various pathogenetic mechanisms are considered possible, including sensitization (lower threshold, hyperexcitability) of peripheral cutaneous nociceptive fibres (in which neurotrophins may play a role); ephaptic transmission ("cross-talk") between large and small (nociceptive) afferent fibres; and abnormal central processing.

The treatment of neuropathic pain is typically with agents such as amitriptyline, duloxetine, gabapentin and pregabalin, or carbamazepine in the case of trigeminal neuralgia. Interruption of sympathetic outflow, for example with regional guanethidine blocks, may sometimes help, but relapse may occur.

Cross References
Hyperalgesia; Hyperpathia

Allographia

This term has been used to describe a peripheral agraphia syndrome characterized by problems spelling both words and nonwords, with case change errors such that upper and lower case letters are mixed when writing, with upper and lower case versions of the same letter sometimes superimposed on one another. Such errors may increase in frequency with word length. Sometimes cursive script is retained whilst writing the same material in upper case is impaired. These defects have been interpreted as a disturbance in selection of allographic forms in response to graphemic information outputted from the graphemic response buffer.

References
De Bastiani P, Barry C. A model of writing performance: evidence from a dysgraphic patient with an "allographic" writing disorder. *Boll Soc Ital Biol Sper*. 1985; **61**: 577–82.
Menichelli A, Rapp B, Semenza C. Allographic agraphia: a case study. *Cortex*. 2008; **44**: 861–8.
Cross Reference
Agraphia

Allokinesia, Allokinesis

Allokinesis has been used to denote a motor response in the wrong limb (*e.g*. movement of the left leg when attempting to move a paretic left arm), or transposition of the intended movement to the contralateral side; the movement may also be in the wrong direction. Others have used the term to denote a form of motor neglect, akin to alloaesthesia and allochiria in the sensory domain, relating to incorrect responses in the limb ipsilateral to a frontal lesion, also labelled disinhibition hyperkinesia.

References
Fisher CM. Neurologic fragments. I. Clinical observations in demented patients. *Neurology*. 1988; **38**: 1868–73. [at 1873].

Heilman KM, Valenstein E, Day A, Watson R. Frontal lobe neglect in monkeys. *Neurology*. 1995; **45**: 1205–10.

Cross References
Alloaesthesia; Allochiria; Neglect

Alternate Cover Test
- see COVER TESTS

Alternating Fist Closure Test
In the alternating fist closure test, patients are asked to open and close the fists alternating (*i.e.* open left, close right, and vice versa) at a comfortable rate. Patients with limb-kinetic apraxia cannot keep pace and lose track.

Cross References
Apraxia; Frontal lobe syndromes

Alternating Sequences Test
- see APRAXIA; FRONTAL LOBE SYNDROMES

Altitudinal Field Defect
Altitudinal visual field defects are horizontal hemianopias, in that they respect the horizontal meridian; they may be superior or inferior. Altitudinal field defects are characteristic of (but not exclusive to) disease in the distribution of the central retinal artery. Central vision may be preserved (macula sparing) because the blood supply of the macula often comes from the cilioretinal arteries. Recognised causes of altitudinal visual field defects include:

- *Monocular*:
  Central retinal artery occlusion (CRAO).
  Acute ischaemic optic neuropathy (AION).
  Retinal detachment.
  Choroiditis.
  Glaucoma.
  Chronic atrophic papilloedema.

- *Bilateral*:
  Sequential CRAO, AION.
  Bilateral occipital (inferior or superior calcarine cortices) lesions.

Cross References
Hemianopia; Macula sparing, Macula splitting; Quadrantanopia; Visual field defects

Amaurosis
Amaurosis describes visual loss, with the implication that this is not due to refractive error or intrinsic ocular disease. The term is most often used in the context of "amaurosis fugax", a transient monocular blindness, which is most often due to embolism from a stenotic ipsilateral internal carotid artery (ocular transient ischaemic attack). Giant cell arteritis, systemic lupus erythematosus and the anti-phospholipid antibody syndrome are also recognised causes. Gaze-evoked amaurosis has been associated with a variety of mass lesions and is thought to result from decreased blood flow to the retina from compression of the central retinal artery on eye movement.

Amblyopia
Amblyopia refers to poor visual acuity, most usually in the context of a "lazy eye", in which the poor acuity results from the failure of the eye to establish normal cortical representation of visual input during the critical period of visual maturation (between the ages of 6 months and 3 years). This may result from:

- Strabismus.
- Uncorrected refractive error.
- Stimulus deprivation.

Amblyopic eyes may demonstrate a relative afferent pupillary defect, and sometimes latent nystagmus.

Amblyopia may not become apparent until adulthood, when the patient suddenly becomes aware of unilateral poor vision. The finding of a latent strabismus (heterophoria) may be a clue to the fact that such visual loss is long-standing.

The word amblyopia has also been used in other contexts: bilateral simultaneous development of central or centrocaecal scotomas in chronic alcoholics has often been referred to as tobacco-alcohol amblyopia, although nutritional optic neuropathy is perhaps a better term.

Cross References

Esotropia; Heterophoria; Nystagmus; Relative afferent pupillary defect (RAPD); Scotoma

Amimia

- see HYPOMIMIA

Amnesia

Amnesia is an impairment of episodic memory, or memory for personally experienced events (autobiographical memory). This is a component of long-term (as opposed to working) memory which is distinct from memory for facts (semantic memory), in that episodic memory is unique to the individual whereas semantic memory encompasses knowledge held in common by members of a cultural or linguistic group. Episodic memory generally accords with the lay perception of memory, although many complaints of "poor memory" represent faulty attentional mechanisms rather than true amnesia. A precise clinical definition for amnesia has not been demarcated, perhaps reflecting the heterogeneity of the syndrome.

Amnesia may be retrograde (for events already experienced) or anterograde (for newly experienced events). Retrograde amnesia may show a temporal gradient, with distant events being better recalled than more recent ones, relating to the duration of anterograde amnesia.

Amnesia may be acute and transient or chronic and persistent. In a pure amnesic syndrome, intelligence and attention are normal and skill acquisition (procedural memory) is preserved. Amnesia may occur as one feature of more widespread cognitive impairments, *e.g.* in Alzheimer's disease.

Various psychometric tests of episodic memory are available. These include the Wechsler Memory Score (WMS-R), the Recognition Memory Test which has both verbal (words) and visual (faces) subdivisions, the Rey Auditory Verbal Learning Test (immediate and delayed free recall of a random word list), and the Rey-Osterrieth Complex Figure (non-verbal memory). Retrograde memory may be assessed with a structured Autobiographical Memory Interview, and with the Famous Faces Test. Poor spontaneous recall, for example of a word list, despite an adequate learning curve, may be due to a defect in either storage or retrieval. This may be further probed with cues: if this improves recall, then a disorder of retrieval is responsible; if cueing leads to no improvement, or false-positive responses to foils (as in the Hopkins Verbal Learning Test) are equal or greater than true positives, then a learning defect (true amnesia) is the cause.

The neuroanatomical substrate of episodic memory is a distributed system in the medial temporal lobe and diencephalon surrounding the third ventricle (the circuit of Papez) comprising the entorhinal area of the parahippocampal gyrus, perforant and alvear pathways, hippocampus, fimbria and fornix, mammillary bodies, mammillothalamic tract, anterior thalamic nuclei, internal capsule, cingulate gyrus, and cingulum. Basal forebrain structures (septal nucleus, diagonal band nucleus of Broca, nucleus basalis of Meynert) are also involved.

Classification of amnesic syndromes into subtypes has been proposed, since lesions in different areas produce different deficits reflecting functional subdivision within the system; thus left temporal lesions produce problems in the verbal domain, right sided lesions affect non-verbal/visual memory. A distinction between medial temporal pathology (*e.g.* hippocampus), leading to difficulty encoding new memories (anterograde amnesia and temporally limited retrograde amnesia), and diencephalic pathology (*e.g.* Korsakoff's syndrome), which causes difficulty retrieving previously acquired memories (extensive retrograde amnesia) with diminished insight and a tendency to confabulation, has been suggested, but overlap may occur. A frontal amnesia has also been suggested, although impaired attentional mechanisms may contribute. Functional imaging studies suggest medial temporal lobe activation is required for encoding with additional prefrontal activation with "deep" processing; medial temporal and prefrontal activation are also seen with retrieval.

Many causes of amnesia are recognised, including:

- Acute/transient:
  - Closed head injury.
  - Drugs.
  - Transient global amnesia.
  - Transient epileptic amnesia.
  - Transient semantic amnesia (very rare).
- Chronic/persistent:
  - Alzheimer's disease (may show isolated amnesia in early disease).
  - Sequela of herpes simplex encephalitis.
  - Limbic encephalitis (paraneoplastic or non-paraneoplastic).
  - Hypoxic brain injury.
  - Temporal lobectomy (bilateral; or unilateral with previous contralateral injury, usually birth asphyxia).
  - Bilateral posterior cerebral artery occlusion.
  - Korsakoff's syndrome.
  - Bilateral thalamic infarction.
  - Third ventricle tumour, cyst.
  - Focal retrograde amnesia (rare).

Few of the chronic persistent causes of amnesia are amenable to specific treatment. Plasma exchange or intravenous immunoglobulin therapy may be helpful in autoimmune limbic encephalitides, for example associated with autoantibodies directed against voltage-gated potassium channels.

Functional or psychogenic or dissociative amnesia may involve failure to recall basic autobiographical details such as name and address. Reversal of the usual temporal gradient of memory loss may be observed (but this may also be the case in the syndrome of focal retrograde amnesia).

References

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Brandt J, Van Gorp WG. Functional ("psychogenic") amnesia. *Semin Neurol*. 2006; **26**: 331–40.

Kopelman MD. Disorders of memory. *Brain*. 2002; **125**: 2152–90.

Papanicolaou AC, editor. The amnesias. A clinical textbook of memory disorders. New York: Oxford University Press; 2006.

Cross References

Confabulation; Dementia; Dissociation

Amphigory

Miller Fisher used this term to describe nonsense speech.

References

Fisher CM. Nonsense speech – amphigory. *Trans Am Neurol Assoc*. 1970; **95**: 238–40.
Fisher CM. Neurologic fragments. I. Clinical observations in demented patients. *Neurology*. 1988; **38**: 1868–73. [at 1872].

Cross Reference

Aphasia

Amusia

Amusia is a loss of the ability to appreciate music despite normal intelligence, memory and language function. Subtypes have been described: receptive or sensory amusia is loss of the ability to appreciate music; and expressive or motor amusia is loss of ability to sing, whistle, etc. Clearly a premorbid appreciation of music is a *sine qua non* for the diagnosis (particularly of the former), and most reported cases of amusia have occurred in trained musicians. Others have estimated that amusia affects up to 4% of the population (presumably expressive; = "tone deafness"). Tests for the evaluation of amusia have been described.

Amusia may occur in the context of more widespread cognitive dysfunction, such as aphasia and agnosia, although aphasia without amusia has been reported. Amusia has been reported in association with pure word deafness, presumably as part of a global auditory agnosia. Isolated amusia has been reported in the context of focal cerebral atrophy affecting the non-dominant temporal lobe. However, functional studies have failed to show strong hemispheric specificity for music perception, but suggest a cross-hemispheric distributed neural substrate. An impairment of pitch processing with preserved awareness of musical rhythm changes has been described in amusics.

Reference

Clark CN, Golden HL, Warren JD. Acquired amusia. *Handb Clin Neurol*. 2015; **129**: 607–31.
Fisher CA, Larner AJ. Jean Langlais (1907–91): an historical case of a blind organist with stroke-induced aphasia and Braille alexia but without amusia. *J Med Biogr*. 2008; **16**: 232–4.

Cross References

Agnosia; Auditory agnosia; Pure word deafness

Amyotrophy

Amyotrophy is a term used to describe thinning or wasting (atrophy) of musculature with attendant weakness. This may result from involvement of:

- Lower motor neurones (in which case fasciculations may also be present):
  - Motor neurone disease/Amyotrophic lateral sclerosis.
  - Benign focal amyotrophy/monomelic amyotrophy.
  - Disinhibition-dementia-parkinsonism-amyotrophy complex (DDPAC).
  - Amyotrophic Creutzfeldt-Jakob disease (obsolete term).
  - "Asthmatic amyotrophy" (Hopkins' syndrome).
- Nerve roots:
  - Diabetic amyotrophy (polyradiculopathy, especially L2-L4).
- Plexus
  - Neuralgic amyotrophy (Parsonage-Turner syndrome).

Hence although the term implies neurogenic (as opposed to myogenic) muscle wasting, its use is non-specific with respect to neuroanatomical substrate.

Cross References

Atrophy; Fasciculation; Neuropathy; Plexopathy; Radiculopathy; Wasting

Anaesthesia

Anaesthesia (anesthesia) is a complete loss of sensation; hypoaesthesia (hypaesthesia, hypesthesia) is a diminution of sensation. Hence in Jacksonian terms, these are negative sensory phenomena. Anaesthesia may involve all sensory modalities (global anaesthesia, as in general surgical anaesthesia) or be selective (*e.g.* thermoanaesthesia, analgesia). Regional patterns of anaesthesia are described, *e.g.* "glove-and-stocking anaesthesia" in peripheral neuropathies, "saddle anaesthesia" involving S3-5 dermatomes resulting from a cauda equina syndrome.

Anaesthesia is most often encountered after resection or lysis of a peripheral nerve segment, whereas paraesthesia or dysaesthesia (positive sensory phenomena) reflect damage to a nerve which is still in contact with the cell body.

Anaesthesia dolorosa, or painful anaesthesia, is a persistent unpleasant pain (*i.e.* a positive sensory phenomenon) which may be experienced in the distribution of a resected nerve, *e.g.* following neurolytic treatment for trigeminal neuralgia, usually with delayed onset. This deafferentation pain may respond to various medications, including tricyclic antidepressants, carbamazepine, gabapentin, pregabalin, and selective serotonin reuptake inhibitors.

Cross References

Analgesia; Dysaesthesia; Neuropathy; Paraesthesia

Analgesia

Analgesia or hypoalgesia refers to a complete loss or diminution, respectively, of pain sensation, or the absence of a pain response to a normally painful stimulus. These negative sensory phenomena may occur as one component of total sensory loss (anaesthesia) or in isolation. Consequences of analgesia include the development of neuropathic ulcers, burns, Charcot joints, even painless mutilation or amputation. Analgesia may occur in:

- Peripheral nerve lesions, *e.g.* hereditary sensory and autonomic neuropathies (HSAN), leprosy.
- Central spinal cord lesions which pick off the decussating fibres of the spinothalamic pathway in the ventral funiculus (with corresponding thermoanaesthesia), *e.g.* syringomyelia.
- Cortical lesions, *e.g.* medial frontal lobe syndrome (akinetic type).

Congenital syndromes of insensitivity to pain were once regarded as a central pain asymbolia (*e.g.* Osuntokun's syndrome), but on further follow-up some have turned out to be variants of HSAN.

Reference

Larner AJ, Moss J, Rossi ML, Anderson M. Congenital insensitivity to pain: a 20 year follow up. *J Neurol Neurosurg Psychiatry*. 1994; **57**: 973–4.

Cross References

Anaesthesia; Frontal lobe syndromes

Anal Reflex

Contraction of the external sphincter ani muscle in response to a scratch stimulus in the perianal region, testing the integrity of the S4/S5 roots, forms the anal or wink reflex. This reflex may be absent in some normal elderly individuals, and absence does not necessarily correlate with urinary incontinence. External anal responses to coughing and sniffing are part of a highly consistent and easily elicited polysynaptic reflex, whose characteristics resemble those of the conventional scratch-induced anal reflex.

Reference

Chan CLH, Ponsford S, Swash M. The anal reflex elicited by cough and sniff: validation of a neglected clinical sign. *J Neurol Neurosurg Psychiatry*. 2004; **75**: 1449–51.

Cross References

Reflexes; Urinary incontinence

Anarchic Hand
- see ALIEN HAND, ALIEN LIMB

Anarithmetia
- see ACALCULIA

Anarthria
Anarthria is the complete inability to articulate words (*cf*. dysarthria). This is most commonly seen as a feature of the bulbar palsy of motor neurone disease.

A motor disorder of speech production with preserved comprehension of spoken and written language has been termed pure anarthria; this syndrome has also been labelled at various times and by various authors as aphemia, phonetic disintegration, apraxic dysarthria, cortical dysarthria, verbal apraxia, subcortical motor aphasia, pure motor aphasia, and small or mini Broca's aphasia. It reflects damage in the left frontal operculum, but with sparing of Broca's area. A pure progressive anarthria or slowly progressive anarthria may result from focal degeneration affecting the frontal operculum bilaterally (so-called Foix-Chavany-Marie syndrome).

References

Broussolle E, Bakchine S, Tommasi M, et al. Slowly progressive anarthria with late anterior opercular syndrome: a variant form of frontal cortical atrophy syndromes. *J Neurol Sci*. 1996; **144**: 44–58.

Lecours AR, Lhermitte F. The "pure" form of the phonetic disintegration syndrome (pure anarthria): anatomo-clinical report of a single case. *Brain Lang*. 1976; **3**: 88–113.

Cross References

Aphemia; Bulbar palsy; Dysarthria

Angioscotoma
Angioscotomata are shadow images of the superficial retinal vessels on the underlying retina, a physiological scotoma.

Cross Reference

Scotoma

Angor Animi
Angor animi describes the sense of dying or the feeling of impending death. It may be experienced on awakening from sleep, or as a somesthetic aura of migraine.

Reference

Ryle JA. Angor animi, or the sense of dying. *Guys Hosp Rep*. 1950; **99**: 230–5.

Cross Reference

Aura

Anhidrosis
Anhidrosis, or hypohidrosis, is a loss or lack of sweating. This may be due to primary autonomic failure, or to pathology within the posterior hypothalamus ("sympathetic area").

Anhidrosis may occur in various neurological disorders, including multiple system atrophy, Parkinson's disease, multiple sclerosis, caudal to a spinal cord lesion, and in some hereditary sensory and autonomic neuropathies. Localised or generalised anhidrosis may be seen in Holmes-Adie syndrome, and unilateral anhidrosis may be seen in Horner's syndrome if the symptomatic lesion is distal to the superior cervical ganglion.

Cross References

Dysautonomia; Holmes-Adie pupil, Holmes-Adie syndrome; Horner's syndrome; Hyperhidrosis

Anismus
Anismus, also known as puborectalis syndrome, is paradoxical contraction of the external anal sphincter during attempted defaecation, leading to faecal retention and a complaint of constipation. This may occur as an idiopathic condition in isolation, or as a feature of the off

periods of idiopathic Parkinson's disease. It is thought to represent a focal dystonia, and may be helped temporarily by local injections of botulinum toxin.

Reference

Ron Y, Avni Y, Lukovetski A, et al. Botulinum toxin type-A in therapy of patients with anismus. *Dis Colon Rectum*. 2001; **44**: 1821–6.

Cross References

Dystonia; Parkinsonism

Anisocoria

Anisocoria describes an inequality of pupil size. This may be physiological (said to occur in up to 15% of the population), in which case the inequality is usually mild and does not vary with degree of ambient illumination; or pathological, with many possible causes.

- Structural:

  Ocular infection, trauma, inflammation, surgery.

- Neurological:

  Anisocoria greater in dim light or darkness suggests a sympathetic innervation defect (darkness stimulates dilatation of the normal pupil). The affected pupil is constricted (miosis; oculosympathetic paresis), as in:

  Horner's syndrome.
  Argyll Robertson pupil.
  Cluster headache.

  Anisocoria greater in bright light/less in dim light suggests a defect in parasympathetic innervation to the pupil. The affected pupil is dilated (mydriasis; oculoparasympathetic paresis), as in:

  Holmes-Adie pupil (vermiform movements of the pupil margin may be visible with a slit-lamp).
  Oculomotor (III) nerve palsy (efferent path from Edinger-Westphal nucleus).
  Mydriatic agents (phenylephrine, tropicamide).
  Anticholinergic agents (*e.g.* asthma inhaler accidentally puffed into one eye).

Clinical characteristics and pharmacological testing may help to establish the underlying diagnosis in anisocoria.

Reference

Bremner FD, Smith SE. Pupil abnormality in autonomic disorders. In: Mathias CJ, Bannister R, editors. Autonomic failure. A textbook of clinical disorders of the autonomic nervous system. 5th ed. Oxford: Oxford University Press; 2013. p. 445–53.

Cross References

Argyll Robertson pupil; Holmes-Adie pupil, Holmes-Adie syndrome; Horner's syndrome; Miosis; Mydriasis

Ankle Jerk, Ankle Reflex

Plantar flexion at the ankle following phasic stretch of the Achilles tendon constitutes the ankle jerk or Achilles (tendon) reflex, mediated through sacral segments S1 and S2 and the sciatic and posterior tibial nerves.

This reflex may be elicited in several ways: by a blow with a tendon hammer directly upon the Achilles tendon (patient supine, or prone with knees flexed, or kneeling) or with a direct plantar strike. The latter, though convenient and quick, is probably the least sensitive method, since absence of an observed muscle contraction does not mean that the reflex is absent; the latter methods are more sensitive, although intra- and interobserver agreement may be better with the plantar strike.

The ankle jerk is typically lost in polyneuropathies and in S1 radiculopathy. Loss of the ankle jerk is increasingly prevalent with normal healthy ageing, beyond the age of 60 years,

although more than 65% of patients are said to retain their ankle jerks; this observation may depend in part on the method of assessment used.

References

O'Keeffe ST, Smith T, Valacio R, Jack CI, Playfer JR, Lye M. A comparison of two techniques for ankle jerk assessment in elderly subjects. *Lancet*. 1994; **344**: 1619–20.

Ross RT. How to examine the nervous system. 4th ed. Totawa: Humana Press; 2006. p. 169–71.

Vrancken AFJE, Kalmijn S, Brugman F, Rinkel GJE, Notermans NC. The meaning of distal sensory loss and absent ankle reflexes in relation to age. A meta-analysis. *J Neurol*. 2006; **253**: 578–89.

Cross References

Age-related signs; Neuropathy; Reflexes

Annular Scotoma

An annular or ring scotoma suggests retinal disease, as in retinitis pigmentosa or cancer-associated retinopathy (paraneoplastic retinal degeneration).

Cross References

Retinopathy; Scotoma; Visual field defects

Anomia

Anomia, or dysnomia, is a deficit in naming or word-finding. This may be detected as abrupt cut-offs in spontaneous speech with circumlocutions and/or paraphasic substitutions. Formal tests of naming are also available (*e.g.* Graded Naming Test). Patients may be able to point to named objects despite being unable to name them, suggesting a problem in word retrieval but with preserved comprehension. They may also be able to say something about the objects they cannot name (*e.g.* "flies in the sky" for kite) suggesting preserved access to the semantic system.

Category-specific anomias have been described, *e.g.* for colour (*cf.* achromatopsia).

Anomia occurs with pathologies affecting the left temporoparietal area, but since it occurs in all varieties of aphasia is of little precise localizing or diagnostic value. The term anomic aphasia is reserved for unusual cases in which a naming problem overshadows all other deficits. Anomia may often be seen as a residual deficit following recovery from other types of aphasia. Anomia may occur with any dominant hemisphere space-occupying lesion, and as a feature of semantic dementia, being more prominent in this condition than in Alzheimer's disease.

References

Benson DF, Ardila A. Aphasia: a clinical perspective. New York: Oxford University Press; 1996. p. 252–61.

Woollams AM, Cooper-Pye E, Hodges JR, Patterson K. Anomia: a doubly typical signature of semantic dementia. *Neuropsychologia*. 2008; **46**: 2503–14.

Cross References

Aphasia; Circumlocution; Paraphasia

Anopsognosia

- see AGNOSOPSIA

Anosmia

Anosmia is the inability to perceive smells due to damage to the olfactory pathways (olfactory neuroepithelium, olfactory nerves, rhinencephalon). Olfaction may be tested with kits containing specific odours (*e.g.* clove, turpentine); each nostril should be separately tested. Unilateral anosmia may be due to pressure on the olfactory bulb or tract, *e.g.* from a subfrontal meningioma.

Anosmia may be congenital (*e.g.* Kallman's syndrome, hypogonadotrophic hypogonadism, a disorder of neuronal migration) or, much more commonly, acquired. Rhinological

disease (allergic rhinitis, coryza) is by far the most common cause; this may also account for the impaired sense of smell in smokers. Head trauma is the most common neurological cause, due to shearing off of the olfactory fibres as they pass through the cribriform plate. Recovery is possible in this situation due to the capacity for neuronal and axonal regeneration within the olfactory pathways. Olfactory dysfunction is also described in Alzheimer's disease and Parkinson's disease, possibly as an early phenomenon, due to pathological involvement of olfactory pathways. Patients with depression may also complain of impaired sense of smell. Loss of olfactory acuity may be a feature of normal ageing.

Reference

Hawkes CH, Doty RL. The neurology of olfaction. Cambridge: Cambridge University Press; 2009.

Cross References

Age-related signs; Ageusia; Cacosmia; Dysgeusia; Mirror movements; Parosmia

Anosodiaphoria

Babinski (1914) used the term anosodiaphoria to describe a disorder of body schema in which patients verbally acknowledge a clinical problem (*e.g*. hemiparesis) but fail to be concerned by it. Anosodiaphoria usually follows a stage of anosognosia.

La belle indifférence describes a similar lack of concern for acknowledged disabilities which are psychogenic.

References

Babinski JM. Contribution à l'étude des troubles mentaux dans l'hémiplégie organique cérébrale (anosognosie). *Rev Neurol*. 1914; **27**: 845–8.

Critchley M. Observations on anosodiaphoria. *L'Encéphalie*. 1957; **46**: 540–6.

Cross References

Anosognosia; *Belle indifférence*; Personification of paralysed limbs

Anosognosia

Anosognosia refers to a patient's unawareness or denial of their illness. The term was first used by von Monakow (1885) and has been used to describe denial of blindness (Anton's syndrome), deafness, hemiplegia (by Babinski), hemianopia, aphasia, and amnesia. Some authorities would question whether this unawareness is a true agnosia, or rather a defect of higher level cognitive integration (*i.e*. perception).

Anosognosia with hemiplegia most commonly follows right hemisphere injury (parietal and temporal lobes) and may be associated with left hemineglect and left-sided hemianopia; it is also described with right thalamic and basal ganglia lesions. Many patients with posterior aphasia (Wernicke type) are unaware that their output is incomprehensible or jargon, possibly through a failure to monitor their own output. Cerebrovascular disease is the most common pathology associated with anosognosia, although it may also occur with neurodegenerative disease, for example the cognitive anosognosia in some patients with Alzheimer's disease (often interpreted by relatives as the patient being "in denial").

The neuropsychological mechanisms of anosognosia are unclear: the hypothesis that it might be accounted for by personal neglect (asomatognosia), which is also more frequently observed after right hemisphere lesions, would seem to have been disproved experimentally by studies using selective hemisphere anaesthesia in which the two may be dissociated, a dissociation which may also be observed clinically. In Alzheimer's disease, anosognosia may be related to memory dysfunction and executive dysfunction

At a practical level, anosognosia may lead to profound difficulties with neurorehabilitation. Temporary resolution of anosognosia has been reported following vestibular stimulation (*e.g*. with caloric testing).

References

Adair JC, Schwartz RL, Barrett AM. Anosognosia. In: Heilman KM, Valenstein E, editors. Clinical neuropsychology. 4th ed. Oxford: Oxford University Press; 2003. p. 185–214.

Babinski JM. Contribution à l'étude des troubles mentaux dans l'hémiplégie organique cérébrale (anosognosie). *Rev Neurol*. 1914; **27**: 845–8.
Prigatano GP, editor. The study of anosognosia. Oxford: Oxford University Press; 2010.
Rosen HJ. Anosognosia in neurodegenerative disease. *Neurocase*. 2011; **17**: 231–41.
Cross References
Agnosia; Anosodiaphoria; Asomatognosia; Cortical blindness; Extinction; Jargon aphasia; Misoplegia; Neglect; Personification of paralysed limbs; Somatoparaphrenia

Anserina
Autonomically mediated piloerection and thermoconstriction may produce "goosebumps", cold and bumpy skin which may be likened to that of a plucked goose. Loss of anserina may be a feature of some autonomic disorders.

Antecollis
Antecollis (or anterocollis) is forward flexion of the neck. It may be a feature of multiple system atrophy (*cf*. retrocollis in progressive supranuclear palsy), a sustained dystonic posture in advanced Parkinson's disease, and, unusually, in spasmodic torticollis.

Forward flexion of the head onto the chest is a feature in the "dropped head syndrome".
Reference
Quinn N. Disproportionate antecollis in multiple system atrophy. *Lancet*. 1989; **1**: 844.
Cross References
Dropped head syndrome; Retrocollis; Torticollis

Anteflexion
Anteflexion is forward flexion of the trunk, as typical of the stooped posture seen in Parkinson's disease.
Cross Reference
Parkinsonism

Anton's Syndrome
Anton's syndrome refers to cortical blindness accompanied by denial of the visual defect (visual anosognosia), with or without confabulation. The syndrome most usually results from bilateral posterior cerebral artery territory lesions causing occipital or occipitoparietal infarctions, but has occasionally been described with anterior visual pathway lesions associated with frontal lobe lesions. It may also occur in the context of dementing disorders or delirium.
References
Abutalebi J, Arcari C, Rocca MA, et al. Anton's syndrome following callosal disconnection. *Behav Neurol*. 2007; **18**: 183–6.
Gassel M, Williams D. Visual function in patients with homonymous hemianopia. III. The completion phenomenon: insight and attitude to the defect: and visual function efficiency. *Brain*. 1963; **86**: 229–60.
Zukic S, Sinanovic O, Zonic L, et al. Anton's syndrome due to bilateral ischemic occipital lobe strokes. *Case Rep Neurol Med*. 2014; **2014**: 474952.
Cross References
Agnosia, Anosognosia, Confabulation, Cortical blindness

Anwesenheit
A vivid sensation of the presence of somebody either somewhere in the room or behind the patient has been labelled as *anwesenheit* (German: presence), presence hallucination, minor hallucination, or extracampine hallucination. This phenomenon is relatively common in Parkinson's disease, occurring in isolation or associated with formed visual hallucinations.

References
Chan D, Rossor MN. "- but who is that on the other side of you?" Extracampine hallucinations revisited. *Lancet*. 2002; **360**: 2064–6.
Fénélon G, Mahieux F, Huon R, Ziegler M. Hallucinations in Parkinson's disease: prevalence, phenomenology and risk factors. *Brain*. 2000; **123**: 733–45.
Cross References
Hallucination; Parkinsonism

Apallic Syndrome
- see VEGETATIVE STATES

Apathy
Apathy is a common neurobehavioural symptom which may be characterized by lack of motivation relative to the patient's previous level of functioning or the standards of age and culture. This manifests as diminished goal-directed behaviour (lack of effort, dependency on others to structure activity), diminished goal-directed cognition (lack of interest, or of concern about personal problems), and diminished concomitants of goal-directed behaviour (unchanging affect, lack of emotional responsiveness). Various scales and inventories are available to measure apathy.

Listlessness, paucity of spontaneous movement (akinesia) or speech (mutism), and lack of initiative, spontaneity and drive, may all be features of apathy. These are also features of the abulic state, and it has been suggested that apathy and abulia represent different points on a continuum of motivational and emotional deficit, abulia being at the more severe end. The diminished motivation of apathy should not be attributable to impaired level of consciousness, emotional distress, or cognitive impairment although it may coexist with the latter, as in Alzheimer's disease. Apathy is a specific neuropsychiatric syndrome, distinct from depression.

Apathy may be observed in various diseases affecting frontal-subcortical structures, for example in the frontal lobe syndrome affecting the frontal convexity, or following multiple vascular insults to paramedian diencephalic structures (thalamus, subthalamus, posterior lateral hypothalamus, mesencephalon) or the posterior limb of the internal capsule; there may be associated cognitive impairment of the so-called "subcortical" type in these situations (*e.g.* in Huntington's disease). Apathy is also extremely common in Alzheimer's disease. It is also described following amphetamine or cocaine withdrawal, in neuroleptic-induced akinesia, and in psychotic depression and schizophrenia.

Because apathy may reflect reward insensitivity, dopaminergic mechanisms may play a role in the pathophysiology, suggesting that dopaminergic agents may sometimes be helpful. Selective serotonin reuptake inhibitors have also been tried.

References
Bonnelle V, Veromann KR, Burnett Heyes S, et al. Characterization of reward and effort mechanisms in apathy. *J Physiol Paris*. 2015; **109**: 16–26.
Levy ML, Cummings JL, Fairbanks LA, et al. Apathy is not depression. *J Neuropsychiatry Clin Neurosci*. 1998; **10**: 314–9.
Levy R, Dubois B. Apathy and the functional anatomy of the prefrontal cortex-basal ganglia circuits. *Cereb Cortex*. 2006; **16**: 916–28.
Stella F, Radanovic M, Aprahamian I, et al. Neurobiological correlates of apathy in Alzheimer's disease and mild cognitive impairment: a critical review. *J Alzheimers Dis*. 2014; **39**: 633–48.
Thobois S, Lhommee E, Klinger H, et al. Parkinsonian apathy responds to dopaminergic stimulation of D2/D3 receptors with piribedil. *Brain*. 2013; **136**: 1568–77.
Cross References
Abulia; Akinetic mutism; Dementia; Frontal lobe syndromes

Aphantasia
This term has been coined to describe a lack of visual imagery.

Reference
Zeman A, Dewar M, Della Sala S. Lives without imagery – congenital aphantasia. *Cortex*. 2015; **73**: 378–80.

Aphasia

Aphasia, or dysphasia, is an acquired loss or impairment of language function. Language may be defined as the complex system of symbols used for communication (including reading and writing), encompassing various linguistic components (phonetic, phonemic, semantic/lexical, syntactic, pragmatic), all of which are dependent on dominant hemisphere integrity. Non-linguistic components of language (emotion, inflection, cadence), collectively known as prosody, may require contributions from both hemispheres.

Language is distinguished from speech (oral communication), disorders of which are termed dysarthria or anarthria. Dysarthria and aphasia may co-exist but are usually separable.

Clinical assessment of aphasia requires analysis of the following features, through listening to the patient's spontaneous speech, asking questions or giving commands, and asking the patient to repeat, name, read and write:

- *Fluency*: is output effortful, laboured, with agrammatism and dysprosody (non-fluent); or flowing, with paraphasias and neologisms (fluent)?
- *Comprehension*: spared or impaired?
- *Repetition*: preserved or impaired?
- *Naming*: preserved or impaired?
- *Reading*: evidence of alexia?
- *Writing*: evidence of agraphia?

These features allow definition of various types of aphasia (see Table and specific entries; although it should be noted that some distinguished neurologists, such as Macdonald Critchley, have taken the view that no satisfactory classification of the aphasias exists). For example, motor ("expressive") aphasias are characterized by non-fluent verbal output, with intact or largely unimpaired comprehension, whereas sensory ("receptive") aphasias demonstrate fluent verbal output, often with paraphasias, sometimes jargon, with impaired comprehension. Conduction aphasia is marked by relatively normal spontaneous speech (perhaps with some paraphasic errors) but a profound deficit of repetition. In transcortical motor aphasia spontaneous output is impaired but repetition is intact.

	Broca	Wernicke	Conduction	Transcortical: motor/sensory
Fluency	↓↓	N	N	↓/N
Comprehension	N	↓↓	N	N/↓
Repetition	↓	↓	↓↓	N/N
Naming	↓	↓	↓	N?/N?
Reading	↓	↓	↓	N?/N?
Writing	↓	↓	↓	N?/N?

Aphasia most commonly follows a cerebrovascular event: the specific type of aphasia may change with time following the event, and discrepancies may be observed between classically defined clinicoanatomical syndromes and the findings of everyday practice.

Aphasia may also occur with space-occupying lesions and in neurodegenerative disorders, often with other cognitive impairments (*e.g.* Alzheimer's disease) but sometimes in isolation. The classification of these primary progressive aphasias (PPA) is still in flux, but broadly they may be divided into primary non-fluent aphasia (agrammatic variant of PPA) and semantic dementia (semantic variant of PPA), both of which usually have a pathological substrate of one of the frontotemporal dementias; and logopenic variant of PPA which most often has the pathological substrate of Alzheimer's disease.

References
Basso A. Aphasia and its therapy. Oxford: Oxford University Press; 2003.
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Caplan D. Aphasic syndromes. In: Heilman KM, Valenstein E, editors. Clinical neuropsychology. 4th ed. Oxford: Oxford University Press; 2003. p. 14–34.
Critchley M. The citadel of the senses and other essays. New York: Raven Press; 1986. p. 96.
LaPointe LL. Aphasia and related neurogenic language disorders. 4th ed. New York: Thieme; 2011.
Spreen O, Risser AH. Assessment of aphasia. Oxford: Oxford University Press; 2003.
Willmes K, Poeck K. To what extent can aphasic syndromes be localized?. *Brain*. 1993; **116**: 1527–40.
Cross References
Agrammatism; Agraphia; Alexia; Anomia; Aprosodia, Aprosody; Broca's aphasia; Circumlocution; Conduction aphasia; *Conduit d'approche*; Crossed aphasia; Dynamic aphasia; Dysarthria; Dysphasia; Jargon aphasia; Neologism; Optic aphasia; Paraphasia; Transcortical aphasias; Wernicke's aphasia

Aphemia

Aphemia was the name originally given by Paul Broca to the language disorder which he observed in his celebrated case, prior to Armand Trousseau's suggestion that the term aphasia was preferable on philological grounds (hence the name "Broca's aphasia" for this phenotype).

The term aphemia is now used to describe a motor disorder of speech production with elements including dysarthria, orofacial apraxia, dysprosody, phonetic and phonemic errors but preserved comprehension of spoken and written language and otherwise preserved cognition. This syndrome, or something akin to it, has also been called phonetic disintegration (*cf*. phonemic disintegration), pure anarthria, apraxic dysarthria, cortical dysarthria, verbal apraxia, subcortical motor aphasia, alalia, pure motor aphasia, small or mini Broca's aphasia, and kinetic speech production disorder, reflecting the differing views as to the nature of the underlying disorder (aphasia, dysarthria, apraxia).

Aphemia probably also encompasses at least some cases of the "foreign accent syndrome", in which altered speech production and/or prosody makes speech output sound foreign to the speaker's native tongue. Such conditions may stand between pure disorders of speech (*i.e*. dysarthrias) and of language (*i.e*. aphasias). They usually reflect damage in the left frontal operculum, but sparing Broca's area. Slowly progressive aphemia may be associated with a left frontal lesion, often affecting the opercular region (*cf*. speech apraxia).

References
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Henderson VW. Alalia, aphemia, and aphasia. *Arch Neurol*. 1990; **47**: 85–8.
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Schiff HB, Alexander MP, Naeser MA, Galaburda AM. Aphemia: clinical-anatomic correlations. *Arch Neurol*. 1983; **40**: 720–7.
Cross References
Anarthria; Aphasia; Aprosodia, Aprosody; Dysarthria; Foreign accent syndrome; Phonemic disintegration; Speech apraxia

Aphonia

Aphonia is loss or diminution of vocal sound volume, necessitating mouthing or whispering of words. As for dysphonia, this most frequently follows laryngeal inflammation, although it may follow bilateral recurrent laryngeal nerve palsy. Dystonia of the abductor muscles of the larynx can result in aphonic segments of speech (spasmodic aphonia, or abductor laryngeal dystonia); this may be diagnosed by hearing the voice fade away to nothing when asking the

patient to keep talking, and patients may volunteer that they cannot hold any prolonged conversation. Aphonia of functional or hysterical origin is also recognised.

Aphonia should be differentiated from mutism, in which patients make no effort to speak, and anarthria in which there is a failure of articulation.

Cross References

Anarthria; Dysphonia; Mutism

Aposiopesis

Critchely used this term to denote a sentence which is started but not finished, as in the aphasia associated with dementia.

Reference

Critchley M. The divine banquet of the brain and other essays. New York: Raven Press; 1979. p. 48.

Cross Reference

Aphasia

Applause Sign

The applause sign, also known as the *signe d'applause*, clapping test or three clap test, is elicited by instructing the patient to clap the hands rapidly three times (the examiner may demonstrate). The tendency to clap more than three times, even when demonstrated by the examiner, is judged abnormal.

The applause sign was first reported (2005) in patients with progressive supranuclear palsy (PSP) and later in other parkinsonian disorders such as Parkinson's disease, dementia with Lewy bodies (DLB), corticobasal degeneration, and multiple system atrophy, consistent with basal ganglia pathology (striatal dysfunction), but it has also been reported in cortical dementias, namely Alzheimer's disease (AD) and frontotemporal lobar degenerations (FTLD), as well as in PSP. In AD, applause sign correlates with frontal lobe dysfunction, hence this may be a motor perseveration.

In consecutive patients attending a cognitive disorders clinic, the applause sign was found to be specific (0.89) but not sensitive (0.36) for identification of any cognitive impairment, and hence it may be useful as a non-canonical sign of cognitive impairment in high prevalence settings.

References

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Cross References

"Attended alone" sign; "Head turning sign"

Apraxia

Apraxia or dyspraxia is a disorder of movement characterized by the inability to perform a voluntary motor act despite an intact motor system (*i.e.* no ataxia, weakness) and without impairment in level of consciousness. Automatic/reflex actions are preserved, hence there is a voluntary-automatic dissociation; some authors see this as critical to the definition of apraxia. Different types of apraxia have been delineated, the standard classification being that of Liepmann (1900):

- *Ideational apraxia*, *conceptual apraxia*:

  A deficit in the conception of a movement; this frequently interferes with daily motor activities and is not facilitated by the use of objects; there is often an associated aphasia.

- *Ideomotor apraxia* (*IMA*):

  A disturbance in the selection of elements that constitute a movement (*e.g.* pantomiming the use of tools); in contrast to ideational apraxia, this is a "clinical" disorder inasmuch as it does not greatly interfere with everyday activities; moreover, use of objects may facilitate movement; it may often be manifest as the phenomenon of using body part as object, *e.g.* in demonstrating how to use a toothbrush or how to hammer a nail, a body part is used to represent the object (finger used as toothbrush, fist as hammer).

- *Limb-kinetic*, *or melokinetic*, *apraxia*:

  Slowness, clumsiness, awkwardness in using a limb, with a temporal decomposition of movement; it usually coexists with ideomotor apraxia but may be differentiated from it as more distal, unilateral, impairing only fine finger movements and hand postures and affecting both transitive and intransitive movements; it may be difficult to disentangle from pure motor deficits associated with corticospinal tract lesions.

Apraxia may also be defined anatomically:

- *Parietal* (*posterior*):

  Ideational and ideomotor apraxia are seen with unilateral lesions of the inferior parietal lobule (most usually of the left hemisphere), or premotor area of the frontal lobe (Brodmann areas 6 and 8).

- *Frontal* (*anterior*):

  Unilateral lesions of the supplementary motor area are associated with impairment in tasks requiring bimanual co-ordination, leading to difficulties with alternating hand movements, drawing alternating patterns (*e.g.* m n m n in joined up writing: alternating sequences test, Luria figures). This may be associated with the presence of a grasp reflex and alien limb phenomena (limb-kinetic type of apraxia).

Apraxia is more common and severe with left hemisphere lesions.

Difficulties with the clinical definition of apraxia persist, as for the agnosias. For example, "dressing apraxia" and "constructional apraxia" are now considered visuospatial problems rather than true apraxias. Likewise, some cases labelled as eyelid apraxia or as gait apraxia are not true ideational apraxias.

References

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Cross References

Alien hand, Alien limb; Body part as object; Crossed apraxia; Dysdiadochokinesia; Eyelid apraxia; Forced groping; Frontal lobe syndromes; Gait apraxia; Grasp reflex; Optic ataxia; Speech apraxia

Aprosexia

Aprosexia is a syndrome of psychomotor inefficiency, characterized by complaints of easy forgetting, for example of conversations as soon as they are finished, material just read or instructions just given. There is difficulty keeping the mind on a specific task, which is forgotten if the patient happens to be distracted by another task. These difficulties, which the patient has insight into and often bitterly complains of, are commonly encountered in the memory clinic. They probably represent a disturbance of attention or concentration, rather than being

a harbinger of dementia. These patients generally achieve normal scores on cognitive screening instruments and formal psychometric tests (and indeed may complain that these assessments do not test the function they are having difficulty with). Concurrent sleep disturbance, irritability, and low mood are common and may reflect an underlying affective disorder (anxiety, depression) which may merit specific treatment.

Cross References

Attention; Dementia

Aprosodia, Aprosody

Aprosodia or aprosody (dysprosodia, dysprosody) is a defect in or absence of the ability to produce or comprehend speech melody, intonation, cadence, rhythm, and accentuations, in other words the non-linguistic aspects of language which convey or imply emotion and attitude. Aprosodia may be classified, in a manner analogous to the aphasias, as:

- *Sensory* (*posterior*):

  Impaired comprehension of the emotional overtones of spoken language or emotional gesturing, also known as affective agnosia; this may be associated with visual extinction and anosognosia, reflecting right posterior temporoparietal region pathology.

- *Expressive*/*Motor* (*anterior*):

  An inability to produce emotional overtones ("emotional dysprosody", sometimes confusingly referred to as speech dyspraxia); this may occur in isolation with right sided anterior lesions, or in association with linguistic aspects of aphasia such as agrammatism with anterior left hemisphere damage.

References

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Cross References

Agnosia: Agrammatism; Anosognosia; Aphasia; Aphemia; Broca's aphasia; Fisher's sign; Foreign accent syndrome; Visual extinction

Arc De Cercle

- see OPISTHOTONOS

Arcuate Scotoma

An arcuate scotoma suggests retinal or optic nerve disease, such as glaucoma, acute ischaemic optic neuropathy, or the presence of drusen.

Cross References

Retinopathy; Scotoma; Visual field defects

Areflexia

Areflexia is an absence or a loss of tendon reflexes. This may be physiological, in that some individuals never demonstrate tendon reflexes; or pathological, reflecting an anatomical interruption or physiological dysfunction at any point along the monosynaptic reflex pathway which is the neuroanatomical substrate of phasic stretch reflexes. Sudden tendon stretch, as produced by a sharp blow from a tendon hammer, activates muscle spindle Ia afferents which pass to the ventral horn of the spinal cord, there activating α-motor neurones, the efferent limb of the reflex, so completing the monosynaptic arc. Hence, although reflexes are typically regarded as part of the examination of the motor system, reflex loss may also occur in "sensory" disorders, affecting the Ia afferents from the muscle spindle. It is often possible to "hear" that reflexes are absent from the thud of tendon hammer on tendon.

Areflexia is most often encountered in disorders of lower motor neurones, specifically radiculopathies, plexopathies and neuropathies (axonal and demyelinating). Areflexia may also occur in neuromuscular junction disorders, such as the Lambert-Eaton myasthenic syndrome, in which condition the reflexes may be "restored" following forced muscular contraction (facilitation). Transient areflexia may be seen in central nervous system disorders such as cataplexy, and in acute spinal cord syndromes ("spinal shock", *e.g.* acute compression, acute inflammatory myelopathy).

Cross References

Cataplexy; Facilitation; Hyporeflexia; Lower motor neurone (LMN) syndrome; Plexopathy; Radiculopathy; Reflexes

Argyll Robertson Pupil (ARP)

The Argyll Robertson pupil is small (miosis) and irregular. It fails to react to light (reflex iridoplegia), but does constrict to accommodation (when the eyes converge). In other words, there is light-near pupillary dissociation (a useful mnemonic is ARP = *a*ccommodation *r*eaction *p*reserved).

Since the light reflex is lost, testing for the accommodation reaction may be performed with the pupil directly illuminated: this can make it easier to see the response to accommodation, which is often difficult to observe when the pupil is small or in individuals with a dark iris. There may be an incomplete response to mydriatic drugs. Although pupil involvement is usually bilateral, it is often asymmetric, causing anisocoria.

The Argyll Robertson pupil was originally described in the context of neurosyphilis, specifically tabes dorsalis. If this pathological diagnosis is suspected, a helpful clinical concomitant is the associated loss of deep pain sensation, as assessed, for example, by vigorously squeezing the Achilles tendon (Abadie's sign).

There are, however, a number of recognized causes of ARP besides neurosyphilis, including:

- Multiple sclerosis.
- Encephalitis.
- Diabetes mellitus.
- Syringobulbia.
- Sarcoidosis.
- Lyme disease.
- Pinealoma.
- Herpes zoster.
- Hereditary motor and sensory neuropathies (Charcot-Marie Tooth disease; Dejerine-Sottas hypertrophic neuropathy).

Miosis and pupil irregularity are inconstant findings in some of these situations, in which case the term "pseudo-Argyll Robertson pupil" may be preferred.

The neuroanatomical substrate of the Argyll Robertson pupil is uncertain. A lesion in the tectum of the (rostral) midbrain proximal to the oculomotor nuclei has been suggested. In multiple sclerosis and sarcoidosis, magnetic resonance imaging has shown lesions in the periaqueductal grey matter at the level of the Edinger-Westphal nucleus, but these cases lacked miosis and may therefore be classified as pseudo-Argyll Robertson pupil. Some authorities think a partial oculomotor (III) nerve palsy or a lesion of the ciliary ganglion is a more likely cause.

References

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Thompson HS, Kardon RH. The Argyll Robertson pupil. *J Neuroophthalmol*. 2006; **26**: 134–8.

Cross References

Abadie's sign; Anisocoria; Light-near pupillary dissociation; Miosis; Pseudo-Argyll Robertson pupil

"Arm Drop"

"Arm drop", or the "face-hand test", has been suggested as a useful diagnostic test if hemiparesis or upper limb monoparesis is suspected to be psychogenic: the examiner lifts the paretic hand directly over the patient's face and drops it. It is said that in organic weakness the hand will hit the face, whereas patients with functional weakness avoid this consequence. However, the validity and reliability of this "avoidance testing manoeuvre" has never been examined; its clinical value is therefore doubtful.

Reference

Stone J, Zeman A, Sharpe M. Functional weakness and sensory disturbance. *J Neurol Neurosurg Psychiatry*. 2002; **73**: 241–5.

Cross References

Babinski's trunk-thigh test; Functional weakness and sensory disturbance; Hoover's sign

"Around the Clock" Paralysis

- see SEQUENTIAL PARESIS

Arthrogryposis

- see CONTRACTURE

Asemasia

Asemasia is an inability to indicate by signs or spoken language. The term was invented in the nineteenth century (Hamilton) as an alternative to aphasia, since in many cases of the latter there is more than a loss of speech, including impairments in pantomime (apraxia) and in symbolizing the relationships of things. Hughlings Jackson approved of the term but feared it was too late to displace the word aphasia.

Reference

Critchley M, Critchley EA. John Hughlings Jackson. Father of English neurology. New York: Oxford University Press; 1998. p. 106.

Cross References

Aphasia; Apraxia

Asomatognosia

Asomatognosia is a lack of regard for a part, or parts, of the body, most typically failure to acknowledge the existence of a hemiplegic left arm. Asomatognosia may be verbal (denial of limb ownership) or non-verbal (failure to dress or wash a limb). All patients with asomatognosia have hemispatial neglect (usually left), hence this would seem to be a precondition for the development of asomatognosia; indeed, for some authorities asomatognosia is synonymous with personal neglect. Attribution of the neglected limb to another person is known as somatoparaphrenia.

The neuroanatomical correlate of asomatognosia is damage to the right supramarginal gyrus and posterior corona radiata, most commonly due to a cerebrovascular event. Cases with right thalamic lesions have also been reported. The predilection of asomatognosia for the left side of the body may simply be a reflection of the aphasic problems associated with left-sided lesions that might be expected to produce asomatognosia for the right side.

Asomatognosia is related to anosognosia (unawareness or denial of illness) but the two are dissociable on clinical and experimental grounds. Some authorities consider asomatognosia as a form of confabulation. Asomatognosia and phantom limb may be conceptualised as converse phenomena (brain representation lost/present even though limb is present/lost).

References
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Cross References
Anosognosia; Confabulation; Neglect; Phantom limb; Somatoparaphrenia

Astasia
- see CATAPLEXY

Astasia-Abasia
Astasia-abasia is a terminology which has sometimes been given to a disorder of gait characterized by impaired balance (disequilibrium), wide base, shortened stride, start/turn hesitation and freezing. The term has no standardized definition and hence may mean different things to different observers; it has also been used to describe a disorder characterised by inability to stand or walk despite normal leg strength when lying or sitting, believed to be psychogenic (although gait apraxia may have similar features).

Modern clinical classifications of gait disorders subsume astasia-abasia under the categories of subcortical disequilibrium and frontal disequilibrium, *i.e.* gait disorders with prominent disequilibrium or impaired postural control. A transient inability to sit or stand despite normal limb strength has been reported after acute infarction of the thalamus (thalamic astasia), caudal cingulate gyrus, or supplementary motor area.

References
Masdeu JC, Gorelick PB. Thalamic astasia: inability to stand after unilateral thalamic lesions. *Ann Neurol*. 1988; **23**: 596–603.
Nutt JG, Marsden CD, Thompson PD. Human walking and higher-level gait disorders, particularly in the elderly. *Neurology*. 1993; **43**: 268–79.
Cross References
Gait apraxia; Presbyastasis

Astereognosis
Astereognosis is the failure to recognize a familiar object, such as a key or a coin, palpated in the hand with the eyes closed, despite intact primary sensory modalities. Description of qualities such as the size, shape and texture of the object may be possible.

Hence, this would seem to be a failure of higher order (*i.e.* cortical) processing and it is most often associated with lesions of the posterior parietal lobe (post central gyrus) association cortex. There may be associated impairments of two-point discrimination and graphaesthesia (cortical sensory syndrome), although the latter can also occur with more peripheral lesions.

Astereognosis was said to be invariably present in the original description of the thalamic syndrome by Dejerine and Roussy. Very occasional cases in which astereognosis has been observed with subcortical lesions, such as cervical meningioma, have been reported, presumably interrupting sensory pathways which ascend to the thalamus.

Some authorities recommend the terms stereoanaesthesia or stereohypaesthesia as more appropriate descriptors of this phenomenon than astereognosis, to emphasize that this may be a disorder of perception rather than a true agnosia (for a similar debate in the visual domain, see Dysmorphopsia).
Cross References
Agnosia; Agraphaesthesia; Dysmorphopsia; Two-point discrimination

Asterixis
Asterixis is a sudden, brief, arrhythmic lapse of sustained posture due to involuntary interruption in muscle contraction. The term was coined by Raymond Adams and Joseph Foley to describe a movement disorder they saw in the context of liver disease.

Asterixis is most easily demonstrated by observing the dorsiflexed hands with arms outstretched (*i.e.* the motion to indicate "stop"), lapses being seen as flicking or flapping movements of the hands ("flapping tremor"). Movement is associated with EMG silence in anti-gravity muscles for 35–200 ms. These features distinguish asterixis from tremor and myoclonus; the phenomenon has been sometimes described as negative myoclonus (a term to which Adams did not object, although it does not help in understanding the movement) or negative tremor. Asterixis may be bilateral or unilateral.

Recognised causes of asterixis include:

- Hepatic encephalopathy ("liver flap").
- Hypercapnia.
- Uraemia.
- Drug-induced, *e.g.* anticonvulsants, levodopa.
- Structural brain lesions: thalamic lesions (haemorrhage, thalamotomy).

Unilateral asterixis has been described in the context of stroke, contralateral to lesions of the midbrain (involving corticospinal fibres, medial lemniscus), thalamus (ventroposterolateral nucleus), primary motor cortex and parietal lobe; and ipsilateral to lesions of the pons or medulla.

References

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Pal G, Lin MM, Laureno R. Asterixis: a study of 103 patients. *Metab Brain Dis*. 2014; **29**: 813–24.

Cross References

Encephalopathy; Myoclonus; Tremor

Asthenopia

Asthenopia, literally "weak vision", is frequently used to describe "eye strain" due to uncorrected or inadequately corrected refractive errors, such as hyperopia (far-sightedness) or overcorrected myopia. Such refractive errors are sometimes blamed for headache.

Asynergia

Asynergia or dyssynergia is lack or impairment of synergy in sequential muscular contractions in the performance of complex or reflex movements, such that they seem to become broken up into their constituent parts, so called decomposition of movement. This may be evident when performing rapid alternating hand movements. Dyssynergy of speech may also occur, a phenomenon sometimes termed scanning speech or scanning dysarthria. This is typically seen in cerebellar syndromes, most often those affecting the cerebellar hemispheres, and may coexist with other signs of cerebellar disease such as ataxia, dysmetria, and dysdiadochokinesia. Detrusor-sphincter dyssynergia may be a cause of urinary symptoms in spinal cord syndromes (injury, demyelination).

Cross References

Ataxia; Cerebellar syndromes; Dysarthria; Dysdiadochokinesia; Dysmetria; Scanning speech

Ataxia

Ataxia (or dystaxia) refers to a lack of co-ordination of voluntary motor acts, impairing their smooth performance. The rate, range, timing, direction, and force of movement may be affected. Ataxia is used most frequently to refer to a cerebellar problem, but sensory ataxia, optic ataxia, and frontal ataxia are also described, so if possible the term should be qualified rather than used in isolation.

- *Cerebellar ataxia*:

  Defective timing of agonist and antagonist muscle contraction (asynergia) produces jerking, staggering, inaccurate movements (decomposition of movement), which may manifest as intention tremor, dysmetria (past pointing), dysdiadochokinesia, ataxic dysarthria (sometimes known as scanning speech, although this also has other connotations), excessive rebound phenomenon, macrographia, head tremor (titubation), gait ataxia, and abnormal eye movements (nystagmus, square-wave jerks, saccadic intrusions). There may be concurrent limb hypotonia. Cerebellar hemisphere lesions cause ipsilateral limb ataxia (hemiataxia; ataxia on finger-to-nose, finger chase, and/or heel-shin testing) whereas midline cerebellar lesions involving the vermis produce selective truncal and gait ataxia. An International Cooperative Ataxia Rating Scale has been developed to assess the efficacy of treatments for cerebellar ataxia.

- *Sensory ataxia*:

  This results from impaired proprioception, and may be seen in disease of the dorsal (posterior) columns of the spinal cord (hence "spinal ataxia"), sensory neuropathies, and neuronopathies affecting the dorsal root ganglia. It is markedly exacerbated by removal of visual cues (*e.g.* as in Romberg's sign), unlike the situation with cerebellar ataxia, and may also lead to pseudoathetosis.

- *Optic ataxia*:

  Misreaching for visually presented targets, with dysmetria, due to a parieto-occipital lesion, as seen in Balint's syndrome.

- "*Frontal ataxia*":

  Similar to, and sometimes indistinguishable from, cerebellar ataxia, but results from lesions of the contralateral frontal cortex or frontopontine fibres, often from tumours invading the frontal lobe or corpus callosum. These fibres run in the corticopontocerebellar tract, synapsing in the pons before passing through the middle cerebellar peduncle to the contralateral cerebellar hemisphere.

Triple ataxia, the rare concurrence of cerebellar, sensory and optic types of ataxia, may be associated with an alien limb phenomenon (sensory type).

There are many causes of cerebellar ataxia, including:

- *Inherited*:

  Autosomal recessive: Friedreich's ataxia, ataxia with isolated vitamin E deficiency, ataxia with oculomotor apraxia (AOA types 1 and 2).
  Autosomal dominant: clinically ADCA types I, II, and III, now reclassified genetically as spinocerebellar ataxias: many types (>50) now described.
  Episodic ataxias: channelopathies involving potassium (type 1) and calcium (type 2) ion channels.
  Mitochondrial disorders.
  Huntington's disease.
  Dentatorubropallidoluysian atrophy (DRPLA).
  Inherited prion diseases, especially Gerstmann-Straussler-Scheinker (GSS) syndrome.

- *Acquired*:

  Cerebrovascular events (infarct, haemorrhage): usually cause hemiataxia; postanoxic cerebellar ataxia.
  Inflammatory: demyelination: multiple sclerosis, Miller Fisher variant of Guillain-Barré syndrome, central pontine and extrapontine myelinolysis.
  Inflammatory: infection: cerebellitis with Epstein-Barr virus; encephalitis with *Mycoplasma*; HIV.

Neoplasia: tumours, paraneoplastic syndromes.
Neurodegeneration: one variant of multiple system atrophy (MSA-C); prion diseases (Brownell-Oppenheimer variant of sporadic Creutzfeldt-Jakob disease, kuru); idiopathic late-onset cerebellar ataxia.
Drugs/toxins, *e.g*. alcohol, phenytoin.
Metabolic: vitamin E deficiency, thiamine deficiency (Wernicke's encephalopathy), gluten ataxia, hypothyroidism (debatable).

References

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Cross References

Alien hand, Alien limb; Asynergia; Balint's syndrome; Cerebellar syndromes; Dysarthria; Dysdiadochokinesia; Dysmetria; Head tremor; Hemiataxia; Hypotonia, Hypotonus; Macrographia; Nystagmus; Optic ataxia; Proprioception; Pseudoathetosis; Rebound phenomenon; Rombergism, Romberg's sign; Saccadic intrusion, Saccadic pursuit; Scanning speech; Square-wave jerks; Tandem walking; Tremor

Ataxic Hemiparesis

Ataxic hemiparesis is a syndrome of ipsilateral hemiataxia and hemiparesis, the latter affecting the leg more severely than the arm (crural paresis). There may be additional dysarthria, nystagmus, paraesthesia and pain.

This syndrome is caused by lacunar (small deep) infarcts in the contralateral basal pons at the junction of the upper third and lower two-thirds. It may also be seen with infarcts in the contralateral thalamocapsular region, posterior limb of the internal capsule (anterior choroidal artery syndrome), red nucleus, and the paracentral region (anterior cerebral artery territory). Sensory loss is an indicator of capsular involvement; pain in the absence of other sensory features is an indicator of thalamic involvement.

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Gorman MJ, Dafer R, Levine SR. Ataxic hemiparesis: critical appraisal of a lacunar syndrome. *Stroke*. 1998; **29** :2549–55.

Cross References

Ataxia; Hemiataxia; Hemiparesis; Pseudochoreoathetosis

Ataxic Nystagmus

- see INTERNUCLEAR OPHTHALMOPLEGIA (INO); NYSTAGMUS

Athetosis

Athetosis is the name sometimes given to an involuntary movement disorder characterized by slow, sinuous, purposeless, writhing movements, often more evident in the distal part of the limbs. Athetosis often co-exists with the more flowing, dance-like movements of chorea, in which case the movement disorder may be described as choreoathetosis. Indeed the term athetosis is now little used except in the context of "athetoid cerebral palsy". Athetoid-like movements of the outstretched hands may also been seen in the presence of sensory ataxia (impaired proprioception) and are known as pseudoathetosis or pseudochoreoathetosis. Choreoathetoid movements result from disorders of the basal ganglia.

References
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Turny F, Jedynak P, Agid Y. Athetosis or dystonia? [in French]. *Rev Neurol (Paris)*. 2004; **160**: 759–64.
Cross References
Chorea, Choreoathetosis; Pseudoathetosis; Pseudochoreoathetosis

Athymhormia

Athymhormia, or athymormia, also known as the robot syndrome, is a name given to a form of abulia or akinetic mutism in which there is loss of self-autoactivation. Clinically there is a marked discrepancy between heteroactivation, behaviour under the influence of exogenous stimulation, which is normal or almost normal, and autoactivation. Left alone, patients are akinetic and mute, a state also known as loss of psychic self-activation or pure psychic akinesia. It is associated with bilateral deep lesions of the frontal white matter or of the basal ganglia, especially the globus pallidus. It has also been described as a feature of Perry syndrome. Athymhormia is thus environment-dependent, since patients normalize initiation and cognition when stimulated, an important differentiation from apathy and akinetic mutism.
References
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Cross References
Abulia; Akinetic mutism; Apathy

Atrophy

Atrophy refers to a wasting or thinning of tissues. The term is often applied to wasted muscles, usually in the context of lower motor neurone pathology (in which case it may be synonymous with amyotrophy), but also with disuse. Atrophy develops more quickly after lower, as opposed to upper, motor neurone lesions. It may also be applied to other tissues, such as subcutaneous tissue (as in hemifacial atrophy). Muscle atrophy may sometimes be remote from the affected part of the neuraxis, hence a false-localising sign, for example the wasting of intrinsic hand muscles sometimes seen with foramen magnum lesions.
Cross References
Amyotrophy; "False-localising signs"; Hemifacial atrophy; Lower motor neurone (LMN) syndrome; Wasting

"Attended Alone" Sign

Collateral history is crucial in assessing cognitive disorders, especially complaints of memory impairment, for which reason individuals referred to memory clinics are usually asked to bring with them a spouse, relative or friend who knows them well to provide such history. Failure to attend with an informant, the "attended alone" sign, is a robust (*i.e.* very sensitive, > 0.95) marker of the absence of dementia and of cognitive health in patients with subjective memory complaints. This may be categorized as one of the non-canonical signs useful in the assessment of cognitive impairment, along with the applause sign and the head turning sign.
References
Larner AJ. Screening utility of the "attended alone" sign for subjective memory impairment. *Alzheimer Dis Assoc Disord*. 2014; **28**: 364–5.
Larner AJ. Neurological signs of possible diagnostic value in the cognitive disorders clinic. *Pract Neurol*. 2014; **14**: 332–335.

Cross References
Applause sign; Dementia; "Head turning sign"

Attention

Attention is a distributed cognitive function, important for the operation of many other cognitive domains; the terms concentration, vigilance, and persistence may sometimes be used synonymously with attention. Attention denotes the preferential allocation of (finite) neuronal resources to relevant events during a specific time period. It is generally accepted that attention is effortful, selective, and closely linked to intention. Distinction may be made between different types of attention, *viz*.:

- Sustained (vigilance): maintenance of behavioural response over a prolonged time period.
- Selective: focus directed to just one source despite competing or distracting stimuli.
- Divided/executive function: processing more than one information source at a time, generally regarded as the highest level of attention.

Impairment of attentional mechanisms may lead to distractability (with a resulting complaint of poor memory, perhaps better termed aprosexia), disorientation in time and place, perceptual problems, and behavioural problems (*e.g*. disinhibition), as in the cardinal disorder of attention, delirium.

The neuroanatomical substrates of attention encompass the ascending reticular activating system of the brainstem, the thalamus, and the prefrontal (multimodal association) cerebral cortex (especially on the right side). Damage to any of these areas of the "attentional matrix" may cause impaired attention.

Attentional mechanisms may be tested in a variety of ways. Those adapted to "bedside" use all essentially look for a defect in selective attention, also known as working memory or short term memory (although this does not necessarily equate with lay use of the term "short term memory"):

- Orientation in time/place.
- Digit span forwards/backwards.
- Reciting months of the year backwards, counting back from 30 to 1.
- Serial sevens (serial subtraction of 7 from 100, = 93, 86, 79, 72, 65).

In the presence of severe attentional disorder (as in delirium) it is difficult to make any meaningful assessment of other cognitive domains (*e.g*. memory).

Besides delirium, attentional impairments may be seen following head injury, and in ostensibly "alert" patients, *e.g*. with Alzheimer's disease (the dysexecutive syndrome of impaired divided attention).

References

Mesulam MM. Attentional and confusional states. *Continuum (Minneap Minn)*. 2010; **16**: 128–39.

Scholey A. Attention. In: Perry E, Ashton H, Young A, editors. Neurochemistry of consciousness: neurotransmitters in mind. Amsterdam: John Benjamins; 2002. p. 43–63.

Cross References
Aprosexia; Delirium; Dementia; Disinhibition; Dysexecutive syndrome; Frontal lobe syndromes; Pseudodementia

Auditory Agnosia

Auditory agnosia refers to an inability to appreciate the meaning of sounds despite normal hearing (perception of pure tones as assessed by audiological examination) and preservation of other cognitive functions such as language. This agnosia may be general (affecting all types

of sound perception) or selective, *e.g*. for verbal material (pure word deafness) or non-verbal material, either sounds (bells, whistles, animal noises) or music (amusia, of receptive or sensory type).

Reference

Robert Slevc L, Shell AR. Auditory agnosia. *Handb Clin Neurol*. 2015; **129**: 573–87.

Cross References

Agnosia; Amusia; Phonagnosia; Pure word deafness

Auditory Perseveration

- see PALINACOUSIS, PALINACUSIS

Auditory-Visual Synaesthesia

This name has been given to the phenomenon of sudden sound-evoked light flashes in patients with optic nerve disorders. This may be equivalent to noise-induced visual phosphenes or sound-induced photisms. Such synaesthetic perceptions might be accounted for by direct cross-modal activation of occipital cortex by auditory stimuli, or by "disinhibited feedback" coupling the inducing stimulus and synaesthetic sensation in a "sensory nexus" area. Evidence from different patients for both possibilities has been reported.

References

Afra P, Anderson J, Funke M, et al. Neurophysiological investigation of idiopathic acquired auditory-visual synesthesia. *Neurocase*. 2012; **18**: 323–9.

Neufeld J, Sinke C, Zedler M, et al. Disinhibited feedback as a cause of synesthesia: evidence from a functional connectivity study on auditory-visual synesthetes. *Neuropsychologia*. 2012; **50**: 1471–7.

Cross References

Phosphene; Synaesthesia

Augmentation

The term augmentation may be used to describe different phenomena.

In Lambert-Eaton myasthenic syndrome (LEMS), augmentation refers to an increase in the strength of affected muscles detected in the first few seconds of maximal voluntary muscle contraction. This may also be known as Lambert's sign. Facilitation is another term used to describe the post-tetancic potentiation seen in LEMS.

Augmentation may also be used to refer to the paradoxical worsening of the symptoms of restless legs syndrome under dopaminergic treatment, manifesting as earlier symptom onset in the evenings or afternoons, shorter periods of rest required to provoke symptom onset, greater intensity of symptoms when they occur, spread of symptoms to other body parts such as the arms, and decreased duration of benefit from medication.

Reference

Trenkwalder C, Winkelmann J, Inoue Y, Paulus W. Restless legs syndrome – current therapies and management of augmentation. *Nat Rev Neurol*. 2015; **11**: 434–45.

Cross References

Facilitation; Lambert's sign

Aura

An aura is a brief feeling or sensation, lasting seconds to minutes, occurring immediately before the onset of a paroxysmal neurological event such as an epileptic seizure or a migraine attack (migraine with aura, "classical migraine"), "warning" of its imminent presentation. Migraine aura may also occur in isolation (migraine aura without headache, "migraine equivalent").

An aura indicates the focal onset of neurological dysfunction. Auras are exclusively subjective, and may be entirely sensory, such as the fortification spectra (teichopsia) of migraine, or more complex, labelled psychosensory or experiential, as in certain seizures. Epileptic auras may be classified into subgroups:

- *Somatosensory*:

  e.g. paraesthesia.
- *Visual*:

  Hallucinations, illusions; occipital or temporal origin; complex hallucinations and a "tunnel vision" phenomenon are exclusive to seizures of anteromedial temporal and occipitotemporal origin, whereas elementary hallucinations, illusions, and visual loss are common to both occipital and temporal lobe seizures.
- *Auditory*:

  May indicate an origin in the superior temporal gyrus.
- *Olfactory*:

  Parosmia may occur in seizures of medial temporal lobe origin (uncus; uncinate fits).
- *Gustatory*
- *Autonomic*
- *Abdominal*:

  Rising epigastic sensation ("visceral aura") of temporal lobe epilepsy.
- *Psychic*:

  Complex hallucinations or illusions that usually affect different senses, *e.g*. distortions of familiarity such as *déjà vu* or *jamais vu* auras of focal-onset epilepsy, indicative of temporal lobe and limbic onset respectively.

References

Bien CG, Benninger FO, Urbach H, Schramm J, Kurthen M, Elger CE. Localizing value of epileptic visual auras. *Brain*. 2000; **123**: 244–253.

Charles A, Hansen JM. Migraine aura: new ideas about cause, classification, and clinical significance. *Curr Opin Neurol*. 2015; **28**: 255–60.

Lüders H, Acharya J, Baumgartner C, et al. Semiological seizure classification. *Epilepsia*. 1998; **39**: 1006–13.

Cross References

"Alice in Wonderland" syndrome; *Déjà vu*; Fortification spectra; Hallucination; Illusion; *Jamais vu*; Parosmia; Seizure; "Tunnel vision"

Automatic Obedience

Automatic obedience may be seen in startle syndromes such as the jumping Frenchmen of Maine, latah, and myriachit, when a sudden shout of, for example, "jump" is followed by a jump. These are sometimes known as the startle-automatic obedience syndromes. Although initially classified (by Gilles de la Tourette) with tic syndromes, there are clear clinical and pathophysiological differences.

Reference

Lajonchere C, Nortz M, Finger S. Gilles de la Tourette and the discovery of Tourette syndrome. Includes a translation of his 1884 article. *Arch Neurol.* 1996; **53**: 567–74.

Cross Reference

Tic

Automatic Writing

Automatic writing is a behaviour characterised by increased writing activity. It has been suggested that it should refer specifically to behaviour which is permanently present or elicitable, characterised by compulsive, iterative and not necessarily complete, written reproduction of visually or orally perceived messages (*cf*. hypergraphia). This may be conceptualised as a particular, sometimes isolated, form of utilization behaviour in which the inhibitory functions of the frontal lobes are suppressed, perhaps due to a disconnection within frontal-subcortical circuits (for example following left inferior capsular genu infarction) leading to motor perseveration in writing.

References
Suzuki K, Miyamoto T, Miyamoto M, Hirata K. Transient automatic writing behavior following a left inferior capsular genu infarction. *Case Rep Neurol*. 2009; **1**: 8–14.
Van Vugt P, Paquier P, Kees L, Cras P. Increased writing activity in neurological conditions: a review and clinical study. *J Neurol Neurosurg Psychiatry*. 1996; **61**: 510–4.
Cross References
Hypergraphia; Utilization behaviour

Automatism

Automatisms are complex motor movements occurring in complex motor seizures, which resemble natural movements but occur in an inappropriate setting. These may occur during a state of impaired consciousness during or shortly after an epileptic seizure. There is usually amnesia for the event.

Automatisms occur in about one-third of patients with complex partial seizures, most commonly those of temporal or frontal lobe origin. Although there are qualitative differences between the automatisms seen in seizures arising from these sites, they are not of sufficient specificity to be of reliable diagnostic value; bizarre automatisms are more likely to be frontal.

Automatisms may take various forms:

- *Oro-facial movements*:

  e.g. lip smacking, chewing and swallowing movements, salivation (especially temporal lobe origin).
- *Gestural*:

  Hand fumbling, foot shuffling, tidying, or more complex actions such as undressing; upper limb movements are said to be more suggestive of temporal lobe origin, lower limb movements (kicking, cycling) of frontal lobe origin; pelvic thrusting (may also be seen in pseudoseizures).
- *Ambulatory*:

  Walking or running around (cursive seizures); prolonged wandering may be termed fugue or poriomania.
- *Emotional*:

  Laughing and, more rarely, crying (gelastic and dacrystic seizures, respectively, although crying may also be a feature of non-epileptic seizures), fear, anger.
- *Verbal*:

  Humming, whistling, grunting, speaking incoherently; vocalization is common in frontal lobe automatisms.

Automatic behaviour and fugue-like states may also occur in the context of narcolepsy, and must be differentiated from the automatisms of complex partial seizures, on the basis of history, examination and EEG findings.

References
Delgado-Escueto AV, Bascal FE, Treiman DM. Complex partial seizures on closed circuit television and EEGs: a study of 691 attacks in 79 patients. *Ann Neurol*. 1982; **11**: 292–300.
Lüders H, Acharya J, Baumgartner C, et al. Semiological seizure classification. *Epilepsia*. 1998; **39**: 1006–13.
Cross References
Absence; Aura; Pelvic thrusting; Poriomania; Seizure

Autophony

Autophony is the perception of the reverberation of ones own voice, which occurs with external or middle, but not inner, ear disease.

Autoscopy

Autoscopy or autoscopia (literally "seeing oneself") is a visual hallucination of ones own face, sometimes with upper body or entire body, likened to seeing oneself in a mirror (hence "mirror hallucination"). The hallucinated image is a mirror image, *i.e*. shows left-right reversal as in a mirror image. Unlike heautoscopy, there is a coincidence of egocentric and body-centred perspectives. Autoscopy may be associated with parieto-occipital space-occupying lesions, epilepsy, migraine, and depression.

References

Blanke O, Landis T, Spinelli L, Seeck M. Out-of-body experience and autoscopy of neurological origin. *Brain*. 2004; **127**: 243–58.

Brugger P. Reflective mirrors: perspective taking in autoscopic phenomena. *Cogn Neuropsychiatry*. 2002; **7**:179–94.

Garry G. A case of autoscopy in a patient with depressive illness. *Prog Neurol Psychiatry*. 2012; **16**(5): 17–8, 20–1.

Cross References

Hallucination; Heautoscopy

Autotopagnosia

Autotopagnosia, or somatotopagnosia, is a rare disorder of body schema characterized by inability to identify parts of the body, either to verbal command or by imitation; this is sometimes localized but may sometimes involve all parts of the body. This may be a form of category-specific anomia with maximum difficulty for naming body parts, or one feature of anosognosia. Finger agnosia and right-left disorientation are partial forms of autotopagnosia, all of which are most often seen following cerebrovascular events involving the left parietal area.

Reference

Ogden JA. Autotopagnosia: occurrence in a patient without nominal aphasia and with an intact ability to point to parts of animals and objects. *Brain*. 1985; **108**: 1009–22.

Cross References

Agnosia; Anosognosia; Finger agnosia; Gerstmann syndrome; Right-left disorientation; Somatoparaphrenia