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## **Abadie's Sign**

Abadie's sign is the absence or diminution of 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).

## **Cross References**

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 maneuver 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 old age

obesity

after abdominal surgery

after multiple pregnancies

in acute abdominal disorders (Rosenbach's sign).

However, absence of all superficial abdominal reflexes may be of localizing 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 primary lateral sclerosis variant of motor neurone disease from multiple sclerosis. However, no prospective study of abdominal reflexes in multiple sclerosis has been reported.

**References**

Dick JPR. The deep tendon and the abdominal reflexes. *Journal of Neurology, Neurosurgery and Psychiatry* 2003; **74**: 150-153

**Cross References**

Bevor's sign; Upper motor neurone (UMN) syndrome

**Abducens (VI) Nerve Palsy**

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

Abducens (VI) nerve palsy may be due to:

Microinfarction in the nerve, due to hypertension, diabetes mellitus

Raised intracranial pressure: a "false-localizing 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).

Isolated weakness of the lateral rectus muscle 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 as "lateral rectus palsy," rather than abducens nerve palsy, until the etiological diagnosis is established.

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

**Cross References**

Diplopia; "False-localizing signs"

**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 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, "trance-like" state, or "glazing over," 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 localization-related complex partial seizures.

**Cross References**

Automatism; Seizures

**Abulia**

Abulia (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 labeled as “pseudodepression.” More plausibly, abulia has been thought of as a minor or partial form of akinetic mutism. There may also be some clinical overlap with catatonia. 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 the brain, 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 lobe dementia

Other causes of frontal lobe disease: tumor, abscess

Metabolic, electrolyte disorders: hypoxia, hypoglycemia, hepatic encephalopathy

Treatment is of the underlying cause where possible. There is anecdotal evidence that the dopamine agonist bromocriptine may help.

**References**

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Bhatia KP, Marsden CD. The behavioral and motor consequences of focal lesions of the basal ganglia in man. *Brain* 1994; **117**: 859-876

Fisher CM. Abulia. In: Bogousslavsky J, Caplan L (eds.). *Stroke syndromes*. Cambridge: CUP, 1995: 182-187

**Cross References**

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

**Acalculia**

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 coexisting 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 part of the Gerstmann syndrome 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 (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 while all others are impaired.

In patients with mild to moderate Alzheimer's disease with dyscalculia but no attentional or language impairments, cerebral glucose metabolism was found to be impaired in the left inferior parietal lobe and inferior temporal gyrus.

Preservation of calculation skills in the face of total language disson (production and comprehension) has been reported with focal left temporal lobe atrophy probably due to Pick's disease.

### References

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- Rossor M, Warrington EK, Cipolotti L. The isolation of calculation skills. *Journal of Neurology* 1995; **242**: 78-81

### Cross References

Agraphia; Alexia; Aphasia; Gerstmann syndrome; Neglect

**Accommodation Reflex**

- see PUPILLARY REFLEXES

**Achilles Reflex**

Plantar flexion at the ankle following phasic stretch of the Achilles tendon, produced by a blow with a tendon hammer either directly upon the Achilles tendon or with a plantar strike, constitutes the ankle or Achilles reflex, mediated through sacral segments S1 and S2 and the sciatic and posterior tibial nerves. This reflex is typically lost in polyneuropathies, S1 radiculopathy, and, possibly, as a consequence of normal ageing.

**Cross References**

Age-related signs; Neuropathy; Reflexes

**Achromatopsia**

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

Achromatopsia is most conveniently tested for clinically using pseudoisochromatic figures (*e.g.*, Ishihara plates), although these were specifically designed for detecting congenital color blindness and test the red-green channel more than blue-yellow. Sorting colors 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 “color 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 and symmetrical and do not change with time.

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 “gray” or “washed out”) and may be confined to only part of the visual field (*e.g.*, hemiachromatopsia).

Optic neuritis typically impairs color vision (red-green > blue-yellow), and this defect may persist while other features of the acute inflammation (impaired visual acuity, central scotoma) remit.

Cerebral achromatopsia results from cortical damage (most usually infarction) to the inferior occipitotemporal area. Area V4 of the visual cortex, which is devoted to color processing, is in the occipitotemporal (fusiform) and lingual gyri. 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 ischemia has been reported.

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

### References

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Zeki S. A century of cerebral achromatopsia. *Brain* 1990; **113**: 1721-1777

### Cross References

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

### Acousticopalpebral Reflex

- see BLINK REFLEX

### Action Dystonia

- see DYSTONIA

### Action Myoclonus

- see MYOCLONUS

### Adiadochokinesia

- see DYSADIADOCHOKINESIA

### Adie's Syndrome, Adie's Tonic Pupil

- see HOLMES-ADIE PUPIL, HOLMES-ADIE SYNDROME

### 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 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 (more details may be found in specific entries) includes:

- 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

Coordination: 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, palmo-mental, grasp reflexes may be more common with ageing

Impairments of gait; parkinsonism

- Sensory system:

Decreased sensitivity to vibratory perception; +/- pain, temperature, proprioception

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 (ed.). *Ageing and dementia: A methodological approach*. London: Edward Arnold, 1993: 144-174

Larner AJ. Neurological signs of aging. In: Pathy MSJ, Morley JE, Sinclair A (eds.). *Principles and practice of geriatric medicine* (4th edition). Chichester: Wiley, 2005: (in press)

**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 fundamental tastes (sweet, sour, bitter, and 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 flavors.

Neurological disorders may also account for ageusia. Afferent taste fibers 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 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. Fibers 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, anesthesia 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, tumor, 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.

### References

Finelli PF, Mair RG. Disturbances of taste and smell. In: Bradley WG, Daroff RB, Fenichel GM, Marsden CD (eds.). *Neurology in clinical practice* (3rd edition). Boston: Butterworth Heinemann, 2000: 263-269  
 Hepburn AL, Lanham JG. Sudden-onset ageusia in the antiphospholipid syndrome. *Journal of the Royal Society of Medicine* 1998; **91**: 640-641

### Cross References

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

### 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 verbally the stimulus (anomia). As a corollary of this last point, there should be no language disorder (aphasia) for the 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.

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, agnostic 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 poorly understood.

### References

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

Ghadiali E. Agnosia. *Advances in Clinical Neuroscience & Rehabilitation* 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; 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 nonsubstantive components of language), whereas nouns and verbs are relatively spared. Despite this impoverishment of language, or “telegraphic speech,” meaning is often still conveyed because of the high information content of verbs and nouns. Agrammatism is encountered in Broca’s type of nonfluent aphasia, associated with lesions of the posterior inferior part of the frontal lobe of the

dominant hemisphere (Broca's area). Agrammatic speech may also be dysprosodic.

### Cross References

Aphasia; Aprosodia, Aprosody

### Agraphesthesia

Agraphesthesia, dysgraphesthesia, or graphanesthesia, is a loss or impairment of the ability to recognize letters or numbers traced on the skin (*i.e.*, of graphanesthesia). Whether this is a perceptual deficit or a tactile agnosia ("agraphognosia") remains a subject of debate. It occurs with damage to the somatosensory parietal cortex.

### Cross References

Agnosia; Tactile agnosia

### 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 kinesthetic 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 and Wernicke/posterior types of aphasia; oral spelling is impaired. From the linguistic viewpoint, two types of paragrammia may be distinguished, *viz.*:  
*surfacelexicalsemantic dysgraphia:* misspelling of irregular words, producing phonologically plausible errors (*e.g.*, sim-tums for symptoms); this is seen with left temporoparietal lesions (*e.g.*, Alzheimer's disease, Pick's disease);  
*deepphonological dysgraphia:* inability to spell unfamiliar words and nonwords; semantic errors; seen with extensive left hemisphere damage.
- *Mechanical agraphia:*  
 Impaired motor control, due to paresis (as in dominant parietal damage), dyspraxia (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 nondominant hemisphere lesion; there may be missing out or misspelling of the left side of words (paragrammia); 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

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Roeltgen DP. Agraphia. In: Heilman KM, Valenstein E (eds.). *Clinical neuropsychology* (4th edition). Oxford: OUP, 2003: 126-145

### Cross References

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

### Agraphognosia

- see AGRAPHESTHESIA

### Agrypnia

Agrypnia is severe, total insomnia of long duration. Recognized causes include trauma to the brainstem and/or thalamus, prion disease (fatal familial and sporadic fatal insomnia), Morvan's syndrome, von Economo's disease, trypanosomiasis, and a relapsing-remitting disorder of possible autoimmune pathogenesis responding to plasma exchange.

### References

Batocchi AP, Della Marca G, Mirabella M *et al.* Relapsing-remitting autoimmune agrypnia. *Annals of Neurology* 2001; **50**: 668-671

### 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. Moaning, humming, and groaning may also be features. Voluntary suppression of the movements may exacerbate inner tension or anxiety.

Recognized associations of akathisia include Parkinson's disease and neuroleptic medication (acute or tardive side effect), suggesting that dopamine depletion may contribute to the pathophysiology; dopamine depleting agents (*e.g.*, tetrabenazine, reserpine) may cause akathisia.

Treatment by reduction or cessation of neuroleptic therapy may help, but can exacerbate coexistent psychosis. Centrally acting  $\beta$ -blockers, such as propranolol, may also help, as may anticholinergic agents, amantadine, clonazepam, and clonidine.

### References

Sachdev P. *Akathisia and restless legs*. Cambridge: CUP, 1995

### Cross References

Parkinsonism; Tic

**Akinesia**

Akinesia is 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, 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 coexist 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 (akineti 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.

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 late in the course of Pick's disease and Alzheimer's disease

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/akineti-rigid syndromes show no or only partial response to these agents.

**References**

Imai H. Clinicophysiological features of akinesia. *European Neurology* 1996; **36(suppl1)**: 9-12

**Cross References**

Akineti mutism; Bradykinesia; Extinction; 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. Abulia has been characterized as a lesser form of akinetic mutism.

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:

anterior cingulate cortex (medial frontal region)

paramedian reticular formation, posterior diencephalon, hypothalamus

Other structures (e.g., globus pallidus) have been implicated but without pathological evidence.

These pathologies may be vascular, neoplastic, or structural (sub-acute communicating hydrocephalus). Akinetic mutism may be the final state common to the end-stages of a number of neurodegenerative pathologies.

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

### References

Cairns H. Disturbances of consciousness with lesions of the brain stem and diencephalon. *Brain* 1952; **75**: 109-146

Freeman FR. Akinetic mutism and bilateral anterior cerebral artery occlusion. *Journal of Neurology, Neurosurgery and Psychiatry* 1971; **34**: 693-698

Ross ED, Stewart RM. Akinetic mutism from hypothalamic damage: successful treatment with dopamine agonists. *Neurology* 1981; **31**: 1435-1439

### Cross References

Abulia; Akinesia; 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 color, 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 it may be associated with acalculia and aphasia.

### References

Zihl J, Von Cramon D, Mai N. Selective disturbance of movement vision after bilateral brain damage. *Brain* 1983; **106**: 313-340

Zeki S. Cerebral akinetopsia (cerebral visual motion blindness). *Brain* 1991; **114**: 811-824

### Cross References

Acalculia; Aphasia; Riddoch's phenomenon

## 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 may be described as an acquired dyslexia.

Alexia may be categorized 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 recognize 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"). 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 color anomia or impaired color 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.
- *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 hemispacial 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, its afferents from early mesial visual areas, or its efferents 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 fibers 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 labored 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 nonwords can be read; this may be seen with left medial +/- lateral temporal lobe pathology (e.g., infarction, temporal lobe Pick’s disease, late Alzheimer’s disease).

- *Phonological dyslexia:*

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

- *Deep dyslexia:*

The inability to translate orthography to phonology, manifesting as an inability to read plausible nonwords (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

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### 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

### Alexithymia

Alexithymia is a reduced ability to identify and express ones feelings. This may contribute to various physical and behavioral 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.

### References

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### “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 somatosensory features 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**

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**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 tumor of the right (nondominant) 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.

**References**

- Silva MT, Howard RS, Kartsounis LD, Ross Russell RW. The alien grasp reflex. *European Neurology* 1996; **36**: 55-56

**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 that manifests slow, involuntary, wandering (levitating), quasipurposive 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 and *le main étranger*). These phenomena are often associated with a prominent grasp reflex, forced groping, intermanual conflict, and magnetic movements (*q.v.*) of the hand.

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

- Anterior or motor types:
  - Callosal*: characterized primarily by intermanual conflict;
  - Frontal*: shows features of environmental dependency, such as forced grasping and groping and utilization behavior.
- 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 tumors, hemorrhage
- Medial frontal cortex infarction (territory of the anterior cerebral artery)

Trauma and hemorrhage affecting both corpus callosum and medial frontal area

Alzheimer's disease (very rare)

Posterior cerebral artery occlusion (sensory variant)

Following commissurotomy (corpus callosotomy alone insufficient).

Functional imaging studies in corticobasal degeneration, along with the evidence from focal vascular lesions, suggest that damage to and/or hypometabolism of the medial frontal cortex (Brodmann area 32) and the supplementary motor area (Brodmann area 6) are associated with alien limb phenomena. More generally, it seems that these areas are involved in the execution of learned motor programs, and damage thereto may lead to the release of learned motor programs from voluntary control.

### References

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Doody RS, Jankovic J. The alien hand and related signs. *Journal of Neurology, Neurosurgery and Psychiatry* 1992; **55**: 806-810

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Josephs KA, Rossor MN. The alien limb. *Practical Neurology* 2004; **4**: 44-45

Sawle GV, Brooks DJ, Marsden CD, Frackowiak RSJ. Corticobasal degeneration: a unique pattern of regional cortical oxygen hypometabolism and striatal fluorodopa uptake demonstrated by positron emission tomography. *Brain* 1991; **114**: 541-556

### Cross References

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

### Allochiria

Allochiria is the transposition of objects 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 hemorrhage, a figure was copied with objects from the left side transposed to the right.

### References

Halligan PW, Marshall JC, Wade DT. Left on the right: allochiria in a case of left visuospatial neglect. *Journal of Neurology, Neurosurgery and Psychiatry* 1992; **55**: 717-719

### Cross References

Alloesthesia; Allokinesia; Neglect

### 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); 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 fibers (in which neurotrophins may play a role); ephaptic transmission (“cross-talk”) between large and small (nociceptive) afferent fibers; and abnormal central processing.

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

### **Cross References**

Hyperalgesia; Hyperpathia

### **Alloesthesia**

Alloesthesia (allesthesia, alloaesthesia) is a 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. Visual alloesthesia, 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 tumors.

Tactile alloesthesia may be seen in the acute stage of right putaminal hemorrhage (but seldom in right thalamic hemorrhage) 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 alloesthesia.”

The mechanism of alloesthesia is uncertain: some consider it a disturbance within sensory pathways, others that it is a sensory response to neglect.

### **References**

Kawamura M, Hirayama K, Shinohara Y, Watanabe Y, Sugishita M. Alloaesthesia. *Brain* 1987; **110**: 225-236

### **Cross References**

Allochiria; Allokinesia; Neglect

### **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 increased in frequency with word length. 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**

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**Cross References**

Agraphia

**Allokinesia**

Allokinesia is a motor response in the wrong limb, or transposition of the intended movement to the contralateral side; the movement may also be in the wrong direction. This may be the motor system counterpart of alloesthesia, and is seen with right hemisphere lesions as part of a neglect syndrome.

**Cross References**

Allochiria; Alloesthesia; Neglect

**Alternate Cover Test**

- see COVER TESTS

**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.

Recognized causes of altitudinal visual field defects include:

- Monocular:
  - Central retinal artery occlusion (CRAO)
  - Acute ischemic optic neuropathy (AION)
  - Retinal detachment
  - Choroiditis
  - Glaucoma
  - Chronic atrophic papilledema
- 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 is 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 ischemic attack). Giant cell arteritis, systemic lupus erythematosus and the antiphospholipid antibody syndrome are also recognized 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 with 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 six months and three 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 centrocecal 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-Osterreith Complex Figure (nonverbal 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 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 recognized, 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 nonparaneoplastic)
  - 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 tumor, 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 nonparaneoplastic limbic encephalitis associated with autoantibodies directed against voltage-gated potassium channels.

Functional or psychogenic 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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### Cross References

Confabulation; Dementia

### 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 (loss of the ability to appreciate music) and expressive or motor (e.g., loss of ability to sing, whistle). 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). 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. It has been found 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 nondominant 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.

### References

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- Wertheim N. The amusias. In: Vinken PJ, Bruyn GW (eds.). *Handbook of clinical neurology, Vol. 4: Disorders of speech, perception, and symbolic behaviour*. Amsterdam: North-Holland Publishing, 1969: 195-206

### 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):
  - 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 nonspecific with respect to neuroanatomical substrate.



**Cross References**

Atrophy; Fasciculation; Neuropathy; Plexopathy; Radiculopathy; Wasting

**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 (anesthesia) 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 fibers of the spinothalamic pathway in the ventral funiculus (with corresponding thermoanesthesia), *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.

**References**

Larner AJ, Moss J, Rossi ML, Anderson M. Congenital insensitivity to pain: a 20 year follow up. *Journal of Neurology, Neurosurgery and Psychiatry* 1994; **57**: 973-974

**Cross References**

Anesthesia; 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.

**References**

Swash M, Chan CLH, Ponsford S. The anal reflex can be elicited by cough and sniff – validation of a clinical sign. *Journal of Neurology, Neurosurgery and Psychiatry* 2004; **75**: 521 (abstract 027)

**Cross References**

Reflexes

**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 in bulbar 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 called 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 may result from focal degeneration affecting the frontal operculum bilaterally (so-called Foix-Chavany-Marie syndrome).

**References**

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

**Cross References**

Aphemia; Bulbar palsy; Dysarthria

**Anesthesia**

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

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

Anesthesia dolorosa, or painful anesthesia, 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; Dysesthesia; Neuropathy; Paresthesia

**Angioscotoma**

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

**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. Localized or generalized 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**

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 defecation, leading to fecal 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 by local injections of botulinum toxin.

**References**

Jost WH, Muller-Lobeck H, Merkle W. Involuntary contractions of the striated anal sphincters as a cause of constipation: report of a case. *Diseases of the Colon and Rectum* 1998; **41**: 258-260

**Cross References**

Dystonia; Parkinsonism

**Anisocoria**

Anisocoria is 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 sympathetic innervation defect (darkness stimulates dilatation of normal pupil). Affected pupil constricted (miosis; oculosympathetic paresis):

- Horner’s syndrome

- Argyll Robertson pupil

- Cluster headache

- Anisocoria greater in bright light/less in dim light suggests defect in parasympathetic innervation to the pupil. Affected pupil dilated (mydriasis; oculoparasympathetic paresis):

- 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.

#### References

Kawasaki A. Approach to the patient with abnormal pupils. In: Biller J (ed.). *Practical neurology* (2nd edition). Philadelphia: Lippincott Williams & Wilkins, 2002: 135-146

#### Cross References

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

### 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 color (*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 as an early feature of Alzheimer's disease, or with any dominant hemisphere space-occupying lesion.

#### References

Benson DF, Ardila A. *Aphasia: a clinical perspective*. New York: OUP, 1996: 252-261

#### Cross References

Aphasia; Circumlocution; Paraphasia

### Anosmia

Anosmia is the inability to perceive smells due to damage to the olfactory pathways (olfactory neuroepithelium, olfactory nerves, rhinen-

cephalon). Olfaction may be tested with kits containing specific odors (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., due to 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 fibers 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 early 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.

#### References

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 Hawkes CH, Shephard BC. Olfactory evoked responses and identification tests in neurological disease. *Annals of the New York Academy of Sciences* 1998; **855**: 608-615

#### 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 (anosognosia). *Revue Neurologique* 1914; **12**: 845-848

#### Cross References

Anosognosia; Belle indifférence

#### Anosognosia

Anosognosia refers to a patient's unawareness or denial of illness. The term was first used by von Monakow (1885) and has been used to describe denial of blindness (Anton's syndrome), deafness, hemiplegia (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.

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 anesthesia 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 (eds.). *Clinical neuropsychology* (4th edition). Oxford: OUP, 2003: 185-214
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#### Cross References

Agnosia; Anosodiaphoria; Asomatognosia; Cortical blindness; Extinction; Jargon aphasia; Misoplegia; Neglect; Somatoparaphrenia

#### Anserina

Autonomically mediated piloerection and thermoconstriction may produce "goose bumps," cold and bumpy skin that may be likened to that of a plucked goose.

#### Antecollis

Antecollis (anterocollis) is forward flexion of the neck. It may be a feature of multiple system atrophy (*cf.* retrocollis in progressive supranu-

clear 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."

#### **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 References**

Parkinsonism

#### **Anton's Syndrome**

- see ANOSOGNOSIA; CONFABULATION; CORTICAL BLINDNESS

#### **Anwesenheit**

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

#### **References**

Fénélon G, Mahieux F, Huon R, Ziegler M. Hallucinations in Parkinson's disease: prevalence, phenomenology and risk factors. *Brain* 2000; **123**: 733-745

#### **Cross References**

Hallucination; Parkinsonism

#### **Apallic Syndrome**

- see VEGETATIVE STATES

#### **Apathy**

Apathy is a neurobehavioral disorder characterized by a lack of interest in environmental stimuli, manifest as listlessness, paucity of spontaneous movement (akinesia) or speech (mutism), and lack of initiative, spontaneity and drive. These are all 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 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 described following amphetamine or cocaine withdrawal, in neuroleptic-induced akinesia and in psychotic depression.

SSRIs may sometimes be helpful in the treatment of apathy.

### References

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### Cross References

Abulia; Akinetic mutism; Dementia; Frontal lobe syndromes

### Aphasia

Aphasia, or dysphasia, is an acquired loss or impairment of language (as opposed to speech) function. Language may be defined as the complex system of symbols used for communication (including reading and writing), encompassing various linguistic components (*viz.* phonology, semantic/lexical, syntax), all of which are dependent on dominant hemisphere integrity. Nonlinguistic 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 coexist 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, labored, with agrammatism and dysprosody (nonfluent); 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). For example, motor (“expressive”) aphasias are characterized by nonfluent 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.

Aphasias most commonly follow 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 (primary progressive aphasia, semantic dementia).

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### Summary of findings in aphasia syndromes

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	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?

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### References

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- Willmes K, Poeck K. To what extent can aphasic syndromes be localized? *Brain* 1993; **116**: 1527-1540

### Cross References

Agrammatism; Agraphia; Alexia; Anomia; Aprosodia, Aprosody; Broca's aphasia; Circumlocution; Conduction aphasia; Conduit D'approche; Crossed aphasia; Dysarthria; Jargon aphasia; Neologism; Optic aphasia; Paraphasia; Transcortical aphasias; Wernicke's aphasia

### Aphemia

Aphemia was the name originally given by Broca to the language disorder subsequently named "Broca's aphasia." The term is now used to

describe a motor disorder of speech production with preserved comprehension of spoken and written language. This syndrome has also been called phonetic disintegration (*cf.* phonemic disintegration), pure anarthria, apraxic dysarthria, cortical dysarthria, verbal apraxia, subcortical motor aphasia, 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 encompasses at least some cases of the "foreign accent syndrome," in which altered speech production and/or prosody makes speech output sound foreign. 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.

#### References

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- Schiff HB, Alexander MP, Naeser MA, Galaburda AM. Aphemia: clinical-anatomic correlations. *Archives of Neurology* 1983; **40**: 720-727

#### Cross References

Anarthria; Aphasia; Aprosodia, Aprosody; Dysarthria; Phonemic disintegration; Speech apraxia

### Aphonia

Aphonia is loss of the sound of the voice, 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; patients may comment that they cannot hold any prolonged conversation. Aphonia of functional or hysterical origin is also recognized. 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

### 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 parts as objects (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; 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 coordination, 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 labeled as eyelid apraxia or gait apraxia are not true ideational apraxias. The exact nosological status of speech apraxia also remains tendentious.

### References

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#### **Cross References**

Alien hand, Alien limb; Body part as object; Crossed apraxia; 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, into which the patient has insight 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 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, the nonlinguistic 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

Monrad-Krohn GH. Dysprosody or altered “melody of language.” *Brain* 1947; **70**: 405-415

Ross ED. The aprosodias: functional-anatomic organization of the affective components of language in the right hemisphere. *Archives of Neurology* 1981; **38**: 561-569

## Cross References

Agnosia; Anosognosia; Aphasia; Aphemia; Broca’s aphasia; Fisher’s sign; Visual extinction

## Arc de Cercle

- see OPISTHOTONOS

## Arcuate Scotoma

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

## Cross References

Retinopathy; Scotoma

## 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  $\alpha$ -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 (ARP = accommodation reaction preserved). 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, especially 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 claimed. In multiple sclerosis and sarcoidosis, magnetic resonance imaging has shown lesions in the periaqueductal gray matter at the level of the Edinger-Westphal nucleus, but these cases lacked miosis and may be classified as pseudo-Argyll Robertson pupil. Some authorities think a partial oculomotor (III) nerve palsy or a lesion of the ciliary ganglion is more likely.

### References

Argyll Robertson D. Four cases of spinal myosis [*sic*]: with remarks on the action of light on the pupil. *Edinburgh Medical Journal* 1869; **15**: 487-493  
 Dacso CC, Bortz DL. Significance of the Argyll Robertson pupil in clinical medicine. *American Journal of Medicine* 1989; **86**: 199-202

### 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 maneuver” has never been examined; its clinical value is therefore doubtful.

### References

Stone J, Zeman A, Sharpe M. Functional weakness and sensory disturbance. *Journal of Neurology, Neurosurgery and Psychiatry* 2002; **73**: 241-245

### 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

### 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 nonverbal (failure to dress or wash 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 anatomical 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.

### References

Feinberg TE, Haber LD, Leeds NE. Verbal asomatognosia. *Neurology* 1990; **40**: 1391-1394

### Cross References

Anosognosia; Confabulation; Neglect; Somatoparaphrenia

### Astasia

- see CATAPLEXY

### Astasia-Abasia

Astasia-abasia is the name that 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 characterized 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 may be seen after an acute thalamic lesion (thalamic astasia).

### References

Nutt JG, Marsden CD, Thompson PD. Human walking and higher-level gait disorders, particularly in the elderly. *Neurology* 1993; **43**: 268-279

### Cross References

Gait apraxia

### 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 is a failure of higher order (*i.e.*, cortical) processing and is associated with lesions of the posterior parietal lobe (post central gyrus) association cortex. There may be associated impairments of two-point discrimination and graphesthesia (cortical sensory syndrome). Astereognosis was said to be invariably present in the original description of the thalamic syndrome by Dejerine and Roussy.

Some authorities recommend the terms stereoaesthesia or stereohypesthesia as more appropriate terms for this phenomenon, 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; Dysmorphopsia; Graphesthesia; Two-point discrimination

### Asterixis

Asterixis is a sudden, brief, arrhythmic lapse of sustained posture due to involuntary interruption in muscle contraction. It 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 antigravity muscles for 35-200 ms. These features distinguish asterixis from tremor and myoclonus; the phenomenon has previously been described as negative myoclonus or negative tremor.

Asterixis may be bilateral or unilateral. Recognized causes of asterixis include:



Hepatic encephalopathy

Hypercapnia

Uremia

Drug-induced, for example, anticonvulsants, levodopa

Structural brain lesions: thalamic lesions (hemorrhage, thalamotomy)

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

### References

Marchini M, Sayegh GA, Caudana R. Unilateral asterixis and stroke in 13 patients: localization of the lesions matching the CT scan images to an atlas. *European Journal of Neurology* 2004; **11(suppl2)**: 56 (abstract P1071)

### Cross References

Encephalopathy; Myoclonus; Tremor

### Asynergia

Asynergia or dyssynergia is lack or impairment of synergy of sequential muscular contraction in the performance of complex 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 (*q.v.*) 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.

### Cross References

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

### Ataxia

Ataxia or dystaxia refers to a lack of coordination 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 it is probably best to qualify ataxia rather than to use the word 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-nose and/or heel-shin testing) whereas midline cerebellar lesions involving the vermis produce selective truncal and gait ataxia.

- *Sensory ataxia:*

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 neuropathies 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 fibers, often from tumors invading the frontal lobe or corpus callosum. These fibers 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

Autosomal dominant: clinically ADCA types I, II, and III, now reclassified genetically as spinocerebellar ataxias, types 1-25 now described

Episodic ataxias: channelopathies involving potassium (type 1) and calcium (type 2) channels

Mitochondrial disorders

Huntington’s disease

Dentatorubropallidolusian atrophy (DRPLA)

Inherited prion diseases, especially Gerstmann-Straussler-Scheinker (GSS) syndrome

- Acquired:

Cerebrovascular events (infarct, hemorrhage): usually cause hemiataxia; postanoxic cerebellar ataxia

Inflammatory: demyelination: multiple sclerosis, Miller Fisher variant of Guillain-Barré syndrome, central pontine myelinolysis

Inflammatory: infection: cerebellitis with Epstein-Barr virus; encephalitis with *Mycoplasma*; HIV  
 Neoplasia: tumors, 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: for example, alcohol, phenytoin  
 Metabolic: vitamin E deficiency, thiamine deficiency (Wernicke's encephalopathy), gluten ataxia, hypothyroidism (debatable)

### References

Klockgether T (ed.). *Handbook of ataxia disorders*. New York: Marcel Dekker, 2000  
 Wood NW, Harding AE. Ataxic disorders. In: Bradley WG, Daroff RB, Fenichel GM, Marsden CD (eds.). *Neurology in clinical practice: principles of diagnosis and management* (3rd edition). Boston: Butterworth-Heinemann, 2000 309-317

### 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 (crual paresis). There may be additional dysarthria, nystagmus, paresthesia and pain.

This syndrome is caused by lacunar (small deep) infarction in the contralateral basis 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 of thalamic involvement.

### References

Bogousslavsky J, Regli F, Ghika J, Feldmeyer JJ. Painful ataxic hemiparesis. *Archives of Neurology* 1984; **41**: 892-893  
 Fisher CM. Ataxic hemiparesis. A pathologic study. *Archives of Neurology* 1978; **35**: 126-128  
 Gorman MJ, Dafer R, Levine SR. Ataxic hemiparesis: critical appraisal of a lacunar syndrome. *Stroke* 1998; **29**: 2549-2555

### Cross References

Ataxia; Hemiataxia; Hemiparesis; Pseudochoreoathetosis

**Ataxic Nystagmus**

- see INTERNUCLEAR OPHTHALMOPLEGIA; 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 coexists 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 be 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**

Pearce JMS. Athetosis and William Alexander Hammond, a founder of American neurology. In: Pearce JMS. *Fragments of neurological history*. London: Imperial College Press, 2003: 413-416

**Cross References**

Chorea, Choreoathetosis; Pseudoathetosis; Pseudochoreoathetosis

**Atrophy**

Atrophy is 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). Atrophy may sometimes be remote from the affected part of the neuraxis, hence a false-localizing sign, for example wasting of intrinsic hand muscles with foramen magnum lesions.

**Cross References**

Amyotrophy; “False-localizing signs”; Hemifacial atrophy; Lower motor neurone (LMN) syndrome; Wasting

**Attention**

Attention is a distributed cognitive function, important for the operation of many other cognitive domains; the terms concentration, vigilance, and persistence may be used synonymously with attention.

A distinction may be made between different types of attention, *viz.*:

- Sustained
- Selective
- Divided/executive function.

It is generally accepted that attention is effortful, selective, and closely linked to intention.

Impairment of attentional mechanisms may lead to distractibility (with a resulting complaint of poor memory, better termed aprosexia,

*q. v.*), disorientation in time and place, perceptual problems, and behavioral problems (*e. g.*, disinhibition), as in the cardinal disorder of attention, delirium (*q. v.*).

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). Damage to any of these areas 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/backward  
Reciting months of the year backward, 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, for example, with Alzheimer’s disease (the dysexecutive syndrome of impaired divided attention).

### References

- Parasuraman R. Attentional functioning in Alzheimer’s disease. In: Morris RG, Becker JT (eds.). *Cognitive neuropsychology of Alzheimer’s disease* (2nd edition). Oxford: OUP, 2004: 81-102
- Perry RJ, Hodges JR. Attention and executive deficits in Alzheimer’s disease: a critical review. *Brain* 1999; **122**: 383-404
- Scholey A. Attention. In: Perry E, Ashton H, Young A (eds.). *Neurochemistry of consciousness: neurotransmitters in mind*. Amsterdam: John Benjamins, 2002: 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 perception of pure tones as assessed by audiological examination. This agnosia may be for either verbal material (pure word deafness) or nonverbal material, either sounds (bells, whistles, animal noises) or music (amusia, of receptive or sensory type).

### Cross References

Agnosia; Amusia; Phonagnosia; Pure word deafness

### Auditory-Visual Synesthesia

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.

#### References

Jacobs L, Karpik A, Bozian D, Gothgen S. Auditory-visual synesthesia: sound-induced photisms. *Archives of Neurology* 1981; **38**: 211-216

#### Cross References

Phosphene; Synesthesia

### 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, although auras may also occur in isolation. 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, labeled psychosensory or experiential, as in certain seizures.

Epileptic auras may be classified into subgroups:

- *Somatosensory*:  
for example, paresthesia.
- *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; uncinatate fits).
- *Gustatory*
- *Autonomic*
- *Abdominal*:  
rising epigastric 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

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

Palmini A, Gloor P. The localizing value of auras in partial seizures. *Neurology* 1992; **42**: 801-808

#### **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.

#### **References**

Lajonchere C, Nortz M, Finger S. Gilles de la Tourette and the discovery of Tourette syndrome. Includes a translation of his 1884 article. *Archives of Neurology* 1996; **53**: 567-574

#### **Cross References**

Tic

#### **Automatic Writing Behavior**

Automatic writing behavior is a form of increased writing activity. It has been suggested that it should refer specifically to a permanently present or elicitable, compulsive, iterative and not necessarily complete, written reproduction of visually or orally perceived messages (*cf.* hypergraphia). This is characterized as a particular, sometimes isolated, form of utilization behavior in which the inhibitory functions of the frontal lobes are suppressed.

#### **References**

Van Vugt P, Paquier P, Kees L, Cras P. Increased writing activity in neurological conditions: a review and clinical study. *Journal of Neurology, Neurosurgery and Psychiatry* 1996; **61**: 510-514

#### **Cross References**

Hypergraphia; Utilization behavior

#### **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:*  
for example, 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 nonepileptic seizures), fear, anger.
- *Verbal:*  
humming, whistling, grunting, speaking incoherently; vocalization is common in frontal lobe automatisms.

Automatic behavior 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.

#### 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. *Annals of Neurology* 1982; **11**: 292-300
- Lüders H, Acharya J, Baumgartner C *et al.* Semiological seizure classification. *Epilepsia* 1998; **39**: 1006-1013

#### Cross References

Absence; Aura; Pelvic thrusting; Poriomania; Seizure

#### Autophony

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

#### Autoscopy

Autoscopy (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-centered perspectives. Autoscopy may be associated with parieto-occipital space-occupying lesions, epilepsy, and migraine.

#### References

- Blanke O, Landis T, Spinelli L, Seeck M. Out-of-body experience and autoscopy of neurological origin. *Brain* 2004; **127**: 243-258
- Brugger P. Reflective mirrors: perspective taking in autoscopic phenomena. *Cognitive Neuropsychiatry* 2002; **7**: 179-194



Maillard L, Vignal JP, Anxionnat R, Taillandier Vespignani L. Semiologic value of ictal autoscopy. *Epilepsia* 2004; **45**: 391-394

**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 at worst involves 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.

**Cross References**

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