D

Dalrymple's Sign

Dalrymple's sign is increased width of the palpebral fissure, often seen in hyperthyroidism.

Cross Reference

Lid retraction

Dazzle

Dazzle describes a painless intolerance of the eyes to bright light (*cf*. photophobia). It may be peripheral in origin (retinal disease; opacities within cornea, lens, vitreous); or central (lesions anywhere from optic nerve to occipitotemporal region).

Cross Reference

Photophobia

Decerebrate Rigidity

Decerebrate rigidity is a posture observed in comatose patients in which there is extension and pronation of the upper extremities, extension of the legs, and plantar flexion of the feet (= extensor posturing), which is taken to be an exaggeration of the normal standing position. Painful stimuli may induce opisthotonos, hyperextension and hyperpronation of the upper limbs.

Decerebrate rigidity occurs in severe metabolic disorders of the upper brainstem (anoxia/ischaemia, trauma, structural lesions, drug-intoxication). A similar picture was first observed by Sherrington (1898) following section of the brainstem of cats at the collicular level, below the red nuclei, such that the vestibular nuclei were intact. The action of the vestibular nuclei, unchecked by higher centres, may be responsible for the profound extensor tone.

Decerebrate rigidity indicates a deeper level of coma than decorticate rigidity; the transition from the latter to the former is associated with a worsening of prognosis.

Reference

Posner JB, Saper CB, Schiff ND, Plum F. Plum and Posner's diagnosis of stupor and coma. 4th ed. Oxford: Oxford University Press; 2007.

Cross References

Coma; Decorticate rigidity; Opisthotonos

De Clérambault Syndrome

- see DELUSION

Decomposition of Movement

- see ASYNERGIA

Decorticate Rigidity

Decorticate rigidity is a posture observed in comatose patients in which there is adduction of the shoulders and arms, and flexion of the elbows and wrists (= flexor posturing). The lesion responsible for decorticate rigidity is higher in the neuraxis than that causing decerebrate rigidity, often being diffuse cerebral hemisphere or diencephalic disease, although, despite the name, it may occur with upper brainstem lesions. Common causes are anoxia/ischaemia, trauma, and drugs.

Cross References

Coma; Decerebrate rigidity

A.J. Larner, *A Dictionary of Neurological Signs*,
DOI 10.1007/978-3-319-29821-4_4

Déjà Entendu
A sensation of familiarity akin to *déjà vu* but referring to auditory (literally "already heard") rather than visual experiences.

Déjà Vécu
- see *DÉJÀ VU*

Déjà Vu
Déjà vu (literally "already seen") is a subjective, inappropriate impression of familiarity for a present experience in relation to an undefined past. However, since the term has passed into the vernacular, not every patient complaining of "*déjà vu*" has a pathological problem. The term may be used colloquially to indicate familiar events or experiences (Yoga Berra: "It's déjà vu all over again!"). Recurrent hallucinations or vivid dream-like imagery may also enter the differential diagnosis. A phenomenon of slight confusion in which all is not clear although it is familiar has sometimes been labelled "*près*que *vu*".

Epileptic *déjà vu* is a complex aura of focal onset epilepsy; specifically, it is indicative of temporal lobe onset of seizures, and is said by some authors to be the only epileptic aura of reliable lateralising significance (right). Epileptic *déjà vu* may last longer and be more frequent than other causes, and may be associated with other features such as depersonalization and derealization, strong emotion such as fear, epigastric aura, or olfactory hallucinations. *Déjà vécu* ("already lived") has been used to denote a broader experience than *déjà vu* but the clinical implications are similar.

Déjà vu has also been reported to occur in several psychiatric disorders, such as anxiety, depression, and schizophrenia.

References

Warren-Gash C, Zeman A. Déjà vu. *Pract Neurol*. 2003; **3**: 106–9.
Wild E. Déjà vu in neurology. *J Neurol*. 2005; **252**: 1–7.

Cross References

Aura; Hallucination; *Jamais vu*

Délire Des Négations
- see COTARD'S SYNDROME

Delirium
Delirium, also sometimes known as acute confusional state, acute organic reaction, acute brain syndrome, or toxic-metabolic encephalopathy, is a neurobehavioural syndrome of which the cardinal feature is a deficit of attention, the ability to focus on specific stimuli. Diagnostic criteria also require a concurrent alteration in level of awareness, which may range from lethargy to hypervigilance, although delirium is not primarily a disorder of arousal or alertness (*cf*. coma, stupor, obtundation). Other features commonly observed in delirium include:

- impaired cognitive function: disorientation in time and place.
- perceptual disorders: illusions, hallucinations.
- behavioural disturbances: agitation, restlessness, aggression, wandering, which may occur as a consequence of perceptual problems (hyperalert type); or unresponsiveness, withdrawal (hypoalert or quiet type).
- language: rambling incoherent speech, logorrhoea.
- altered sleep-wake cycle: "sundowning" (restlessness and confusion at night)
- tendency to marked fluctuations in alertness/activity, with occasional lucid intervals.
- delusions: often persecutory.

Hence this abnormal mental state shows considerable clinical heterogeneity. Subtypes or variants are described, one characterised by hyperactivity ("agitated"), the other by withdrawal and apathy ("quiet").

The course of delirium is usually brief (seldom more than a few days, often only hours). On recovery the patient may have no recollection of events, although islands of recall may be preserved, corresponding with lucid intervals (a useful, if retrospective, diagnostic feature).

Delirium is often contrasted with dementia, a "chronic brain syndrome", in which attention is relatively preserved, the onset is insidious rather than acute, the course is stable over the day rather than fluctuating, and which generally lasts months to years. However, it should be noted that delirium is often superimposed on dementia, especially in the elderly. Dementia is a predisposing factor for the development of delirium, perhaps reflecting impaired cerebral reserve.

The pathophysiology of delirium is not well understood. Risk factors for the development of delirium may be categorised as either predisposing or precipitating.

- *Predisposing factors* include:
  - Age: frailty, physiological age rather than chronological.
  - Sex: men > women.
  - Neurological illness: dementia.
  - Burden of co-morbidity; dehydration.
  - Drugs: especially anticholinergic medication.
  - Primary sensory impairment (hearing, vision).
- *Precipitating factors* include:
  - Drugs/toxins: benzodiazepines, opiates.
  - Alcohol, especially withdrawal from, as in delirium tremens.
  - Intercurrent illness:
    - Infection: primary CNS (encephalitis, meningitis), or systemic (urinary tract, chest, septicaemia).
    - Metabolic: hypoxia, hypo-/hyperglycaemia, hepatic failure, uraemia, porphyria.
    - CNS disorders: head injury, cerebrovascular disease, epilepsy (*e.g.* some forms of status), inflammatory disorders (*e.g.* collagen vascular disease).
    - Iatrogenic events: surgery (especially cardiac, orthopaedic).

These precipitating factors merit treatment in their own right, and investigations should be tailored to identify these aetiological factors. The EEG may show non-specific slowing in delirium, the degree of which is said to correlate with the degree of impairment, and reverses with resolution of delirium.

It is suggested that optimal nursing of delirious patients should aim at environmental modulation to avoid both under- and over-stimulation; a side room is probably best (if possible). Drug treatment is not mandatory, the evidence base for pharmacotherapy is slim. However, if the patient poses a risk to him/herself, other patients, or staff which cannot be addressed by other means, regular low dose oral haloperidol may be used, probably in preference to atypical neuroleptics, benzodiazepines (lorazepam), or cholinesterase inhibitors. Prevention of delirium by screening patients for risk factors is advocated.

References

Fong TG, Davis D, Growdon ME, Albuquerque A, Inouye SK. The interface between delirium and dementia in elderly adults. *Lancet Neurol*. 2015; **14**: 823–32.

Inouye SK. Current concepts: delirium in older persons. *N Engl J Med*. 2006; **354**: 1157–65.

Siddiqi N, House AO, Holmes JD. Occurrence and outcome of delirium in medical in-patients: a systematic literature review. *Age Ageing*. 2006; **35**: 350–64.

Cross References

Agraphia; Attention; Coma; Delusion; Dementia; Hallucination; Illusion; Logorrhoea; Obtundation; Stupor; "Sundowning"

Delusion

A delusion is a fixed false belief, not amenable to reason (*i.e*. held despite evidence to the contrary), and not culturally sanctioned. There are a number of common forms of delusion, including:

- persecutory (paranoia).
- reference: important events or people being influenced by a patient's thoughts, ideas.
- grandiose/expansive: occurs particularly in mania.
- guilt/worthlessness: occurs particularly in depression.
- hypochondria.
- thought broadcast and thought insertion.
- control by an external agency.

Specific, named, delusional syndromes are those of:

- *Capgras*: the "delusion of doubles", a familiar person or place is thought to be an impostor, or double; this resembles the reduplicative paramnesia described in neurological disorders such as Alzheimer's disease.
- *Fregoli*: a familiar person is identified in other people, even though they bear no resemblance; this may occur in schizophrenia.
- *De Clérambault* (erotomania): the belief (usually of a single woman) that a famous person is secretly in love with her ("hope"), followed by the belief that that person is persecuting her ("resentment"); may occur in schizophrenia.

Delusions are a feature of primary psychiatric disease (psychoses such as schizophrenia; neuroses such as depression), but may also be encountered in neurological disease with secondary psychiatric features ("organic psychiatry"), *e.g*. delirium, and dementing syndromes such as Alzheimer's disease, dementia with Lewy bodies.

References

Moore DP, Puri BK. Textbook of clinical neuropsychiatry and behavioral neuroscience. 3rd ed. London: Hodder Arnold; 2012.

Tekin S, Cummings JL. Hallucinations and related conditions. In: Heilman KM, Valenstein E, editors. Clinical neuropsychology. 4th ed. Oxford: Oxford University Press; 2003. p. 479–94.

Cross References

Delirium; Dementia; Ekbom's syndrome; Hallucination; Illusion; Intermetamorphosis; Misidentification syndromes; Reduplicative paramnesia

Delusional Parasitosis

- see EKBOM'S SYNDROME

Dementia

Dementia is a syndrome characterised by loss of intellectual (cognitive) functions sufficient to interfere with social and occupational functioning. Cognition encompasses multiple functions including language, memory, perception, praxis, attentional mechanisms and executive function (planning, reasoning). These elements may be affected selectively or globally: older definitions of dementia requiring global cognitive decline have now been superseded. Amnesia may or may not, depending on the classification system used, be a *sine qua non* for the diagnosis of dementia. Attentional mechanisms are largely preserved, certainly in comparison with delirium, a condition which precludes meaningful neuropsychological assessment because of profound attentional deficits. Multiple neuropsychological tests are available to test different areas of cognition.

Although more common in the elderly, dementia can also occur in the presenium, and in children who may lose cognitive skills as a result of hereditary metabolic disorders. Failure to

develop cognitive skills is termed learning disability. The heterogeneity of dementia is further exemplified by the fact that it may be acute or insidious in onset, and its course may be progressive, stable, or, in some instances, reversible ("dysmentia").

A distinction is drawn by some authors between cortical and subcortical dementia: in the former the pathology is predominantly cortical and neuropsychological findings are characterized by amnesia, agnosia, apraxia, and aphasia (*e.g.* Alzheimer's disease); in the latter pathology is predominantly frontal-subcortical and neuropsychological deficits include psychomotor retardation, attentional deficits, with relative preservation of memory and language; movement disorders may also be apparent (*e.g.* progressive supranuclear palsy, Huntington's disease). However, not all authors subscribe to this distinction, and considerable overlap may be observed clinically.

Cognitive deficits also occur in affective disorders such as depression, usually as a consequence of impaired attentional mechanisms. This syndrome is often labelled as "pseudodementia" since it is potentially reversible with treatment of the underlying affective disorder. It may be difficult to differentiate dementia originating from depressive or neurodegenerative disease, since depression may also be a feature of the latter. Impaired attentional mechanisms may account for the common complaint of not recalling conversations or instructions immediately after they happen (aprosexia). Behavioural abnormalities are common in dementias due to degenerative brain disease, and may require treatment in their own right.

Recognised causes of a dementia syndrome include:

- Neurodegenerative diseases:

  Alzheimer's disease, frontotemporal lobar degenerations (encompassing behavioural variant frontotemporal dementia and the agrammatic and semantic variants of primary progressive aphasia, the latter two previously known as primary nonfluent aphasia and semantic dementia), Parkinson's disease dementia; dementia with Lewy bodies, Huntington's disease, progressive supranuclear palsy, corticobasal degeneration, prion disease, Down's syndrome, dementia pugilistica.
- Cerebrovascular disease:

  focal strategic infarcts (*e.g.* paramedian thalamic infarction), multiple infarcts, subcortical vascular disease, Binswanger's disease.
- Inflammatory disorders: multiple sclerosis, systemic lupus erythematosus.
- Structural disease: normal pressure hydrocephalus, subdural haematoma, tumours, dural arteriovenous fistula.
- Infection: HIV dementia, neurosyphilis, Whipple's disease.
- Metabolic causes: Wernicke-Korsakoff syndrome, vitamin B_{12} deficiency, hypothyroidism, hyperparathyroidism/hypercalcaemia, leucodystrophies, Wilson's disease.

Cognitive dysfunction may be identified in many other neurological illnesses.

Investigation of patients with dementia aims to identify its particular cause. Because of the possibility of progression, reversible causes are regularly sought though very rarely found. A focus on early identification at the pre-dementia stage, variously defined as mild cognitive impairment, cognitive impairment no dementia, or mild cognitive disorder, is now promoted, although disease-modifying treatments are lacking. Specific treatments for established dementia are few: cholinesterase inhibitors have been licensed for the treatment of mild to moderate Alzheimer's disease and may find a role in other conditions, such as Parkinson's disease dementia, dementia with Lewy bodies and vascular dementia, for behavioural as well as mnestic features. Memantine is licensed in some jurisdictions for moderate to severe dementia.

References

Ames D, Burns A, O'Brien J, editors. Dementia. 4th ed. London: Hodder Arnold; 2010.

Clarfield AM. The decreasing prevalence of reversible dementia: an updated meta-analysis. *Arch Intern Med*. 2003; **163**: 2219–29.

Dickerson B, Atri A, editors. Dementia. Comprehensive principles and practice. Oxford: Oxford University Press; 2014.
Larner AJ. Neuropsychological neurology. The neurocognitive impairments of neurological disorders. 2nd ed. Cambridge: Cambridge University Press; 2013.
Cross References
Agnosia; Amnesia; Aphasia; Apraxia; Aprosexia; Attention; Delirium; Dysmentia; Pseudodementia; Psychomotor retardation

De Musset's Sign
- see HEAD TREMOR

Depersonalisation
Depersonalisation, a form of dissociation, is the experience of feeling detached or alienated from oneself, such that the body feels strange, lacking control, or being viewed from the outside. There may be concurrent derealisation. Depersonalisation is a very common symptom in the general population, and may contribute to neurological presentations described as dizziness, numbness, and forgetfulness, with the broad differential diagnoses that such symptoms encompass. Such symptoms may also occur in the context of meditation and self-suggestion.
Reference
Stone J. Dissociation: what is it and why is it important? *Pract Neurol*. 2006; **6**: 308–13.
Cross References
Derealization; Dissociation

Derealisation
Derealisation, a form of dissociation, is the experience of feeling that the world around is unreal. There may be concurrent depersonalization.
Reference
Stone J. Dissociation: what is it and why is it important? *Pract Neurol*. 2006; **6**: 308–13.
Cross References
Depersonalsation; Dissociation

Dermatomal Sensory Loss
Dermatome refers to the area of skin innervated by a particualr neural element, such as a nerve root or spinal segment. Mapping out an area of sensory loss or impairment may correspond to a particular dermatome and hence assist in localisation. Few neurology textbooks neglect to include a dermatomal map of the body, but it should be realised that these maps are only approximate and adjacent dermatomes may show significant overlap.
Reference
Apok V, Gurusinghe NT, Mitchell JD, Emsley HCA. Dermatomes and dogma. *Pract Neurol*. 2011; **11**: 100–5.
Cross Reference
Radiculopathy

Dermo-Optical Perception
Dermo-optical perception, or fingertip sight, describes the rare ability to read print, describe pictures, and recognise colours purely by way of touch. This may be a form of paroptic vision; other forms have been decribed, for example though the nose. Experiments have suggested that in some cases fingertip sight for colours may be due to minute differences in surface texture or reflected heat, but correct identification of colours through a glass plate by some subjects would seem to rule out these mechanisms.

Colour-touch synaesthesia might also account for some of these phenomena, whose exact physiology remains uncertain.
Reference
Brugger P, Weiss PH. Dermo-optical perception: the non-synesthetic "palpability of colors" a comment on Larner (2006). *J Hist Neurosci*. 2008; **17**: 253–5.

Cross Reference
Synaesthesia

Developmental Signs
- see FRONTAL RELEASE SIGNS; PRIMITIVE REFLEXES

Diagonistic Dyspraxia
This term referes to a dissociative phenomenon observed after callosotomy, probably identical to intermanual conflict.
Reference
Akelaitis AJ. Studies on the corpus callosum, IV: diagonistic dyspraxia in epileptics following partial and complete section of the corpus callosum. *Am J Psychiatry*. 1944–1945; **101**: 594–9.
Cross References
Alien hand, Alien limb; Intermanual conflict

Diamond on Quadriceps Sign
Diamond on quadriceps sign may be seen in patients with dysferlinopathies (limb girdle muscular dystrophy type 2B, Miyoshi myopathy): with the knees slightly bent so that the quadriceps are in moderate action, an asymmetric diamond shaped bulge may be seen, with wasting above and below, indicative of the selectivity of the dystrophic process in these conditions.
Reference
Pradhan S. Diamond on quadriceps: a frequent sign in dysferlinopathy. *Neurology*. 2008; **70**: 322.
Cross Reference
Calf head sign

Diaphoresis
Diaphoresis describes sweating, either physiological as in sympathetic activation (*e.g.* during hypotension, hypoglycaemia), or pathological (hyperhidrosis, *q.v.*). Diaphoresis may be seen in syncope, delirium tremens, or may be induced by certain drugs (*e.g.* cholinesterase inhibitors) or drug withdrawal (*e.g.* opiates in dependent individuals). Anticholinergics decrease diaphoresis but increase core temperature, resulting in a warm dry patient.
Cross Reference
Hyperhidrosis

Diaphragm Weakness
Diaphragm weakness is a feature of certain myopathies such as acid maltase deficiency, and of cervical cord lesions (C3–C5) affecting phrenic nerve function. Forced vital capacity measured in the supine and sitting positions is often used to assess diaphragmatic function, a drop of 25% being taken as indicating diaphragmatic weakness.
Reference
Allen SM, Hunt B, Green M. Fall in vital capacity with posture. *Br J Diseases Chest*. 1985; **79**: 267–71.
Cross Reference
Paradoxical breathing

Digital Reflex
- see HOFFMANN'S SIGN; TRÖMNER'S SIGN

Diplophonia
Diplophonia, the simultaneous production of two pitch levels when phonating, occurs in unilateral vocal cord paralysis because each vocal fold has a different vibration frequency.
Cross References
Bovine cough; Dysphonia

Diplopia

Diplopia is double vision, *viz*., seeing two images of a single object. The spatial and temporal characteristics of the diplopia may help to ascertain its cause.

Diplopia may be monocular, in which case ocular causes are most likely (although monocular diplopia may be cortical or functional in origin), or binocular, implying a divergence of the visual axes of the two eyes. With binocular diplopia, it is of great importance to ask the patient whether the images are separated horizontally, vertically, or obliquely (tilted), since this may indicate the extraocular muscle(s) most likely to be affected. Whether the two images are separate or overlapping is important when trying to ascertain the direction of maximum diplopia.

The experience of diplopia may be confined to, or particularly noticeable during, the performance of particular activities, reflecting the effect of gaze direction; for example, diplopia experienced on coming downstairs may reflect a trochlear (IV) nerve palsy; or only on looking to the left may reflect a left abducens (VI) nerve palsy. Double vision experienced on looking at a distant object after looking down (*e.g*. reading) may occur with bilateral abducens (VI) nerve palsies. The effect of gaze direction on diplopia should always be sought, since images are most separated when looking in the direction of a paretic muscle. Conversely, diplopia resulting from the breakdown of a latent tendency for the visual axes to deviate (latent strabismus, squint) results in diplopia in all directions of gaze.

Examination of the eye movements should include asking the patient to look at a target, such as a pen, in the various directions of gaze (versions) to ascertain where diplopia is maximal. Ductions are tested monocularly with the opposite eye covered. Then, each eye may be alternately covered to try to demonstrate which of the two images is the false one, namely that from the non-fixing eye. The false image is also the most peripheral image. Thus in a left abducens (VI) nerve palsy, diplopia is maximum on left lateral gaze; when the normal right eye is covered the inner image disappears; the non-fixing left eye is responsible for the remaining false image, which is the more peripheral and which disappears when the left eye is covered.

Other clues to the cause of diplopia include the presence of any other neurological signs, for example ptosis (unilateral: oculomotor (III) nerve palsy; bilateral: myasthenia gravis), or head tilt or turn (to the right suggests a weak right lateral rectus muscle suggesting a right abducens (VI) nerve palsy; tilt to the left shoulder suggests a right trochlear (IV) nerve palsy, = Bielschowsky's sign).

Manifest squints (heterotropia) are obvious but seldom a cause of diplopia if longstanding. Latent squints may be detected using the cover-uncover test, when the shift in fixation of the eyes indicates an imbalance in the visual axes; this may account for diplopia if the normal compensation breaks down. This produces diplopia in all directions of gaze (comitant). Patients may with an effort be able to fuse the two images.

Transient diplopia (minutes to hours) suggests the possibility of myasthenia gravis. There are many causes of persistent diplopia, including the breakdown of a latent strabismus, development of oculomotor (III), trochlear (IV) or abducens (VI) nerve palsy (singly or in combination), orbital myopathy (thyroid), and mass lesions of the orbit (tumour, pseudotumour).

Divergence of the visual axes or ophthalmoplegia without diplopia suggests a longstanding problem, such as amblyopia or chronic progressive external ophthalmoplegia. Some eye movement disorders are striking for the lack of associated diplopia, *e.g*. internuclear ophthalmoplegia.

References

Danchaivijitr C, Kennard C. Diplopia and eye movement disorders. *J Neurol Neurosurg Psychiatry*. 2004; **75**(Suppl IV): iv24–31.

Low L, Shah W, MacEwen CJ. Double vision. *BMJ*. 2015; **351**: h5385.

Rucker JC, Tomsak RL. Binocular diplopia. A practical approach. *Neurologist*. 2005; **11**: 98–110.

Yee RD. Approach to the patient with diplopia. In: Biller J, editor. Practical neurology. 2nd ed. Philadelphia: Lippincott Williams & Wilkins; 2002. p. 147–61.

Cross References
Abducens (VI) nerve palsy; Amblyopia; Bielschowsky's sign, Bielschowsky's test; Cover tests; Heterophoria; Heterotropia; Internuclear ophthalmoplegia (INO); Oculomotor (III) nerve palsy

Directional Hypokinesia

Directional hypokinesia describes a reluctance to move towards contralesional space seen in the neglect syndrome.

Cross Reference
Motor neglect; Neglect

Disc Swelling

Swelling or oedema of the optic nerve head may be visualized by ophthalmoscopy. It produces haziness of the nerve fibre layer obscuring the underlying vessels; there may also be disc haemorrhages and loss of spontaneous retinal venous pulsation at the disc margin. Whether vision is affected is dependent upon the precise cause of disc swelling.

The clinical history, visual acuity and visual fields may help determine the cause of disc swelling.

Disc swelling due to raised intracranial pressure (papilloedema, *q.v.*) may occur without specific visual complaint but with an enlarged blind spot on visual field testing. Local inflammation of the optic nerve (papillitis) may be associated with marked impairment of vision, as for example in optic neuritis.

Disc swelling due to oedema must be distinguished from pseudopapilloedema, elevation of the optic disc not due to oedema, in which the nerve fibre layer is clearly seen.

Recognised causes of disc swelling include:

- *Unilateral*:
  Optic neuritis.
  Acute ischaemic optic neuropathy (arteritic, non-arteritic).
  Orbital compressive lesions, *e.g.* optic nerve sheath meningioma (Foster Kennedy syndrome).
  Graves' ophthalmopathy (through compression of retinal veins by myositis).
  Central retinal vein occlusion.
  Infiltration: carcinoma, lymphoma, granuloma.
  Raised intracranial pressure (papilloedema; more usually bilateral).
- *Bilateral*:
  Raised intracranial pressure (papilloedema).
  Malignant hypertension.
  Hypercapnia.
  High CSF protein, as in Guillain-Barré syndrome.
  Any of the unilateral causes.

Cross References
Foster Kennedy syndrome; Papilloedema; Pseudopapilloedema; Retinal venous pulsation; Visual field defects

Disinhibition

Disinhibited behaviour is impulsive, showing poor judgment and insight, and may transgress normal cultural or social bounds. There is a loss of normal emotional and/or behavioural control. The disinhibited patient may be inappropriately jocular (*witzelsucht*), short-tempered (verbally abusive, physically aggressive), distractible (impaired attentional mechanisms), and show emotional lability. A Disinhibition Scale encompassing various domains (motor, intellectual, instinctive, affective, sensitive) has been described.

Disinhibition is a feature of frontal lobe, particularly orbitofrontal, dysfunction. This may be due to neurodegenerative disorders (behavioural variant frontotemporal dementia, Alzheimer's disease), mass lesions, or be a feature of epileptic seizures.

Cross References

Attention; Emotionalism, Emotional lability; Frontal lobe syndromes; Osculation; *Witzelsucht*

Dissociated Sensory Loss

Dissociated sensory loss refers to impairment of selected sensory modalities with preservation, or sparing, of others. It is usually an indication of an intramedullary spinal cord lesion. For example, a focal central cord pathology such as syringomyelia will, in the early stages, selectively involve decussating fibres of the spinothalamic pathway within the ventral commissure, thus impairing pain and temperature sensation (often in a suspended, "cape-like", "bathing suit", "vest-like", or cuirasse distribution), whilst the dorsal columns are spared, leaving proprioception intact. The anterior spinal artery syndrome also leaves the dorsal columns intact. Conversely, pathologies confined, largely or exclusively, to the dorsal columns (classically tabes dorsalis and subacute combined degeneration of the cord from vitamin B_{12} deficiency, but probably most commonly seen with compressive cervical myelopathy) impair proprioception, sometimes sufficient to produce pseudoathetosis or sensory ataxia, whilst pain and temperature sensation is preserved. A double dissociation of sensory modalities on opposite sides of the trunk is seen in the Brown-Séquard syndrome.

Small fibre peripheral neuropathies may selective affect the fibres which transmit pain and temperature sensation, leading to a glove-and-stocking impairment to these modalities. Neuropathic (Charcot) joints and skin ulceration may occur in this situation; tendon reflexes may be preserved.

Cross References

Analgesia; Ataxia; Brown-Séquard syndrome; Charcot joint; *Main succulente*; Myelopathy; Proprioception; Pseudoathetosis; Sacral sparing

Dissociation

Dissociation is an umbrella term for a wide range of symptoms involving feelings of disconnection from the body (depersonalsation) or the surroundings (derealization). Common in psychiatric disorders (depression, anxiety, schizophrenia), these symptoms are also encountered in neurological conditions (epilepsy, migraine, presyncope), conditions such as functional weakness and non-epilpetic attacks, and in isolation by a significant proportion of the general population. Symptoms of dizziness and blankness may well be the result of dissociative states rather than neurological disease.

Reference

Stone J. Dissociation: what is it and why is it important? *Pract Neurol*. 2006; **6**: 308–13.

Cross References

Depersonalsation; Derealization

Divisional Palsy

The oculomotor (III) nerve divides into superior and inferior divisions, usually at the superior orbital fissure. The superior division or ramus supplies the superior rectus and levator palpebrae superioris muscles; the inferior division or ramus supplies medial rectus, inferior rectus and inferior oblique muscles. Isolated dysfunction of these muscle groups allows diagnosis of divisional palsy and suggests pathology at the superior orbital fissure or anterior cavernous sinus. However, occasionally the division may occur more proximally, at the fascicular level (*i.e*. within the midbrain) or within the subarachnoid space, giving a false-localising divisional palsy. This may reflect the topographic arrangement of axons within the oculomotor nerve.

Reference
Larner AJ. Proximal superior division oculomotor nerve palsy from metastatic subarachnoid infiltration. *J Neurol*. 2002; **249**: 343–4.
Cross References
"False-localising signs"; Oculomotor (III) nerve palsy

Dix-Hallpike Positioning Test
- see HALLPIKE MANOEUVRE, HALLPIKE TEST

Doll's Eye Manoeuvre, Doll's Head Manoeuvre
This test of the vestibulo-ocular reflex (VOR) is demonstrated by rotating the patient's head and looking for a conjugate eye movement in the opposite direction. Although this can be done in a conscious patient focusing on a visual target, smooth pursuit eye movements may compensate for head turning; hence the head impulse test (*q.v.*) may be required. The manoeuvre is easier to do in the unconscious patient, when testing for the integrity of brainstem reflexes.

A slow (0.5–1.0 Hz) doll's head manoeuvre may be used in conscious patients to assess vestibulo-ocular reflexes. Whilst directly observing the eyes, "catch up" saccades may be seen in the absence of VOR. Measuring visual acuity with head movement compared to visual acuity with the head still (dynamic visual acuity, or illegible E test), two to three lines may be dropped if VOR is impaired. On ophthalmoscopy, the disc moves with the head if VOR is lost.
Reference
Roberts TA, Jenkyn LR, Reeves AG. On the notion of doll's eyes. *Arch Neurol*. 1984; **41**: 1242–3.
Cross References
Bell's phenomenon, Bell's sign; Caloric testing; Coma; Head impulse test; Oculocephalic response; Supranuclear gaze palsy; Vestibulo-ocular reflexes

"Dorsal Guttering"
Dorsal guttering refers to the marked prominence of the extensor tendons on the dorsal surface of the hand when intrinsic hand muscles (especially interossei) are wasted, as may occur in an ulnar nerve lesion, a lower brachial plexus lesion, or a T1 root lesion. Benign extramedullary tumours at the foramen magnum may also produce this picture (remote atrophy, a "false-localising sign"). In many elderly people the extensor tendons are prominent in the absence of significant muscle wasting.
Cross References
"False-localising signs"; Wasting

"Double Elevator Palsy"
This name has been given to monocular elevation paresis (apparent hypotropia). This may be congenital or acquired, the latter may occur in association with pretectal supranuclear lesions (*e.g.* pineal mass lesion) either contralateral or ipsilateral to the paretic eye interrupting efferents from the rostral interstitial nucleus of the medial longitudinal fasciculus to the superior rectus and inferior oblique subnuclei. Bell's phenomenon may be preserved.
Reference
Thömke F, Hopf HC. Acquired monocular elevation paresis. An asymmetric up-gaze palsy. *Brain*. 1992; **115**: 1901–10.
Cross References
Bell's phenomenon, Bell's sign; Hypotropia

Downbeat Nystagmus
- see NYSTAGMUS

Dressing Apraxia
- see APRAXIA

Drooling

- see SIALORRHOEA

Dropped Head Syndrome

Dropped head syndrome (head droop or head drop) refers to forward flexion of the head on the neck, such that the chin falls on to the chest (*cf*. antecollis) and the head cannot be voluntarily extended. This syndrome has a broad differential diagnosis, encompassing disorders which may cause axial truncal muscle weakness, especially of upper thoracic and paraspinous muscles.

- Neuropathy/Neuronopathy:
  Motor neurone disease (the author has also seen this syndrome in a patient with frontotemporal lobar degeneration with motor neurone disease, FTLD/MND).
  Guillain-Barré syndrome, chronic inflammatory demyelinating polyneuropathy.
  Paraneoplastic motor neuronopathy.
- Neuromuscular junction disorder:
  Myasthenia gravis.
- Myopathy:
  Polymyositis.
  Myotonic dystrophy.
  Myopathy with rimmed vacuoles.
  "Dropped head syndrome", or "isolated neck extensor myopathy", a condition of uncertain aetiology but which may on occasion be steroid-responsive ("bent spine syndrome" or camptocormia may be related forms of axial myopathy).
- Extrapyramidal disorders:
  Parkinson's disease.
  Multiple system atrophy.
  Progressive supranuclear palsy.

Of these, probably MND and myasthenia gravis are the most common causes.

Treatment of the underlying condition may be possible, hence investigation is mandatory. If not treatable (*e.g*. MND), a head brace may keep the head upright.

References

Katz JS, Wolfe GI, Burns DK, Bryan WW, Fleckenstein JL, Barohn RJ. Isolated neck extensor myopathy. A common cause of dropped head syndrome. *Neurology*. 1996; **46**: 917–21.

Nicholas RS, Lecky BRF. Dropped head syndrome: the differential diagnosis. *J Neurol Neurosurg Psychiatry*. 2002; **73**: 218 (abstract 26).

Rose MR, Levin KH, Griggs RC. The dropped head plus syndrome: quantitation of response to corticosteroids. *Muscle Nerve*. 1999; **22**: 115–8.

Swash M. Dropped-head and bent-spine syndromes: axial myopathies? *Lancet*. 1998; **352**: 758.

Cross References

Antecollis; Camptocormia; Myopathy

Drusen

Drusen are hyaline bodies that are typically seen on and around the optic nerve head, and may be mistaken for papilloedema ("pseudopapilloedema"). Drusen are thought to result from altered axonal flow with axonal degeneration. They occur sporadically or may be inherited in an autosomal dominant fashion, and are common, occurring in 2% of the population. In children the drusen are buried whilst in adults they are on the surface of the disc.

Drusen are usually asymptomatic but can cause visual field defects (typically an inferior nasal visual field loss) or occasionally transient visual obscurations, but not changes in visual acuity which require investigation for an alternative cause. When there is doubt whether

papilloedema or drusen is the cause of a swollen optic nerve head, retinal fluorescein angiography is required.

Reference

Arbabi EM, Fearnley TE, Carrim ZI. Drusen and the misleading optic disc. *Pract Neurol*. 2010; **10**: 27–30.

Cross References

Disc swelling; Papilloedema; Pseudopapilloedena; Visual field defects

Durkan's Compression Test

Durkan's compression test, sometimes called the carpal compression test, is a provocative test for carpal tunnel syndrome. With the wrist in a supine position on a table, the examiner places three fingers over the carpal tunnel and compresses the area for 30 s. If the patient reports tingling, numbness, or altered sensation in the thumb or index finger, middle finger, or radial half of the ring finger, then the test is positive and suggestive of a diagnosis of carpal tunnel syndrome.

Cross References

Phalen's sign; Tinel's sign

Dynamic Aphasia

Dynamic aphasia refers to an aphasia characterized by difficulty initiating speech output, ascribed to executive dysfunction. There is a reduction in spontaneous speech, but on formal testing there are no paraphasias, minimal anomia, preserved repetition, reading, and automatic speech. "Incorporational echolalia", when the patient uses the examiner's question to help form an answer, may be observed.

Dynamic aphasia has been conceptualised as a variant of transcortical motor aphasia, and may be seen with lesions of dorsolateral prefrontal cortex ("frontal aphasia"). It has also been reported in progressive supranuclear palsy, and postulated to be a variant of primary progressive aphasia. A division into pure and mixed forms has been suggested, with additional phonological, lexical, syntactical and articulatory impairments in the latter.

Reference

Robinson GA. Primary progressive dynamic aphasia and Parkinsonism: generation, selection and sequencing deficits. *Neuropsychologia*. 2013; **51**: 2534–47.

Cross References

Aphasia; Echolalia; Transcortical aphasias

Dysaesthesia

Dysaesthesia (or dysesthesia) refers to an unpleasant, abnormal or unfamiliar, sensation, often with a burning and/or "electrical" quality. Some authorities reserve the term for provoked positive sensory phenomena, as opposed to spontaneous sensations (paraesthesia). Dysaesthesia differs from paraesthesia in its unpleasant quality, but may overlap in some respects with allodynia, hyperalgesia and hyperpathia (the latter phenomena are provoked by stimuli, either non-noxious or noxious).

There are many causes of dysaesthesia, both peripheral (including small fibre neuropathies, neuroma, nerve trauma) and central (*e.g.* spinal multiple sclerosis).

Dysaesthetic sensations may be helped by agents such as carbamazepine, amitriptyline, gabapentin and pregabalin.

Cross References

Allodynia; Hyperalgesia; Hyperpathia; Paraesthesia

Dysarthria

Dysarthria is a disorder of speech, as opposed to language (*cf.* aphasia), because of impairments in the actions of the speech production apparatus *per se*, due to paralysis, ataxia, tremor or spasticity, in the presence of intact mental function, comprehension and memory for words. In its most extreme form, anarthria, there is no speech output.

Dysarthria is a symptom which may be caused by a number of different conditions, all of which ultimately affect the function of pharynx, palate, tongue, lips and larynx, be that at the level of the cortex, lower cranial nerve nuclei or their motor neurones, neuromuscular junction or bulbar muscles themselves. Dysarthrias affect articulation in a highly reliable and consistent manner, the errors reflecting the muscle group involved in the production of specific sounds. There are various syndromes of dysarthria, which have been classified as follows:

- *Flaccid or nasal dysarthria*:

  hypernasal, breathy, whining output, as in bulbar palsy, *e.g.* myasthenia gravis.
- *Spastic dysarthria*:

  slow, strained ("strangled") output, monotonous, as in pseudobulbar palsy; may coexist with Broca's aphasia.
- *Ataxic or cerebellar dysarthria*:

  altered rhythm of speech, uneven irregular output, slurred speech (as if inebriated), improper stresses; seen in acute cerebellar damage due to asynergia of speech muscle contractions (*cf.* scanning speech).
- *Hypokinetic dysarthria*:

  monotonic pitch, hypophonic volume, as in parkinsonism.
- *Hyperkinetic dysarthria*:

  several varieties are described, including choreiform (as in Huntington's disease), dystonic (as in tardive dyskinesia, and other dystonic syndromes), tremulous (tremor syndromes), and the dysarthria with vocal tics (including coprolalia) in Tourette syndrome.
- *Mixed dysarthria*:

  combination of any of above.

Recognised causes of dysarthria include:

- Muscle disease:

  e.g. oculopharyngeal muscular dystrophy: nasal speech; weak pharynx/drooling.
- Neuromuscular disorder:

  e.g. myasthenia gravis: nasal speech; fatiguability (development of hypophonia with prolonged conversation, or counting).
- Lower motor neurone disease = bulbar palsy:

  e.g. motor neurone disease (rasping monotones, wasted and fasciculating tongue), poliomyelitis, Guillain-Barré syndrome, diphtheria.
- Upper motor neurone disease = pseudobulbar palsy:

  e.g. motor neurone disease (spastic tongue), cerebrovascular disease.
- Cortical dysarthria:

  damage to left frontal cortex, usually with associated right hemiparesis; may be additional aphasia.
- Extrapyramidal disease:

  e.g. hypokinetic disorders: Parkinson's disease: slow, hypophonic, monotonic; multiple system atrophy (may have vocal cord palsy).
  e.g. hyperkinetic disorders: Huntington's disease: loud, harsh, variably stressed, and poorly co-ordinated with breathing; myoclonus of any cause (hiccup speech); dystonia of any cause.
- Ataxic dysarthria:

  disease of or damage to the cerebellum: slow, slurred, monotonous, with incoordination of speech with respiration; may therefore be quiet and then explosive; unnatural separation of syllables; slow tongue movements.

- Acquired stuttering:

  involuntary repetition of letters or syllables, may be acquired with aphasia; developmental stutter, the more common cause, usually affects the beginnings of words and with plosive sounds, whereas the acquired form may be evident throughout sentences and affect all speech sounds.

Treatment of the underlying cause may improve dysarthria (*e.g.* nasal dysarthria of myasthenia gravis). Baclofen has been suggested for dysarthria of upper motor neurone type. Speech and language therapy may provide symptomatic benefit.

References

Darley FL, Aronson AE, Brown JR. Motor speech disorders. Philadelphia: Saunders; 1975.
LaMonte MP, Erskine MC, Thomas BE. Approach to the patient with dysarthria. In: Biller J, editor. Practical neurology. 2nd ed. Philadelphia: Lippincott Williams & Wilkins; 2002. p. 236–43.
Murdoch BE, editor. Dysarthria. A physiological approach to assessment and treatment. Cheltenham: Stanley Thornes; 1998.

Cross References

Anarthria; Aphasia; Asynergia; Broca's aphasia; Bulbar palsy; Coprolalia; Dysphonia; Fatigue; Lower motor neurone (LMN) syndrome; Parkinsonism; Pseudobulbar palsy; Scanning speech; Stutter; Upper motor neurone (UMN) syndrome

Dysautonomia

Dysautonomia describes autonomic nervous system dysfunction which may result from either pre- or post-ganglionic lesions of either the sympathetic or parasympathetic pathways, or both (pandysautonomia).

Clinical features of dysautonomia include:

- Visual blurring; pupillary areflexia.
- Orthostatic hypotension.
- Cardiac arrhythmia.
- Abdominal pain, diarrhoea, vomiting, constipation, ileus, pseudo-obstruction.
- Sweating dysfunction (*e.g.* anhidrosis).

Autonomic dysfunction may be:

- Congenital.
- Acquired:

  acute (*e.g.* after a viral infection such as infectious mononucleosis).
  subacute (*e.g.* the "autonomic-only" form of Guillain-Barré syndrome).
  chronic (*e.g.* pure autonomic failure, multiple system atrophy, certain hereditary neuropathies).

As regards aetiology, in addition to hereditary (genetic) and neurodegenerative disorders, autoimmune forms of autonomic neuropathy are described, for example a pandysautonomia associated with antibodies to ganglionic neuronal nicotinic acetylcholine receptors (*cf.* antibodies to muscarinic acetylcholine receptors in myasthenia gravis) which may respond well to immunomodulatory therapies.

References

Goldstein DS, Holmes C, Dendi R, Li ST, Brentzel S, Vernino S. Pandysautonomia associated with impaired ganglionic neurotransmission and circulating antibody to the neuronal nicotinic receptor. *Clin Auton Res*. 2002; **12**: 281–5.
Mathias CJ, Bannister R, editors. Autonomic failure. A textbook of clinical disorders of the autonomic nervous system. 5th ed. Oxford: Oxford University Press; 2013.

Dyscalculia
- see ACALCULIA

Dyschromatopsia
- see ACHROMATOPSIA

Dysdiadochokinesia
Dysdiadochokinesia, or adiadochokinesia, is a difficulty in performing rapid alternating movements, for example pronation/supination of the arms, tapping alternately with the palm and dorsum of the hand, tapping the foot on the floor.

Dysdiadochokinesia is a sign of cerebellar dysfunction, especially hemisphere disease, and may be seen in association with asynergia, ataxia, dysmetria, and excessive rebound phenomenon. It may reflect the impaired checking response seen in cerebellar disease. Dysdiadochokinesia may also be seen with disease of the frontal lobes ("frontal apraxia") or basal ganglia.

Cross References
Asynergia; Apraxia; Ataxia; Cerebellar syndromes; Dysmetria; Rebound phenomenon

Dysexecutive Syndrome
The term executive function encompasses a range of cognitive processes including sustained attention, fluency and flexibility of thought, problem solving skills, planning and regulation of adaptive and goal-directed behaviour. Some authors prefer to use these individual terms, rather than "lump" them together as executive function. Deficits in these various functions, the dysexecutive syndrome, are typically seen with lateral prefrontal cortex lesions.

Reference
Knight RT, D'Esposito M. Lateral prefrontal syndrome: a disorder of executive control. In: D'Esposito M, editor. Neurological foundations of cognitive neuroscience. Cambridge: MIT Press; 2003. p. 259–79.

Cross References
Attention; Frontal lobe syndromes

Dysgeusia
Dysgeusia is a complaint of distorted taste perception. It may occur along with anosmia as a feature of upper respiratory tract infections, and has also been described with various drug therapies, in psychiatric diseases, and as a feature of zinc deficiency.

Reference
Henkin RI, Patten BM, Pe RK, Bronzert DA. A syndrome of acute zinc loss. Cerebellar dysfunction, mental changes, anorexia and taste and smell dysfunction. *Arch Neurol*. 1975; **32**: 745–51.

Cross References
Ageusia; Anosmia

Dysgraphaesthesia
- see AGRAPHOGNOSIA; GRAPHAESTHESIA

Dysgraphia
- see AGRAPHIA

Dyskinesia
Dyskinesia may be used as a general term for excessive involuntary movements, encompassing tremor, myoclonus, chorea, athetosis, tics, stereotypies, and hyperekplexia. The term may be qualified to describe a number of other syndromes of excessive movement, *e.g.*:

- *Drug-induced dyskinesia*:

  Fluid, restless, fidgety movements seen in patients with Parkinson's disease after several years of levodopa therapy, and often described according to their relationship to timing of tablets (*e.g.* peak dose, diphasic), although others are unpredictable (freezing, yo-yo-ing). In MPTP-induced parkinsonism, dyskinesias tend to occur early, hence it may be the depth of dopamine deficiency rather than chronicity of treatment which is the key determinant; reduction in overall levodopa use (increased frequency of smaller doses, controlled-release preparations, addition of dopamine agonists) may reduce these effects; amantadine is sometimes helpful.

- *Tardive dyskinesia*:

  A form of drug-induced dyskinesia developing after long-term use of neuroleptic (dopamine antagonist) medication, typically involving orolingual musculature (buccolingual syndrome, rabbit syndrome, "bon-bon sign") and occasionally trunk and arms; usually persists after withdrawal of causative therapy; clonazepam, baclofen, and tetrabenazine may sometimes help.

- *Paroxysmal dyskinesias*:

  Paroxysmal kinesigenic choreoathetosis/dystonia (PKC; usually responds to carbamazepine), and paroxysmal non-kinesigenic dystonia/choreoathetosis (PDC; does not respond to carbamazepine).

- *Focal dyskinesias*:

  Orofacial dyskinesia, belly-dancer's dyskinesia, moving ear syndrome.

References

Fahn S. The paroxysmal dyskinesias. In: Marsden CD, Fahn S, editors. Movement disorders. 3rd ed. Oxford: Butterworth-Heinemann; 1994. p. 310–45.

Schelosky LD. Paroxysmal dyskinesias. In: Schmitz B, Tettenborn B, Schomer DL, editors. The paroxysmal disorders. Cambridge: Cambridge University Press; 2010. p. 113–29.

Wojcieszek J. Drug-induced movement disorders. In: Biller J, editor. Iatrogenic neurology. Boston: Butterworth-Heinemann; 1998. 215–31.

Cross References

Athetosis; "Bon-bon sign"; Chorea, Choreoathetosis; Dystonia; Hyperekplexia; Moving ear; Myoclonus; Parkinsonism; Stereotypy; Tic; Yo-yo-ing

Dyslexia

Dyslexia is difficulty or impairment in reading, usually applied to developmental abnormalities of reading ability. A loss of previously acquired reading ability is probably better termed alexia.

Cross Reference

Alexia

Dysmentia

The term dysmentia has been suggested as an alternative to dementia, to emphasize the possibility that cognitive impairment and decline may be amenable to treatment and prevention, thereby reversing the therapeutic nihilism sometimes associated with the diagnostic label of dementia.

Reference

Chiu E. What's in a name: dementia or dysmentia? *Int J Geriatr Psychiatry*. 1994; **9**: 1–4.

Cross Reference

Dementia

Dysmetria

Dysmetria, or past-pointing, is a disturbance in the control of range of movement in voluntary muscular action, and is one feature of the impaired checking response seen in cerebellar lesions (especially cerebellar hemisphere lesions).

Dysmetria may also be evident in saccadic eye movements: hypometria (undershoot) is common in parkinsonism; hypermetria (overshoot) is more typical of cerebellar disease (lesions of dorsal vermis and fastigial nuclei).

In cerebellar disorders, dysmetria reflects the asynergia of co-ordinated muscular contraction.

References

Bötzel K, Rottach K, Büttner U. Normal and pathological saccadic dysmetria. *Brain*. 1993; **116**: 337–53.

Büttner U, Straube A, Spuler A. Saccadic dysmetria and "intact" smooth pursuit eye movements after bilateral deep cerebellar nuclei lesions. *J Neurol Neurosurg Psychiatry*. 1994; **57**: 832–4.

Cross References

Asynergia; Cerebellar syndromes; Dysdiadochokinesia; Parkinsonism; Rebound phenomenon; Saccades

Dysmorphopsia

The term dysmorphopsia has been proposed for impaired vision for shapes, a visual recognition defect in which visual acuity, colour vision, tactile recognition and visually-guided reaching movements are intact. These phenomena have been associated with bilateral lateral occipital cortical damage (*e.g*. after carbon monoxide poisoning) and are thought to reflect a selective loss of the magnocellular visual pathway. Whether this condition is an agnosia for shape or visual form, or a perceptual problem ("pseudoagnosia"), remains a subject of debate and the term dysmorphopsia has been suggested as a compromise between the different strands of thought.

Reference

Milner AD, Perrett DI, Johnston RS, et al. Perception and action in "visual form agnosia". *Brain*. 1991; **114**: 405–28.

Cross References

Agnosia; Visual agnosia; Visual form agnosia

Dysnomia

- see ANOMIA

Dysphagia

Dysphagia is difficulty swallowing. This may have local mechanical causes which are usually gastroenterological in origin (tumour; peptic ulceration/stricture, in which case there may be additional pain on swallowing – odynophagia) but sometimes vascular (aberrant right subclavian artery – dysphagia lusoria) or due to connective tissue disease (systemic sclerosis).

Dysphagia of neurological origin may be due to pathology occurring anywhere from cerebral cortex to muscle. Neurological control of swallowing is bilaterally represented and so unilateral upper motor neurone lesions may cause only transient problems. Poststroke dysphagia is common, but there is evidence of cortical reorganization (neuroplasticity) underpinning recovery. Bilateral upper motor neurone lesions cause persistent difficulties. Dysphagia of neurological origin may be accompanied by dysphonia, palatal droop, and depressed or exaggerated gag reflex.

Dysphagia may be:

- *Neurogenic*:
  - CNS:
    - Cerebrovascular disease: hemisphere, brainstem stroke.
    - Extrapyramidal disease: Parkinson's disease, progressive supranuclear palsy, Huntington's disease, Wilson's disease, tardive dyskinesia, dystonia.

Inflammatory disease: multiple sclerosis.
Neoplasia: primary, secondary; cerebral, brainstem (skull base).
Other structural disorders of the brainstem: syringobulbia, cerebellar disease.
Developmental disorders: cerebral palsy syndromes, Chiari malformations.

Neuronopathy:

Motor neurone disease.

Neuropathy:

Guillain-Barré syndrome.
Autonomic neuropathy (diabetes mellitus, amyloidosis, Chagas' disease, autonomic failure, Riley Day syndrome).
Lower motor neurone pathology: bulbar palsy, isolated vagus (X) nerve palsy, jugular foramen syndrome.

Neuromuscular:

Myasthenia gravis.

- *Myogenic*:

Inflammatory muscle disease: polymyositis, inclusion body myositis.
Myotonia: myotonic dystrophy.
Muscular dystrophy: oculopharyngeal muscular dystrophy.
Symptomatic oesophageal peristalsis ("nutcracker oesophagus").

- *Functional*:

"Hysterical", globus hystericus (diagnosis of exclusion).

Gastrointestinal causes of dysphagia include:

- *Intrinsic*:

Oesophageal carcinoma.
Metastatic or extrinsic tumour spread.
Peptic (post-inflammatory) stricture.
Hiatus hernia.

- *Extrinsic*:

Thoracic aortic aneurysm.
Abnormal origin of right subclavian artery (dysphagia lusoria).
Posterior mediastinal mass.
Large goitre.
Retropharyngeal mass.

If swallowing is compromised with a risk of aspiration, feeding may need to be undertaken via nasogastric tube, percutaneous gastrostomy or jejunostomy placed endoscopically (PEG or PEJ), or even parenterally.

Reference

Abdel Jalil AA, Katzka DA, Castell DO. Approach to the patient with dysphagia. *Am J Med.* 2015; **128**: 1138.e17–23.

Cross References

Bulbar palsy; Dysphonia; Gag reflex; Jugular foramen syndrome; Pseudobulbar palsy

Dysphasia

"The inaccuracy of applying an absolute negation [i.e. aphasia] to a partial effect [of language] has led to the suggestion of 'dysphasia' as a substitute. The term does not, however, seem likely to come into use, a matter of little regret, since the word has not the merit of unimpeachable exactness, and it has an unfortunate resemblance in sound to 'dysphagia'."

Reference

Gowers WR. Manual of diseases of the nervous system. Vol. 2; 2nd ed. London: J&A Churchill; 1893. p. 110n1

Cross Reference
Aphasia

Dysphonia
Dysphonia is a disorder of the volume, pitch or quality of the voice resulting from dysfunction of the larynx, *i.e*. a disorder of phonation or sound generation. Hence this is a motor speech disorder and could be considered as a type of dysarthria if of neurological origin.

Dysphonia manifests as hoarseness, or a whispering breathy quality to the voice. Diplophonia may occur. At the extreme, there may be complete loss of the voice (aphonia).

Recognised causes of dysphonia include:

- infection (laryngitis).
- structural abnormalities, *e.g*. polyp, nodule, papilloma of vocal cord.
- neurological causes:

  Focal dystonic syndrome: spasmodic dysphonia or laryngeal dystonia (either abductor or adductor); the voice may have a strained and harsh quality, with low volume and pitch, vocal tremor, and irregularly distributed stoppages; with continuing speech, or if holding a single note, the voice may fade away entirely. These syndromes may be amenable to treatment with botulinum toxin.

  Flaccid dysphonia, due to superior laryngeal nerve or vagus nerve (recurrent laryngeal nerve) palsy, bulbar palsy.

Reference
Blitzer A, Brin MF, Stewart CF. Botulinum toxin management of spasmodic dysphonia (laryngeal dystonia): a 12-year experience in more than 900 patients. *Laryngoscope*. 2015; **125**: 1751–7.

Cross References
Aphonia; Bulbar palsy; Diplophonia; Dysarthria; Dystonia; Hypophonia; Vocal tremor, Voice tremor

Dyspraxia
Dyspraxia is difficulty or impairment in the performance of a skilled voluntary motor act despite an intact motor system and level of consciousness. This may be developmental in origin ("clumsy child"), but in adult practice reflects a loss of function, hence apraxia is a better term.

Cross Reference
Apraxia

Dysprosodia, Dysprosody
- see APROSODIA, APROSODY

Dyssynergia
- see ASYNERGIA

Dystaxia
- see ATAXIA

Dystextia
Dystextia is a neologism coined to describe difficulty writing mobile phone texts. Acute dystextia has been described in the context of dominant hemisphere stroke with concurrent aphasia and in migraine. It may reflect not only linguistic difficulties but also visual and motor problems (as for agraphia). The tendency of predictive text functions to garble meaning, and the peculiar argot used by frequent users of mobile texts, should not be forgotten when considering the possibility of dystextia.

References
Burns B, Randall M. "Dystextia": onset of difficulty writing mobile phone texts determines the time of acute ischaemic stroke allowing thrombolysis. *Pract Neurol*. 2014; **14**: 256–7.
Cawood TJ, King T, Sreenan S. Dystextia – a sign of the times? *Irish Med J*. 2006; **99**: 157.
Cross References
Agraphia; Aphasia; Dystypia

Dystonia

Dystonia, a term first used by Oppenheim in 1911, is a motor syndrome of sustained involuntary muscle contractions causing twisting and repetitive movements, sometimes tremor, and/or abnormal postures. Dystonic movements may initially appear with voluntary movement of the affected part ("action dystonia") but may eventually occur with voluntary movement elsewhere in the body ("overflow"). The severity of dystonia may be reduced by sensory tricks (*geste antagoniste*), using tactile or proprioceptive stimuli to lessen or eliminate posturing; this feature is unique to dystonia. Dystonia may develop after muscle fatiguing activity, and patients with focal dystonias show more rapid fatigue than normals. Dystonic disorders may be classified according to:

- *Age of onset*: the most significant predictor of prognosis: worse with earlier onset.
- *Distribution*: focal, segmental, multifocal, generalised; hemidystonia.
- *Aetiology*: primary/idiopathic *vs*. secondary/symptomatic.

Primary/idiopathic dystonias include:

- Primary torsion dystonia (idiopathic torsion dystonia).
- Severe generalized dystonia (dystonia musculorum deformans).
- Segmental, multifocal and focal dystonias (*e.g*. torticollis, blepharospasm, writer's cramp).
- Dopa-responsive dystonia (DRD; Segawa's syndrome).
- Myoclonic dystonia.

Secondary/symptomatic dystonia: the differential diagnosis is broad, with more than 40 known causes, including:

- Heredodegenerative disorders: Wilson's disease, Huntington's disease, neurodegeneration with brain iron accumulation, mitochondrial disorders, X-linked dystonia-parkinsonism (lubag).
- Paroxysmal dystonias/dyskinesias: paroxysmal kinesigenic choreoathetosis/dystonia (PKC; usually responds to carbamazepine), and paroxysmal non-kinesigenic dystonia/choreoathetosis (PDC; does not respond to carbamazepine).
- Metachromatic leukodystrophy.
- Gangliosidoses (GM1, GM2).
- Perinatal cerebral injury.
- Encephalitis.
- Head trauma.
- Multiple sclerosis.
- Drugs/toxins, *e.g*. antipsychotic, antiemetic, and antidepressant drugs.
- Psychogenic.

Appropriate investigations to exclude these symptomatic causes (especially Wilson's disease) are appropriate.

The pathogenesis of dystonia is incompletely understood. Different mechanisms may apply in different conditions. Peripheral focal dystonias such as torticollis and writer's cramp have been suggested to result from abnormal afferent information relayed from "stiff" muscle spindles. The genetic characterisation of various dystonic syndromes may facilitate understanding of pathogenesis.

From the therapeutic point of view, one of the key questions relates to response to levodopa: dopa-responsive dystonia (DRD) responds very well to levodopa (and response fluctuations do not develop over time; *cf*. Parkinson's disease). Other treatments which are sometimes helpful include anticholinergics, dopamine antagonists, dopamine agonists, and baclofen. Drug-induced dystonia following antipsychotic, antiemetic, or antidepressant drugs is often relieved within 20 min by intramuscular biperiden (5 mg) or procyclidine (5 mg). Botulinum toxin may be very helpful in some focal dystonias (*e.g*. blepharospasm). Surgery for dystonia using deep brain stimulation is still at the experimental stage.

References

Fahn S, Marsden CD, Calne DB. Classification and investigation of dystonia. In: Marsden CD, Fahn S, editors. Movement disorders 2. London: Butterworth; 1987. p. 332–58.

Moore P, Naumann M, editors. Handbook of botulinum toxin treatment. 2nd ed. Oxford: Blackwell Scientific; 2003.

Phukan J, Albanese A, Gasser T, Warner T. Primary dystonia and dystonia-plus syndromes: clinical characteristics, diagnosis, and pathogenesis. *Lancet Neurol*. 2011; **10**: 1074–85.

Van Harten PN, Hoek HW, Kahn RS. Acute dystonia induced by drug treatment. *BMJ*. 1999; **319**: 623–6.

Warner TT, Bressman SB, editors. Clinical diagnosis and management of dystonia. Abingdon: Informa Healthcare; 2007.

Cross References

Anismus; Blepharospasm; Dysphonia; Eyelid apraxia; Fatigue; Gaping; Hemidystonia; Sensory tricks; Torticollis; Writer's cramp

Dystypia

Dystypia describes a specific impairment in typewriting, particular using a personal computer, in the absence of aphasia, agraphia, apraxia or other neuropsychological deficit. Two types have been suggested according to whether typing errors are predominantly linguistic (frontal type) or spatial (parietal type)

References

Cook FA, Makin SD, Wardlaw J, Dennis MS. Dystypia in acute stroke not attributable to aphasia or neglect. *BMJ Case Rep*. 2013; **2013**. pii: bcr2013200257.

Otsuki M, Nakagawa Y, Imamura H, Ogata A. Dystypia: frontal type and parietal type. *J Neurol*. 2011; **258**(Suppl 1): S189 (abstract P696).

Cross References

Agraphia; Aphasia; Dystextia