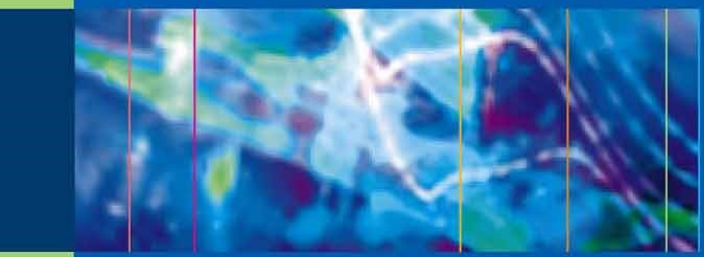
A.J. Larner



A Dictionary of Neurological Signs

Second Edition

Springer

A DICTIONARY OF NEUROLOGICAL SIGNS

SECOND EDITION

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To Philippa, Thomas, and Elizabeth

" ... there are many works useful and even necessary, which require no genius at all; and dictionary making is one of these."

James Burnet, Lord Monboddo.
Of the origin and progress of language: 1773-1792: V, 273

"I know ... that Writers of Travels, like Dictionary-Makers, are sunk into Oblivion by the Weight and Bulk of those who come after, and therefore lie uppermost."

Jonathan Swift
Gulliver's Travels: 1726

FOREWORD TO THE FIRST EDITION

Neurology has always been a discipline in which careful physical examination is paramount. The rich vocabulary of neurology replete with eponyms attests to this historically. The decline in the importance of the examination has long been predicted with the advent of more detailed neuroimaging. However, neuroimaging has often provided a surfeit of information from which salient features have to be identified, dependent upon the neurological examination. A dictionary of neurological signs has a secure future.

A dictionary should be informative but unless it is unwieldy, it cannot be comprehensive, nor is that claimed here. Andrew Larner has decided sensibly to include key features of the history as well as the examination. There is no doubt that some features of the history can strike one with the force of a physical sign. There are entries for "palinopsia" and "environmental tilt" both of which can only be elicited from the history and yet which have considerable significance. There is also an entry for the "head turning sign" observed during the history taking itself as well as the majority of entries relating to details of the physical examination.

This book is directed to students and will be valuable to medical students, trainee neurologists, and professions allied to medicine. Neurologists often speak in shorthand and so entries such as "absence" and "freezing" are sensible and helpful. For the more mature student, there are the less usual as well as common eponyms to entice one to read further than the entry which took you first to the dictionary.

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PREFACE TO THE SECOND EDITION

As in the first edition, the belief in signs as signifiers underpins this text. The aim is to be true to the "methode anatomo-clinique" pioneered in neurology by Charcot,[1] but also integrating, where possible, data from newer sources such as neuroimaging and neurogenetics.

Certain omissions in the first edition have become evident to me, necessitating a second edition (moreover, according to Michael Holroyd, "Collectors like first editions, authors like second editions"). Just under 700 entries have now expanded to a little under 1000. Most signs should be elicitable with the kit typically carried by neurologists.[2]

New features include a greater emphasis on change in signs with ageing and more medical history. Perspective on which signs are "really important" has been addressed elsewhere.[3] Details of neurological conditions associated with the various neurological signs are not discussed in any depth. Readers are encouraged to consult appropriate texts, for one of which the author has a particular, and hopefully excusable, bias.[4]

Neurological signs, like neurological diagnoses, are medical constructs and, hence, cultural artefacts liable to change with time. Hence, all definitions are seen as provisional rather than fixed. Systematic studies which "operationalize" signs, both how to elicit them and how to rate responses,[5] alone will define their utility in terms of sensitivity and specificity.

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PREFACE TO THE FIRST EDITION

In writing a book devoted to neurological signs and their meaning, it is not my intention to undervalue in any way the skill of neurological history taking. This remains the key element of the doctor-patient encounter both in the neurological clinic and on the ward, and is clearly crucial in order to formulate diagnostic hypotheses, guide clinical examination, and help decide on the nature of the pathological process (if one is present). However, having sat through several thousand neurological consultations, I do not subscribe to the view that all one need do is listen carefully and the patient will "tell you the diagnosis", although this may happen on rare (and often memorable) occasions. Clearly, history taking is not simply a passive recording of symptoms ("what the patient complains of"), but also an active process of seeking information of possible diagnostic significance through appropriate questions; this might be called the "historical examination". This latter facet of history taking, much the more difficult skill to learn, may disclose certain neurological signs which are not available to *physical* examination (principally in the sensory domain, but also intermittent motor phenomena). Hence, my use of the term "sign" in this book is a broad one, encompassing not only findings in physical examination (its traditional use) but also from focused history taking. My operational definition of sign is therefore simply a "signifier", in the sense of phenomena of semiologic value, giving information as to anatomical location and/or pathological cause.

Most neurological textbooks adopt an approach which is either symptom-based, beginning with what the patient complains of and then offering a structured differential diagnosis; or disease-based, assuming that a diagnosis has already been established. Although such texts are of great value, it seems to me that this does leave a place for a book devoted to neurological signs. Signs, elicited in either the historical or neurological examination, bridge the gap between the patient's symptoms, and the selection of appropriate investigations to confirm or refute the examiner's diagnostic formulations and thus establish a diagnosis.

Although it has been mooted whether the dramatic technological advances in neurological practice, for example in neuroimaging, might render neurological examination redundant, others maintain the central importance of neurological examination in patient management.[1,2]

It will come as little surprise to the reader that I am emphatically of the latter persuasion. However, this book does not aim to be a handbook of neurological examination technique (one reason for the absence of pictures), or neurological investigation, many excellent examples of which already exist. Rather, it seeks to elucidate the interpretation of neurological signs ("neurosemiology"): their anatomical, physiological, and pathological significance (where these are known). It should be added quickly that this is not to suggest that neurological signs are peculiarly objective (as some systems of clerking might suggest): as with all clinical observations, neurological signs are subject to both inter- and intra-observer variation and are biased by prior knowledge of the history and other examination findings.[3-5] As with other elements of clinical examination, relatively little study of the accuracy and precision of neurological signs has been undertaken; a methodology to remedy this situation has been suggested.[6] It is hoped that the current work might encourage more such studies. To those who might suggest that, in an age of molecular genetics, such an undertaking is passé, and rather nineteenth-century in its outlook, I would argue that precision in the definition of clinical signs is of relevance if meaningful genotype/phenotype correlations are to be established.

An attempt has been made to structure the entries in this volume in the following way:

- a definition of the sign, or the common usage of the term (subtypes italicized);
- a brief account of the clinical technique required to elicit the sign;
- a description of other neurological signs which may accompany the index sign (cross referenced as appropriate).

Where known, there is appended:

- a brief account of the neuroanatomical basis of the sign;
- an explanation, where possible, of the pathophysiological and/or pharmacological basis of the sign;
- the neuropathological basis of sign;
- a differential diagnosis of the commonest clinical diseases causing or associated with the sign (bulleted);
- brief details of specific treatments of these disorders, if available.

Using this schema, it will hopefully prove possible to integrate clinical phenomenology with the underlying neuroscience (anatomy, physiology, and pathology) in an accessible manner which will facilitate assimilation by the reader. Clearly not all these factors are known or applicable for every sign, and hence definitions vary quite considerably in length, the longer entries generally being for signs of greater clinical importance. Salient references from the primary and secondary literature are given, particularly for the more uncommon signs, for those wishing to pursue topics further. Entries are cross-referenced to other relevant signs.

Clearly such an undertaking cannot hope to be (and does not claim to be) comprehensive, such is the diversity of neurological function. Moreover, the limitations of my personal clinical experience means that selections are inevitably somewhat arbitrary, precluding (at the very least!) inclusion of signs familiar in pediatric neurological practice. Dermatological signs of potential neurological relevance have also been largely overlooked, and after much consideration "bruit" has been omitted. Nonetheless, it is hoped that this book will be of use to all students of neurology, both undergraduate and postgraduate, both dedicated neurology trainees and those required, perhaps against their personal inclinations, to develop some familiarity with neurology for examination purposes (*e.g.* candidates for the MRCP). It may also serve as a book of reference for more experienced clinicians. Since the majority of patients with neurological symptoms and signs in the United Kingdom are currently seen by general practitioners and general physicians, a situation which is likely to persist for some time, if not indefinitely,[7] it is very much hoped that these groups will also find the book of use, as indeed may members of ancillary professions: nursing, physiotherapy, speech and language therapy, occupational therapy, radiography.

The definitions given are not conceived of as in any way immutable. Language, after all, is plastic with respect to meaning and usage, and my aim is certainly not to "fix" the language. Nor do I suppose, despite my indebtedness to many distinguished colleagues, that I have been free from errors, all of which are my own doing. I shall be happy to hear from those who find errors, disagree with my suggested definitions, or feel that important signs have been omitted.

A.J. Larner

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CONTENTS

A

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.

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Cross References
Beevor'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.

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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 lobule and inferior temporal gyrus.

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

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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.

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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 DYSDIADOCHOKINESIA

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, palmomental, 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.

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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.

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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, agnosic defects merge into anterograde amnesia (failure to learn new information).

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

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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 paragraphia may be distinguished, *viz.*:
  surface/lexical/semantic dysgraphia: misspelling of irregular words, producing phonologically plausible errors (*e.g.*, simtums for symptoms); this is seen with left temporoparietal lesions (*e.g.*, Alzheimer's disease, Pick's disease);
  deep/phonological dysgraphia: inability to spell unfamiliar words and 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 (paragraphia); oral spelling may be spared.
- *Pure agraphia*:
  A rare syndrome in which oral language, reading and praxis are normal.

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

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

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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

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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 β-blockers, such as propranolol, may also help, as may anticholinergic agents, amantadine, clonazepam, and clonidine.

References

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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 (akinetic mutism) may occur if pathology is bilateral. Pure akinesia, without rigidity or tremor, may occur: if levodopa-responsive, this is usually due to Parkinson's disease; if levodopa-unresponsive, it may be the harbinger of progressive supranuclear palsy.

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/akinetic-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

Akinetic 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 (subacute 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

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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 hemispatial neglect.

The various forms of peripheral alexia may coexist; following a stroke, patients may present with global alexia which evolves to a pure alexia over the following weeks. Pure alexia is caused by damage to the left occipito-temporal junction, 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.

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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
Larner AJ. The neurology of "Alice." *Advances in Clinical Neuroscience & Rehabilitation* 2005; **4(6)**: 35-36
Todd J. The syndrome of Alice in Wonderland. *Canadian Medical Association Journal* 1955; **73**: 701-704
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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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

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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 nonverbal/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).

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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.

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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
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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
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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

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Cross References

Aphemia; Bulbar palsy; Dysarthria

Anesthesia
Anesthesia (anaesthesia) is a complete loss of sensation; hypoesthesia (hypoaesthesia, 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

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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

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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

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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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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

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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

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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.

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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).

Summary of findings in aphasia syndromes

	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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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.

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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.

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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
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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 α-motor neurones, the efferent limb of the reflex, so completing the monosynaptic arc. Hence, although reflexes are typically regarded as part of the examination of the motor system, reflex loss may also occur in "sensory" disorders, affecting the Ia afferents from the muscle spindle. It is often possible to "hear" that reflexes are absent from the thud of tendon hammer on tendon.

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

Argyll Robertson Pupil (ARP)
The Argyll Robertson pupil is small (miosis) and irregular. It fails to react to light (reflex iridoplegia), but does constrict to accommodation

(when the eyes converge). In other words, there is light-near pupillary dissociation (ARP = **a**ccommodation **r**eaction **p**reserved). Since the light reflex is lost, testing for the accommodation reaction may be performed with the pupil directly illuminated: this can make it easier to see the response to accommodation, which is often difficult to observe when the pupil is small or in individuals with a dark iris. There may be an incomplete response to mydriatic drugs. Although pupil involvement is usually bilateral, it is often asymmetric, causing anisocoria.

The Argyll Robertson pupil was originally described in the context of neurosyphilis, 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

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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

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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 stereoanesthesia 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 neuronopathies affecting the dorsal root ganglia. It is markedly exacerbated by removal of visual cues (*e.g.*, as in Romberg's sign), unlike the situation with cerebellar ataxia, and may also lead to pseudoathetosis.
- *Optic ataxia*:
  Misreaching for visually presented targets, with dysmetria, due to a parieto-occipital lesion, as seen in Balint's syndrome.
- "*Frontal ataxia*":
  Similar to, and sometimes indistinguishable from, cerebellar ataxia, but results from lesions of the contralateral frontal cortex or frontopontine 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
  Dentatorubropallidoluysian 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

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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 (crural 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

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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 been seen in the presence of sensory ataxia (impaired proprioception) and are known as pseudoathetosis or pseudochoreoathetosis.

Choreoathetoid movements result from disorders of the basal ganglia.

References

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

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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; uncinate 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.

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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

B

Babinski's Sign (1)

Babinski's sign is a polysynaptic cutaneous reflex consisting of an extensor movement (dorsiflexion) of the big toe on eliciting the plantar response, due to contraction of extensor hallucis longus. There may be in addition fanning (abduction) of the other toes (fan sign; *signe de l'éventail*) but this is neither necessary nor sufficient for Babinski's sign to be present. There may be simultaneous contraction of other limb flexor muscles, consistent with the notion that Babinski's sign forms part of a flexion synergy (withdrawal) of the leg. The use of the term "negative Babinski sign" to indicate the normal finding of a downgoing (flexor; plantar flexion) big toe is incorrect, "flexor plantar response" being the appropriate description.

The plantar response is most commonly performed by stroking the sole of the foot, although many other variants are described (*e.g.*, Chaddock's sign, Gordon's sign, Oppenheim's sign, *q.v.*).

Babinski's sign is normal in infants with immature (unmyelinated) corticospinal tracts; persistence beyond three years of age, or reemergence in adult life, is pathological. In this context, Babinski's sign is considered a reliable ("hard") sign of corticospinal (pyramidal) tract dysfunction (upper motor neurone pathology), and may coexist with other signs of upper motor neurone dysfunction (*e.g.*, weakness in a so-called pyramidal distribution, spasticity, hyperreflexia). However, if weakness of extensor hallucis longus is one of the features of upper motor neurone dysfunction, or from any other cause, Babinski's sign may be unexpectedly absent although anticipated on clinical grounds. In the presence of extrapyramidal signs, it is important to distinguish Babinski's sign, a "pyramidal sign," from a striatal toe (spontaneous upgoing plantar).

References

Lance JW. The Babinski sign. *Journal of Neurology, Neurosurgery and Psychiatry* 2002; **73**: 360-362

Van Gijn J. *The Babinski sign: a centenary*. Utrecht: Universiteit Utrecht, 1996

Cross References

Chaddock's sign; Gordon's sign; Hyperreflexia; Oppenheim's sign; Parkinsonism; Plantar response; Spasticity; Striatal toe; Upper motor neurone (UMN) syndrome; Weakness

Babinski's Sign (2)

Babinski (1905) described the paradoxical elevation of the eyebrow in hemifacial spasm as orbicularis oris contracts and the eye closes, a synkinesis which is not reproducible by will. This observation indicated to Babinski the peripheral (facial nerve) origin of hemifacial spasm. It may assist in differentiating hemifacial spasm from other craniofacial movement disorders.

References

Devoize JL. "The other" Babinski sign: paradoxical raising of the eyebrow in hemifacial spasm. *Journal of Neurology, Neurosurgery and Psychiatry* 2001; **70**: 516

Cross References

Hemifacial spasm

Babinski's Trunk-Thigh Test

Babinski's trunk-thigh test is suggested to be of use in distinguishing organic from functional paraplegia and hemiplegia (Hoover's sign may also be of use in the latter case). The recumbent patient is asked to sit up with the arms folded on the front of the chest. In organic hemiplegia there is involuntary flexion of the paretic leg; in paraplegia both legs are involuntarily raised. In functional paraplegic weakness neither leg is raised, and in functional hemiplegia only the normal leg is raised.

Cross References

Functional weakness and sensory disturbance; Hemiplegia; Hoover's sign; Paraplegia

"Bag of Worms"

- see MYOKYMIA

Balaclava Helmet

A pattern of facial sensory loss resembling in distribution a balaclava helmet, involving the outer parts of the face but sparing the nose and mouth, may be seen with central lesions, such as syringobulbia which progress upwards from the neck, such that the lowermost part of the spinal nucleus of the trigeminal nerve which serves the outer part of the face is involved while the upper part of the nucleus which serves the central part of the face is spared. This pattern of facial sensory impairment may also be known as onion peel or onion skin.

Cross References

Onion peel, Onion skin

Balint's Syndrome

Balint's syndrome, first described by a Hungarian neurologist in 1909, consists of:

- *Simultanagnosia* (*q.v.*; dorsal type):
  A constriction of visual attention, such that the patient is aware of only one object at a time; visual acuity is preserved, and patients can recognize single objects placed directly in front of them; they are unable to read or distinguish overlapping figures.
- *Spatial disorientation*:
  Loss of spatial reference and memory, leaving the patient "lost in space."
- *Disorders of oculomotor function*:
  Specifically visually guided eye movements (fixation, pursuit, saccades); Balint's "psychic paralysis of gaze," or "sticky

fixation," refers to an inability to direct voluntary eye movements to visual targets, despite a full range of eye movements; this has also been characterized as a form of oculomotor apraxia. Accurate eye movements may be programmed by sound or touch. Loss of spontaneous blinking has also been reported.

- *Optic ataxia*:

A failure to grasp or touch an object under visual guidance. Not all elements may be present; there may also be coexisting visual field defects, hemispatial neglect, visual agnosia, or prosopagnosia. Balint's syndrome results from bilateral lesions of the parieto-occipital junction causing a functional disconnection between higher order visual cortical regions and the frontal eye fields, with sparing of the primary visual cortex. Brain imaging, either structural (CT, MRI) or functional (SPECT, PET), may demonstrate this bilateral damage, which is usually of vascular origin, for example due to watershed or border zone ischemia, or top-of-the-basilar syndrome. Balint syndrome has also been reported as a migrainous phenomenon, following traumatic brain injury and in association with Alzheimer's disease, tumor (butterfly glioma), radiation necrosis, progressive multifocal leukoencephalopathy, Marchiafava-Bignami disease with pathology affecting the corpus callosum, and X-linked adrenoleukodystrophy.

References

Husein M, Stein J. Rezso Balint and his most celebrated case. *Archives of Neurology* 1988; **45**: 89-93

Rafal R. Bálint's syndrome: a disorder of visual cognition. In: D'Esposito M (ed.). *Neurological foundations of cognitive neuroscience*. Cambridge: MIT Press, 2003: 27-40

Cross References

Apraxia; Blinking; Ocular apraxia; Optic ataxia; Simultanagnosia

Ballism, Ballismus

Ballism or ballismus is a hyperkinetic involuntary movement disorder characterized by wild, flinging, throwing movements of a limb. These movements most usually involve one half of the body (*hemiballismus*), although they may sometimes involve a single extremity (*monoballismus*) or both halves of the body (*paraballismus*). The movements are often continuous during wakefulness but cease during sleep. Hemiballismus may be associated with limb hypotonia. Clinical and pathophysiological studies suggest that ballism is a severe form of chorea. It is most commonly associated with lesions of the contralateral subthalamic nucleus.

Cross References

Chorea, Choreoathetosis; Hemiballismus; Hypotonia, Hypotonus

Bathing Suit Sensory Loss

- see SUSPENDED SENSORY LOSS

Battle's Sign

Battle's sign is a hematoma overlying the mastoid process, which indicates an underlying basilar skull fracture extending into the mastoid portion of the temporal bone. It appears 48-72 hours after the trauma that causes the fracture.

Beevor's Sign

Beevor's sign is an upward movement of the umbilicus in a supine patient attempting either to flex the head onto the chest against resistance (*e.g.*, the examiner's hand) or performing a sit-up. It indicates a lesion causing rectus abdominis muscle weakness below the umbilicus. This may occur with a spinal lesion (*e.g.*, tumor, syringomyelia) between T10 and T12 causing isolated weakness of the lower part of the muscle, or myopathies affecting abdominal muscles, particularly facioscapulohumeral muscular dystrophy. Lower cutaneous abdominal reflexes are also absent, having the same localizing value.

Downward movement of the umbilicus ("inverted Beevor's sign") due to weakness of the upper part of rectus abdominis is less often seen.

References

Hilton-Jones D. Beevor's sign. *Practical Neurology* 2004; **4**: 176-177

Tashiro K. Charles Edward Beevor (1854-1908) and Beevor's sign. In: Rose FC (ed.). *A short history of neurology: the British contribution 1660-1910*. Oxford: Butterworth Heinemann, 1999: 222-225

Cross References

Abdominal reflexes

Belle Indifférence

La belle indifférence refers to a patient's seeming lack of concern in the presence of serious symptoms. This was first defined in the context of "hysteria," along with exaggerated emotional reactions, what might now be termed functional or somatoform illness. However, the sign is a poor discrimator against "organic" illness. Some patients' coping style is to make light of serious symptoms; they might be labeled stoical.

Patients with neuropathological lesions may also demonstrate a lack of concern for their disabilities, either due to a disorder of body schema (anosodiaphoria) or due to incongruence of mood (typically in frontal lobe syndromes, sometimes seen in multiple sclerosis).

References

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

Cross References

Anosodiaphoria; Frontal lobe syndromes; Functional weakness and sensory disturbance

Bell's Palsy

Bell's palsy is an idiopathic peripheral (lower motor neurone) facial weakness (prosopoplegia). It is thought to result from viral inflammation

of the facial (VII) nerve. Other causes of lower motor neurone facial paresis (*q.v.*) may need to be excluded before a diagnosis of Bell's palsy can be made.

In the majority of patients with Bell's palsy (idiopathic facial paresis), spontaneous recovery occurs over three weeks to two months. Poorer prognosis is associated with older age (over 40 years) and if no recovery is seen within four weeks of onset. The efficacy of steroid treatment remains uncertain, but it is often prescribed; it may improve facial functional outcome.

References

Grogan PM, Gronseth GS. Practice parameter: steroids, acyclovir, and surgery for Bell's palsy (an evidence-based review). Report of the Quality Standards Subcommittee of the American Academy of Neurology. *Neurology* 2001; **56**: 830-836

Williamson IG, Whelan TR. The clinical problem of Bell's palsy: is treatment with steroids effective? *British Journal of General Practice* 1996; **46**: 743-747

Cross References

Bell's phenomenon, Bell's sign; Facial paresis; Lower motor neurone (LMN) syndrome

Bell's Phenomenon, Bell's Sign

Bell's phenomenon or sign is reflex upward, and slightly outward, deviation of the eyes in response to forced closure, or attempted closure, of the eyelids. This is a synkinesis of central origin involving superior rectus and inferior oblique muscles. It may be very evident in a patient with Bell's palsy (idiopathic facial nerve paralysis) attempting to close the paretic eyelid. The reflex indicates intact nuclear and infranuclear mechanisms of upward gaze, and hence that any defect of upgaze is supranuclear. However, in making this interpretation it should be remembered that perhaps 10-15% of the normal population do not show a Bell's phenomenon.

Bell's phenomenon is usually absent in progressive supranuclear palsy and is only sometimes spared in Parinaud's syndrome

References

Bell C. On the motions of the eye, in illustration of the use of the muscles and nerves of the orbit. *Philosophical Transactions of the Royal Society, London* 1823; **113**: 166-186.

Cross References

Bell's palsy; Gaze palsy; Parinaud's syndrome; Supranuclear gaze palsy; Synkinesia, synkinesis

Benediction Hand

Median nerve lesions in the axilla or upper arm cause weakness in all median nerve innervated muscles, including flexor digitorum profundus. On attempting to make a fist, impaired flexion of the index and middle fingers, complete and partial respectively, results in a hand posture likened to that of a priest saying benediction.

A somewhat similar, but not identical, appearance may occur with ulnar nerve lesions: hyperextension of the metacarpophalangeal joints

of the ring and little fingers with slight flexion at the interphalangeal joints. The index and middle fingers are less affected because of the intact innervation of their lumbrical muscles (median nerve).

Cross References

Claw hand; Simian hand

Bent Spine Syndrome

- see CAMPTOCORMIA

Bielschowsky's Sign, Bielschowsky's Test

Bielschowsky's sign is head tilt toward the shoulder, typically toward the side contralateral to a trochlear (IV) nerve palsy. The intorsion of the unaffected eye brought about by the head tilt compensates for the double vision caused by the unopposed extorsion of the affected eye. Very occasionally, head tilt is paradoxical (*i.e.*, toward the involved side: presumably the greater separation of images thus produced allows one of them to be ignored).

Bielschowsky's (head tilt) test consists of the examiner tipping the patient's head from shoulder to shoulder to see if this improves or exacerbates double vision, as will be the case when the head is respectively tilted away from or toward the affected side in a unilateral trochlear (IV) nerve lesion. The test is usually negative in a skew deviation causing vertical divergence of the eyes. This test may also be used as part of the assessment of vertical diplopia to see whether hypertropia changes with head tilt to left or right; increased hypertropia on left head tilt suggests a weak intortor of the left eye (superior rectus); increased hypertropia on right head tilt suggests a weak intortor of the right eye (superior oblique).

Cross References

Diplopia; Hypertropia; Skew deviation

Bitemporal Hemianopia

- see HEMIANOPIA; VISUAL FIELD DEFECTS

Blepharoptosis

- see PTOSIS

Blepharospasm

Blepharospasm is a focal dystonia of the orbicularis oculi resulting in repeated involuntary forced eyelid closure, with failure of voluntary opening. It may be sufficiently severe to result in functional blindness. The condition typically begins in the sixth decade of life, and is commoner in women than men. Blepharospasm may occur in isolation or in combination with other involuntary movements which may be dystonic (orobuccolingual dystonia or Meige syndrome; limb dystonia) or dyspraxic (eyelid apraxia).

Blepharospasm is usually idiopathic but may be associated with lesions (usually infarction) of the rostral brainstem, diencephalon, and striatum; it has been occasionally reported with thalamic lesions. The

pathophysiological mechanisms underlying blepharospasm are not understood, but may reflect dopaminergic pathway disruption causing disinhibition of brainstem reflexes.

Local injections of botulinum toxin into orbicularis oculi are the treatment of choice, the majority of patients deriving benefit and requesting further injection. Failure to respond to botulinum toxin may be due to concurrent eyelid apraxia or dopaminergic therapy with levodopa.

References

Grandas F, Elston J, Quinn N, Marsden CD. Blepharospasm: a review of 264 patients. *Journal of Neurology, Neurosurgery and Psychiatry* 1988; **51**: 767-772

Hallett M, Daroff RB. Blepharospasm: report of a workshop. *Neurology* 1996; **46**: 1213-1218

Cross References

Blinking; Dystonia; Eyelid apraxia; Gaping; Yawning

Blind Spot

The blind spot is defined anatomically as the point on the retina at which axons from the retinal ganglion cells enter the optic nerve; since this area is devoid of photoreceptors there is a physiological blind spot. This area may be mapped clinically by confrontation with the examiner's blind spot, or mechanically. Enlargement of the blind spot (peripapillary scotoma) is observed with raised intracranial pressure causing papilledema: this may be helpful in differentiating papilledema from other causes of disc swelling, such as optic neuritis, in which a central scotoma is the most common field defect. Enlargement of the blind spot may also be a feature of peripapillary retinal disorders including big blind spot syndrome.

Cross References

Disc swelling; Papilledema; Scotoma

Blinking

Involuntary blinking rate is decreased in idiopathic Parkinson's disease (and may be improved by dopaminergic therapy) and progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome). In contrast, blink rate is normal in multiple system atrophy and dopa-responsive dystonia, and increased in schizophrenia and postencephalitic parkinsonism. These disparate observations are not easily reconciled with the suggestion that blinking might be a marker of central dopaminergic activity.

Loss of spontaneous blinking has been reported in Balint's syndrome. In patients with impaired consciousness, the presence of involuntary blinking implies an intact pontine reticular formation; absence suggests structural or metabolic dysfunction of the reticular formation. Blinking decreases in coma. Functional disorders may be accompanied by an increase in blinking.

Cross References

Balint's syndrome; Blink reflex; Coma; Corneal reflex; Parkinsonism; Sighing; Yawning

Blink Reflex

The blink reflex consists of bilateral reflex contraction of the orbicularis oculi muscles. This may be induced by:

- Mechanical stimulus:
  Examples include percussion over the supraorbital ridge (glabellar tap reflex, Myerson's sign, nasopalpebral reflex): this quickly habituates with repetitive stimulation in normal individuals; touching the cornea (corneal reflex); stroking the eyelashes in unconscious patients with closed eyes ("eyelash reflex").
- Visual stimulus:
  Sudden visual stimulus approaching the eyes (menace reflex, threat reflex, visuopalpebral reflex): the stimulus should be unexpected since the reflex can be voluntarily suppressed; failure to respond to a stimulus moving into the temporal field of vision may indicate a hemianopic field defect in patients unable to comply with standard confrontation visual field testing. Care should be taken to avoid generating air currents with the hand movement as this may stimulate the corneal reflex which may simulate the visuopalpebral reflex. It is probable that this reflex requires cortical processing: it is lost in persistent vegetative states. Loss of this reflex may occur in Balint's syndrome, ascribed to inability to recognize the nearness of the threatening object.
- Acoustic stimulus:
  Sudden loud sounds (acousticopalpebral reflex).

The final common (efferent) pathway for these responses is the facial nerve nucleus and facial (VII) nerve, the afferent limbs being the trigeminal (V), optic (II), and auditory (VIII) nerves respectively.

Electrophysiological study of the blink reflex may demonstrate peripheral or central lesions of the trigeminal (V) nerve or facial (VII) nerve (afferent and efferent pathways, respectively). It has been reported that in the evaluation of sensory neuronopathy the finding of an abnormal blink reflex favors a nonparaneoplastic etiology, since the blink reflex is normal in paraneoplastic sensory neuronopathies.

References

Auger RG, Windebank AJ, Lucchinetti CF, Chalk CH. Role of the blink reflex in the evaluation of sensory neuronopathy. *Neurology* 1999; **53**: 407-408

Liu GT, Ronthal M. Reflex blink to visual threat. *Journal of Clinical Neuro-ophthalmology* 1992; **12**: 47-56

Cross References

Balint's syndrome; Blinking; Corneal reflex; Glabellar tap reflex

Body Part as Object

In this phenomenon, apraxic patients use a body part when asked to pantomime certain actions, such as using the palm when asked to demonstrate the use of a hair brush or comb, or fingers when asked to demonstrate use of scissors or a toothbrush.

References

Goodglass H, Kaplan E. Disturbance of gesture and pantomime in aphasia. *Brain* 1963; **86**: 703-720

Kato M, Meguro K, Sato M *et al*. Ideomotor apraxia in patients with Alzheimer's disease: why do they use their body parts as objects? *Neuropsychiatry Neuropsychology and Behavioral Neurology* 2001; **14**: 45-52

Cross References

Apraxia

"Bon-Bon Sign"

Involuntary pushing of the tongue against the inside of the cheek, the "bon-bon sign," is said to be typical of the stereotypic orolingual movements of tardive dyskinesia, along with chewing and smacking of the mouth and lips, and rolling of the tongue in the mouth. These signs may help to distinguish tardive dyskinesia from chorea, although periodic protrusion of the tongue (flycatcher, trombone tongue) is common to both.

Cross References

Chorea, Choreoathetosis; Trombone tongue

Bouche de Tapir

Patients with facioscapulohumeral (FSH) dystrophy have a peculiar and characteristic facies, with puckering of the lips when attempting to whistle. The pouting quality of the mouth, unlike that seen with other types of bilateral (neurogenic) facial weakness, has been likened to the face of the tapir (*Tapirus* sp.).

Cross References

Facial paresis

Bovine Cough

A bovine cough lacks the explosive character of a normal voluntary cough. It may result from injury to the distal part of the vagus nerve, particularly the recurrent laryngeal branches which innervate all the muscles of the larynx (with the exception of cricothyroid) with resultant vocal cord paresis. Because of its longer intrathoracic course, the left recurrent laryngeal nerve is more often involved. A bovine cough may be heard in patients with tumors of the upper lobes of the lung (Pancoast tumor) due to recurrent laryngeal nerve palsy. Bovine cough may also result from any cause of bulbar weakness, such as motor neurone disease, Guillain-Barré syndrome, and bulbar myopathies.

References

Arcasoy SM, Jett JR. Superior pulmonary sulcus tumors and Pancoast's syndrome. *New England Journal of Medicine* 1997; **337**: 1370-1376

Cross References

Bulbar palsy; Diplophonia; *Signe de rideau*

Bradykinesia

Bradykinesia is a slowness in the initiation and performance of voluntary movements, one of the typical signs of parkinsonian

syndromes, in which situation it is often accompanied by difficulty in the initiation of movement (akinesia, hypokinesia) and reduced amplitude of movement (hypometria) which may increase with rapid repetitive movements (fatigue). It may be overcome by reflexive movements or in moments of intense emotion (*kinesis paradoxica*). Bradykinesia in parkinsonian syndromes reflects dopamine depletion in the basal ganglia. It may be improved by levodopa and dopaminergic agonists, less so by anticholinergic agents.

Slowness of voluntary movement may also be seen with psychomotor retardation, frontal lobe lesions producing abulia, and in the condition of obsessive slowness.

Cross References

Abulia; Akinesia; Fatigue; Hypokinesia; Hypometria; *Kinesis paradoxica*; Parkinsonism; Psychomotor retardation

Bradylalia

Bradylalia is slowness of speech, typically seen in the frontal-subcortical types of cognitive impairment, with or without extrapyramidal features, or in depression.

Cross References

Palilalia; Tachylalia

Bradyphrenia

Bradyphrenia is a slowness of thought, typically seen in the frontal-subcortical types of cognitive impairment, *e.g.*, progressive supranuclear palsy, vascular dementia, Huntington's disease. Such patients typically answer questions correctly but with long response times.

Cross References

Abulia; Dementia

Bragard's Test

- see LASÈGUE'S SIGN

Broca's Aphasia

Broca's aphasia is the classic "expressive aphasia," in distinction to the "receptive aphasia" of Wernicke; however, there are problems with this simple classification, since Broca's aphasics may show comprehension problems with complex material, particularly in relation to syntax.

Considering each of the features suggested for the clinical classification of aphasias (see Aphasia), Broca's aphasia is characterized by:

- *Fluency*: slow, labored, effortful speech (nonfluent) with phonemic paraphasias, agrammatism, and aprosody; the patient knows what s/he wants to say and usually recognizes the paraphasic errors (*i.e.*, patients can "self-monitor").
- *Comprehension*: comprehension for simple material is preserved, but there may be problems with more complex syntax.
- *Repetition*: impaired.

- *Naming*: impaired (anomia, dysnomia); may be aided by phonemic or contextual cueing (*cf*. Wernicke's aphasia).
- *Reading*: alexia with labored oral reading, especially of function words and verb inflections. Silent reading may also be impaired (deep dyslexia) as reflected by poor text comprehension.
- *Writing*: similarly affected.

Aphemia was the name originally given by Broca to the language disorder subsequently named "Broca's aphasia." The syndrome may emerge during recovery from a global aphasia. Broca's aphasia is sometimes associated with a right hemiparesis, especially affecting the arm and face; there may also be bucco-lingual-facial dyspraxia. Depression may be a concurrent feature.

Classically Broca's aphasia is associated with a vascular lesion of the third frontal gyrus in the inferior frontal lobe (Broca's area), but in practice such a circumscribed lesion is seldom seen. More commonly there is infarction in the perisylvian region affecting the insula and operculum (Brodmann areas 44 and 45), which may include underlying white matter and the basal ganglia (territory of the superior branch of the middle cerebral artery).

The terms "small Broca's aphasia," "mini-Broca's aphasia," and "Broca's area aphasia," have been reserved for a more circumscribed clinical and neuroanatomical deficit than Broca's aphasia, wherein the damage is restricted to Broca's area or its subjacent white matter. There is a mild and transient aphasia or anomia which may share some of the characteristics of aphemia/phonetic disintegration (*i.e*., a motor disorder of speech production with preserved comprehension of spoken and written language).

References

Alexander MP, Naeser MA, Palumbo C. Broca's area aphasias: aphasia after lesions including the frontal operculum. *Neurology* 1990; **40**: 353-362

Mohr JP, Pessin MS, Finkelstein S, Funkenstein HH, Duncan GW, Davis KR. Broca aphasia: pathologic and clinical aspects. *Neurology* 1978; **28**: 311-324

Pearce JMS. Paul Broca and aphasia. In: Pearce JMS. *Fragments of neurological history*. London: Imperial College Press, 2003: 84-89

Cross References

Agrammatism; Agraphia; Alexia; Aphasia; Aphemia; Aprosodia, Aprosody; Paraphasia; Wernicke's aphasia

Brown-Séquard Syndrome

The Brown-Séquard syndrome is the consequence of anatomical or, more usually, functional hemisection of the spinal cord (spinal hemisection syndrome), producing the following pattern of clinical findings:

- *Motor*:

  Ipsilateral spastic weakness, due to corticospinal tract involvement

Segmental lower motor neurone signs at the level of the lesion, due to root and/or anterior horn cell involvement.

- *Sensory*:

  A dissociated sensory loss, i.e.:

  Ipsilateral loss of proprioception, due to dorsal column involvement;

  Contralateral loss of pain and temperature sensation, due to crossed spinothalamic tract involvement.

Spinal cord lesions producing this syndrome may be either extramedullary (*e.g.*, prolapsed cervical intervertebral disc, extrinsic spinal cord tumor) or intramedullary (*e.g.*, multiple sclerosis, intrinsic spinal cord tumor); the former group is said to be the more common cause.

References

Aminoff MJ. *Brown-Séquard. A visionary of science*. New York: Raven, 1993: 112-131

Engelhardt P, Trostdorf E. Zur Differentialdiagnose des Brown-Séquard-Syndroms. *Nervenarzt* 1997; **48**: 45-49

Tattersall R, Turner B. Brown-Séquard and his syndrome. *Lancet* 2000; **356**: 61-63

Cross References

Dissociated sensory loss; Myelopathy; Proprioception; Spasticity; Weakness

Brudzinski's (Neck) Sign

Brudzinski described a number of signs, but the one most often used in clinical practice is the neck sign, which is sometimes evident in cases of meningeal irritation, for example due to meningitis. Passive flexion of the neck to bring the head onto the chest is accompanied by flexion of the thighs and legs. As with nuchal rigidity and Kernig's sign, Brudzinski's sign may be absent in elderly or immunosuppressed patients with meningeal irritation.

References

Pearce JMS. Kernig and Brudzinski. In: Pearce JMS. *Fragments of neurological history*. London: Imperial College Press, 2003: 365-366

Cross References

Kernig's sign; Meningism; Nuchal rigidity

Bruit

Bruits arise from turbulent blood flow causing arterial wall vibrations that are audible at the body surface with the unassisted ear or with a stethoscope (diaphragm rather than bell, better for detecting higher frequency sounds). They are associated with stenotic vessels or fistulae where there is arteriovenous shunting of blood. Dependent on the clinical indication, various sites may be auscultated: eye for orbital bruit in carotico-cavernous fistula; head for bruit of AV fistula; but probably the most frequently auscultated region is the carotid bifurcation, high up under the angle of the jaw, in individuals thought to have had

a transient ischemic attack or ischemic stroke. Examination for carotid bruits in asymptomatic individuals is probably best avoided, other than in the clinical trial setting, since the optimal management of asymptomatic carotid artery stenosis has yet to be defined.

References

Sandercock PAG, Kavvadia E. The carotid bruit. *Practical Neurology* 2002; **2**: 221-224

Bruxism

Bruxism is forcible grinding or gnashing of the teeth. This is common in children, and as a parasomnia, said to occur in 5-20% of the population during nonREM sleep. Masseter hypertrophy may become apparent in persistent grinders. Bruxism may also occur in encephalopathic disorders (*e.g.*, hepatic encephalopathy) and occasionally in disorders of the basal ganglia (multiple system atrophy, basal ganglia infarcts). Dysfunction of efferent and/or afferent thalamic and striatopallidal tracts has been suggested as the neural substrate.

If necessary, a rubber device or bite may be worn in the mouth to protect the teeth. Botulinum toxin injections have also been tried.

References

Glaros AG, Rao SM. Bruxism: a critical review. *Psychological Bulletin* 1977; **84**: 767-781

Cross References

Encephalopathy; Masseter hypertrophy

Buccofacial Dyspraxia

- see OROFACIAL DYSPRAXIA

Bulbar Palsy

Bulbar palsy is weakness of bulbar musculature of lower motor neurone origin. This may be differentiated clinically from bulbar weakness of upper motor neurone origin (pseudobulbar palsy).

Clinical features of bulbar palsy include:
Dysarthria of flaccid/nasal type
Dysphonia
Dysphagia, often with nasal regurgitation
Weak ("bovine") cough; risk of aspiration
+/– Wasted, fasciculating tongue
+/– absent jaw jerk
+/– absent gag reflex.

Bulbar palsy is usually neurogenic. Recognized causes include:

- Brainstem disorders affecting cranial nerve motor nuclei (intrinsic):
  - motor neurone disease (which may also cause a pseudobulbar palsy)
  - poliomyelitis
  - glioma
  - syringobulbia

- Cranial nerve lesions outside the brainstem (there may be associated sensory signs):
  Infiltration by carcinoma, granuloma
- Neuromuscular junction transmission defect:
  myasthenia gravis.

A myogenic bulbar palsy may be seen in oculopharyngeal muscular dystrophy, inclusion body myositis, or polymyositis.

Cross References

Bovine cough; Dysarthria; Dysphagia; Dysphonia; Fasciculation; Gag reflex; Jaw jerk; Lower motor neurone (LMN) syndrome; Pseudobulbar palsy; Upper motor neurone (UMN) syndrome

Bulbocavernosus Reflex

A test of the integrity of the S2, S3 and S4 spinal roots, looking for contraction of the anal sphincter (may be felt with a gloved finger in the rectum) when squeezing the glans penis or clitoris. The reflex may be abolished in lesions of the cauda equina.

Cross References

Cauda equina syndrome; Reflexes

Buphthalmos

Buphthalmos, or ox-eye, consists of a large and bulging eye caused by raised intraocular pressure due to congenital or secondary glaucoma. This is one of the ophthalmological features of Sturge-Weber syndrome.

"Butt-First Maneuver"

- see GOWERS' SIGN

C

Cacogeusia
Sensation of a disagreeable taste, often associated with parosmia.

Cacosmia
- see PAROSMIA

Calf Hypertrophy
Calf enlargement has many causes; it may reflect true hypertrophy (enlargement of muscle fibers) or, more commonly, pseudohypertrophy, due to infiltration with tissue elements other than muscle.

Hypertrophy may be due to neuromuscular disorders producing:

- Chronic partial denervation, for example:
  radiculopathy
  peripheral neuropathy
  spinal muscular atrophy
  following paralytic poliomyelitis.
- Continuous muscle activity, for example:
  myotonia congenita
  Isaacs syndrome (neuromyotonia)
  generalized myokymia.

Pseudohypertrophy may be due to:

- Dystrophinopathies (Duchenne muscular dystrophy, Becker dystrophy), due to excess connective tissue.
- Infection/inflammation: myositis
- Infiltration: amyloidosis, tumor, cysticercosis

References
Coles A, Dick D. Unilateral calf hypertrophy. *Journal of Neurology, Neurosurgery and Psychiatry* 2004; **75**: 1606
Wilson H, Kidd D, Howard RS, Williams AJ, Spencer GT. Calf hypertrophy following paralytic poliomyelitis. *Postgraduate Medical Journal* 2000; **76**: 179-181
Cross References
Gowers' sign; Muscle hypertrophy; Myokymia; Myotonia; Neuromyotonia

Caloric Testing
Caloric tests examine the vestibulo-ocular reflexes (VOR). They are mainly used in two circumstances: to identify vestibular pathology in the assessment of dizziness/vertigo when clinical tests of VOR are unhelpful and to assess brainstem integrity in coma. Each labyrinth may be separately assessed by irrigating each outer ear. Head flexion to 30° above the

horizontal allows maximum stimulation of the horizontal semicircular canals, whereas 60° below horizontal maximally stimulates the lateral semicircular canals. Water 7°C above and below body temperature (*i.e.,* 30°C and 44°C) is used, applied for 30-40 seconds. Induced nystagmus is then timed both with and without visual fixation (in the dark, Frenzel glasses). This method is cheap but has poor patient acceptability.

Normally, the eyes show conjugate deviation toward the ear irrigated with cold water, with corrective nystagmus in the opposite direction; with warm water the opposite pattern is seen. (The direction of nystagmus may thus be recalled by the mnemonic COWS: **c**old **o**pposite, **w**arm **s**ame.) Dysconjugate responses suggest brainstem damage or depression. A reduced duration of induced nystagmus is seen with canal paresis; enhancement of the nystagmus with removal of visual fixation suggests this is peripheral in origin (labyrinthine, vestibulocochlear nerve), whereas no enhancement suggests a central lesion.

In coma the deviation may be present but without corrective saccades, even at a time when the oculocephalic responses elicited by the doll's head maneuver are lost. As coma deepens even the caloric reflexes are lost as brainstem involvement progresses.

References

Rudge P, Bronstein AM. Investigations of disorders of balance. In: Hughes RAC (ed.). *Neurological Investigations*. London: BMJ Publishing, 1997: 283-314

Cross References

Coma; Nystagmus; Oculocephalic response; Vertigo; Vestibulo-ocular reflexes

Camptocormia

Camptocormia, or "bent spine syndrome," was first described as a psychiatric phenomenon in men facing armed conflict (a "war neurosis"). It has subsequently been realized that reducible lumbar kyphosis may also result from neurological disorders, including muscle disease (paravertebral myopathy, nemaline myopathy), Parkinson's disease, dystonia, motor neurone disease, and, possibly, as a paraneoplastic phenomenon. Cases with associated lenticular (putaminal) lesions have also been described. Camptocormia may be related in some instances to dropped head syndrome.

References

Djaldetti R, Mosberg-Galili R, Sroka H, Merims D, Melamed E. Camptocormia (bent spine) in patients with Parkinson's disease: characterization and possible pathogenesis of an unusual phenomenon. *Movement Disorders* 1999; **14**: 443-447

Oerlemans WGH, de Visser M. Dropped head syndrome and bent spine syndrome: two separate entities or different manifestations of axial myopathy? *Journal of Neurology, Neurosurgery and Psychiatry* 1998; **65**: 258-259

Cross References

Dropped head syndrome; Dystonia

Camptodactyly

Camptodactyly, literally "bent finger," is a flexion deformity at the proximal interphalangeal joint, especially affecting the little fingers. A distinction is sometimes drawn between camptodactyly and streblodactyly: in the latter, several fingers are affected by flexion contractures (streblo = twisted, crooked), but it is not clear whether the two conditions overlap or are separate. The term streblomicrodactyly has sometimes been used to designate isolated crooked little fingers. Camptodactyly is not accompanied by any sensory or motor signs. Although some papers report camptodactyly to be usually unilateral, of 27 cases seen by the author in general neurology outpatient clinics over a 5 year period (2000-2004), most (24) referred for reasons other than finger deformity, 20 had bilateral changes, albeit asymmetric in some. The condition may be familial: in the author's series, other family members were affected by report or by examination in 11 out of 26 families represented. The condition is commoner in women (M:F = 9:18). X-linked dominant transmission has been suggested but there are occasional reports of father-to-child transmission. Camptodactyly may occur as part of a developmental disorder with other dysmorphic features or, as in all the cases observed by the author, in isolation.

It is important to differentiate camptodactyly, a nonneurogenic cause of clawing, from neurological diagnoses, such as:

Ulnar neuropathy
C8/T1 radiculopathy
Cervical rib
Syringomyelia

Awareness of the condition is important to avoid unnecessary neurological investigation.

References

Larner AJ. Camptodactyly in a neurology outpatient clinic. *International Journal of Clinical Practice* 2001; **55**: 592-595

Cross References

Claw hand

Capgras Syndrome

- see DELUSION

Carphologia

Carphologia, or floccillation, is an aimless plucking at clothing, as if picking off pieces of thread. This may sometimes be seen in psychiatric illness, delirium, Alzheimer's disease, or vascular dementia particularly affecting the frontal lobe.

Cross References

Delirium; Dementia

Carpopedal Spasm

- see *MAIN D'ACCOUCHEUR*

Catalepsy

This term has been used to describe increased muscle tone, leading to the assumption of fixed postures which may be held for long periods without fatigue. Clearly this term is cognate with or overlaps with waxy flexibility which is a feature of catatonic syndromes. The term should not be confused with cataplexy, a syndrome in which muscle tone is lost.

Cross References

Cataplexy; Catatonia

Cataplexy

Cataplexy is a sudden loss of limb tone which may lead to falls (drop attacks) without loss of consciousness, usually lasting less than 1 minute. Attacks may be precipitated by strong emotion (laughter, anger, embarrassment, surprise). Sagging of the jaw and face may occur, as may twitching around the face or eyelids. During an attack there is electrical silence in antigravity muscles, which are consequently hypotonic, and transient areflexia. Rarely status cataplecticus may develop, particularly after withdrawal of tricyclic antidepressant medication.

Cataplexy may occur as part of the narcoleptic syndrome of excessive and inappropriate daytime somnolence, hypnagogic hallucinations and sleep paralysis (Gélineau's original description of narcolepsy in 1877 included an account of "astasia" which corresponds to cataplexy). Symptomatic cataplexy occurs in certain neurological diseases including brainstem lesions, von Economo's disease (postencephalitic parkinsonism), Niemann-Pick disease type C, and Norrie's disease.

Therapeutic options for cataplexy include: tricyclic antidepressants, such as protriptyline, imipramine and clomipramine; serotonin reuptake inhibitors, such as fluoxetine; and noradrenaline and serotonin reuptake inhibitors, such as venlafaxine.

References

Houghton WC, Scammell TE, Thorpy M. Pharmacotherapy for cataplexy. *Sleep Medicine Reviews* 2004; **8**: 355-366

Cross References

Areflexia; Hypersomnolence; Hypotonia, Hypotonus

Catathrenia

Catathrenia is expiratory groaning during sleep, especially its later stages. Although sufferers are unaware of the condition, it does alarm relatives and bed partners. There are no associated neurological abnormalities, and no identified neurological or otorhinolaryngological cause. Catathrenia is a type of parasomnia.

References

Vetrugno R, Provini F, Plazzi G, Vignatelli L, Lugaresi E, Montagna P. Catathrenia (nocturnal groaning): a new type of parasomnia. *Neurology* 2001; **56**: 681-683

Catatonia

Catatonia is a clinical syndrome, first described by Kahlbaum (1874), characterized by a state of unresponsiveness but with maintained,

immobile, body posture (sitting, standing; *cf*. stupor), mutism, and refusal to eat or drink, with or without staring, grimacing, limb rigidity, maintained abnormal postures (waxy flexibility or *flexibilitas cerea*), negativism, echophenomena (imitation behavior), stereotypy, and urinary incontinence or retention. After recovery patients are often able to recall events which occurred during the catatonic state (*cf*. stupor). "Lethal catatonia," in which accompanying fever and collapse lead to death, was described in the 1930's, and seems to resemble neuroleptic malignant syndrome; the name "malignant catatonia" has been proposed for this syndrome. Catatonia may be confused clinically with abulia.

Kraeplin classified catatonia as a subtype of schizophrenia but most catatonic patients in fact suffer a mood or affective disorder. Furthermore, although initially thought to be exclusively a feature of psychiatric disease, catatonia is now recognized as a feature of structural or metabolic brain disease. The original account contains descriptions suggestive of extrapyramidal disease.

Catatonia of psychiatric origin often responds to lorazepam; there are also advocates of ECT.

References

Fink M, Taylor MA. *Catatonia: a clinician's guide to diagnosis and treatment*. Cambridge: CUP, 2003

Kahlbaum K. *Catatonia*. Levij Y, Pridan T (trans.). Baltimore: Johns Hopkins University Press, 1973

Muqit MMK, Rakshi JS, Shakir RA, Larner AJ. Catatonia or abulia? A difficult differential diagnosis. *Movement Disorders* 2001; **16**: 360-362

Cross References

Abulia; Akinetic mutism; Imitation behavior; Mutism; Negativism; Rigidity; Stereotypy; Stupor

Cauda Equina Syndrome

A cauda equina syndrome results from pathological processes affecting the spinal roots below the termination of the spinal cord around L1/L2, hence it is a syndrome of multiple radiculopathies.

Depending on precisely which roots are affected, this may produce symmetrical or asymmetrical sensory impairment in the buttocks (saddle anesthesia; sacral anesthesia) and the backs of the thighs, radicular pain, and lower motor neurone type weakness of the foot and/or toes (even a flail foot). Weakness of hip flexion (L1) does not occur, and this may be useful in differentiating a cauda equina syndrome from a conus lesion which may otherwise produce similar features. Sphincters may also be involved, resulting in incontinence, or, in the case of large central disc herniation at L4/L5 or L5/S1, acute urinary retention.

Causes of a cauda equina syndrome include:
Central disc herniation
Tumor: primary (ependymoma, meningioma. Schwannoma), metastasis
Hematoma

Abscess
Lumbosacral fracture
Inflammatory disease, *e.g.,* sarcoidosis (rare)
Ankylosing spondylitis (rare).

The syndrome needs to be considered in any patient with acute (or acute-on-chronic) low back pain, radiation of pain to the legs, altered perineal sensation, and altered bladder function. Missed diagnosis of acute lumbar disc herniation may be costly, from the point of view of both clinical outcome and resultant litigation.

References

Ahn UM, Ahn NU, Buchowski JM, Garrett ES, Sieber AN, Kostiuk JP. Cauda equina syndrome secondary to lumbar disc herniation: a meta-analysis of surgical outcomes. *Spine* 2000; **25**: 1515-1522

Markham DE. Cauda equina syndrome: diagnosis, delay and litigation risk. *Journal of the Medical Defence Union* 2004; **20(1)**: 12-15

Cross References

Bulbocavernosus reflex; Foot Drop; Incontinence; Radiculopathy; Urinary retention

Central Scotoma, Centrocecal Scotoma

These visual field defects are typical of retinal or optic nerve pathology. They may be mapped by confrontation testing or automatically.

- *Central scotoma*:

Field defect occupying the macula, due to involvement of the macula or the papillomacular bundle; this is the typical (but not exclusive) finding in optic neuritis, but may also be seen with disease of the macula, optic nerve compression, Leber's hereditary optic neuropathy. Examination for a concurrent contralateral superior temporal defect should be undertaken: such junctional scotomas may be seen with lesions at the anterior angle of the chiasm.

- *Centrocecal or cecocentral scotoma*:

Field defect involving both the macula and the blind spot; seen in optic nerve disease, such as Leber's hereditary optic neuropathy, toxic or nutritional optic neuropathies (said to be typical of vitamin B_{12} deficiency optic neuropathy), sometimes in optic neuritis.

Cross References

Junctional scotoma, Junctional scotoma of traquair; Scotoma; Visual field defects

Cerebellar Syndromes

Differing clinical pictures may be seen with pathology in different parts of the cerebellum. Broadly speaking, a midline cerebellar syndrome (involving the vermis) may be distinguished from a hemispheric cerebellar syndrome (involving the hemispheres). Their clinical characteristics are:

- *Midline cerebellar syndrome*:

Gait ataxia but with little or no limb ataxia, hypotonia, or nystagmus (because the vestibulocerebellum is spared), or dysarthria;

causes include alcoholic cerebellar degeneration, tumor of the midline (*e.g.,* medulloblastoma), paraneoplastic cerebellar degeneration.

- *Hemispheric cerebellar syndrome*:

Limb ataxia (*e.g.,* ataxia on finger-nose and/or heel-shin testing), dysdiadochokinesia, dysmetria, dysarthria, nystagmus; usual causes are infarcts, hemorrhages, demyelination, and tumors.

- *Pancerebellar syndrome*:

Affecting all parts of the cerebellum, and showing a combination of the above signs (*e.g.,* cerebellar degenerations).

References

Holmes G. The Croonian lectures on the clinical symptoms of cerebellar disease and their interpretation. *Lancet* 1922: **i**: 1177-1182; 1231-1237; **ii**: 59-65; 111-115

Cross References

Asynergia; Ataxia; Dysarthria; Dysdiadochokinesia; Dysmetria; Hemiataxia; Hypotonia, Hypotonus; Nystagmus

Cerebellopontine Angle Syndrome

Lesions of the cerebellopontine angle produce a constellation of ipsilateral signs:

Depressed corneal reflex, impaired corneal sensation (early sign)
Lower motor neurone facial (VII) weakness
Sensorineural hearing loss (VIII)
Hemiataxia.

The most common causes of this syndrome are acoustic neuroma (schwannoma) or meningioma; occasional causes include dermoids, epidermoids (cholesteatoma), and chordoma.

Cross References

Corneal reflex; Facial paresis; Hemiataxia

Chaddock's Sign

Chaddock's sign, or the external malleolar sign, is a variant method for eliciting the plantar response, by application of a stimulus in a circular direction around the external malleolus, or the lateral aspect of the foot, moving from heel to little toe. Extension of the hallux (upgoing plantar response, Babinski's sign) is pathological, indicating corticospinal tract (upper motor neurone) pathology. The development of Babinski's sign always predates that of Chaddock's sign.

References

Chaddock CG. A preliminary communication concerning a new diagnostic nervous sign. *Interstate Medical Journal* 1911; **18**: 742-746

Van Gijn J. *The Babinski sign: a centenary*. Utrecht: Universiteit Utrecht, 1996

Cross References

Babinski's sign (1); Gordon's sign; Oppenheim's sign; Plantar response; Upper motor neurone (UMN) syndrome

Charcot Joint

Charcot joint, or neuropathic joint, describes a destructive arthropathy seen following repeated injury to an anesthetic joint in patients with impaired or absent pain sensation. There is trophic change, with progressive destruction of articular surfaces with disintegration and reorganization of joint structure. Although the destruction is painless, the Charcot joint itself may be painful. There may be concurrent skin ulceration. Charcot joints were originally described in the context of tabes dorsalis (knees, shoulders, elbows, hips, ankles) but they may also be seen in:

Syringomyelia (elbow)
Hereditary sensory (and autonomic) neuropathies (HSAN, "congenital insensitivity to pain"; ankles)
Leprosy
Diabetes mellitus

References

Pearce JMS. Charcot joints. In: Pearce JMS. *Fragments of neurological history*. London: Imperial College Press, 2003: 563-565

Cross-References

Analgesia*; Main succulente*

Charles Bonnet Syndrome

Described by the Swiss naturalist and philosopher Charles Bonnet in 1760, this syndrome consists of well-formed (complex), elaborate and often stereotyped visual hallucinations, of variable frequency and duration, in a partially sighted (usually elderly) individual who has insight into their unreality. Hallucinations may disappear on eye closure. Predisposing visual disorders include cataract, macular degeneration, and glaucoma. There are no other features of psychosis or neurological disease, such as dementia. The pathogenesis of the visual hallucinations is uncertain. Reduced stimulation of the visual system leading to increased cortical hyperexcitability is one possible explanation (the deafferentation hypothesis), although the syndrome may occasionally occur in people with normal vision. Treatment consists primarily of reassurance. Pharmacological treatment with atypical antipsychotics or anticonvulsants may be tried but there is no secure evidence base.

References

Jacob A, Prasad S, Boggild M, Chandratre S. Charles Bonnet syndrome – elderly people and visual hallucinations. *BMJ* 2004; **328**: 1552-1554

Menon GJ, Rahman I, Menon SJ, Dutton GN. Complex visual hallucinations in the visually impaired: the Charles Bonnet syndrome. *Survey of Ophthalmology* 2003; **48**: 58-72

Cross References

Hallucinations

Chasm

- see YAWNING

Cheiro-Oral Syndrome
- see PSEUDORADICULAR SYNDROME

Cherry Red Spot at the Macula
The appearance of a "cherry red spot at the macula," caused by the contrast of a red macula against retinal pallor, occurs in a number of metabolic storage disorders, including:

Sialidosis (type I = cherry red spot-myoclonus syndrome)
Gangliosidoses (*e.g.,* Tay-Sachs disease: Tay's sign)
Metachromatic leukodystrophy
Niemann-Pick disease (especially type A)

Storage of sphingolipids or other substances in ganglion cells in the perimacular region gives rise to the appearance.

References

Kivlin JD, Sanborn GE, Myers GG. The cherry-red spot in Tay-Sachs and other storage diseases. *Annals of Neurology* 1985; **17**: 356-360

Cross References
Maculopathy

Cheyne-Stokes Breathing
- see PERIODIC RESPIRATION

"Chicken Wings"
In facioscapulohumeral (FSH) muscular dystrophy, the bulk of the deltoid and forearm muscles is normally well preserved, while biceps and triceps are wasted (and may be weak), thus giving rise to an appearance of the upper limbs sometimes labeled as "chicken wings" or "Popeye arms."

Cross References
Winging of the scapula

Chorea, Choreoathetosis
Chorea is an involuntary movement disorder characterized by jerky, restless, purposeless movements (literally dance-like) that tend to flit from one part of the body to another in a rather unpredictable way, giving rise to a fidgety appearance. There may also be athetoid movements (slow, sinuous, writhing), jointly referred to as choreoathetosis. Severe proximal choreiform movements of large amplitude ("flinging") are referred to as ballism or ballismus. When, as is often the case, such movements are confined to one side of the body they are referred to as hemichorea-hemiballismus. There may be concurrent abnormal muscle tone, either hypotonia or rigidity. Hyperpronation of the upper extremity may be seen when attempting to maintain an extended posture.

The pathophysiology of chorea (as for ballismus) is unknown; movements may be associated with lesions of the contralateral subthalamic nucleus, caudate nucleus, putamen, and thalamus. One model

of basal ganglia function suggests that reduced basal ganglia output to the thalamus disinhibits thalamic relay nuclei leading to increased excitability in thalamocortical pathways which passes to descending motor pathways resulting in involuntary movements.

Recognized causes of chorea and choreoathetosis are many, including:

- Hereditary:
  - Huntington's disease (HD)
  - Dentatorubropallidoluysian atrophy (DRPLA)
  - Neuroacanthocytosis
  - Benign hereditary chorea

Paroxysmal dyskinesias: paroxysmal kinesigenic choreoathetosis (PKC) and paroxysmal dystonic choreoathetosis (PDC)

- Sporadic:
  - Drugs: levodopa therapy in later stages of idiopathic Parkinson's disease
  - Pregnancy: chorea gravidarum
  - Hyperthyroidism
  - Systemic lupus erythematosus (SLE)
  - Sydenham's chorea (post-infectious, rheumatic chorea, St. Vitus dance, PANDAS)
  - Polycythemia rubra vera (hyperviscosity)
  - AIDS
  - Hyperosmolality (hyperglycemia, hypernatremia)
  - CNS tumor
  - Multiple sclerosis (rare)
  - Variant Creutzfeldt-Jakob disease
  - "Senile chorea" (diagnosis of exclusion, especially of HD)

Where treatment is necessary, antidopaminergic agents, such as dopamine receptor antagonists (*e.g.,* neuroleptics, sulpiride, risperidone) and dopamine depleting agents (*e.g.,* tetrabenazine, reserpine) may help, although they may cause parkinsonism, akathisia, neuroleptic malignant syndrome, and sedation. Chronic neuroleptic use may also cause chorea, but these movements are repetitive and predictable, unlike "classic" chorea.

References

Barker R. Chorea: diagnosis and management. *Advances in Clinical Neuroscience & Rehabilitation* 2003; **3(4)**: 19-20

Sawle G. Chorea. In: Sawle G (ed.). *Movement disorders in clinical practice*. Oxford: Isis Medical Media, 1999: 119-133

Schrag A, Quinn N. Huntington's disease and other choreas. *Journal of Neurology* 1998; **245**: 709-716

Cross References

Athetosis; Ballism, Ballismus; Dyskinesia; Hypotonia, Hypotonus; Milkmaid's grip; PseudoChoreoathetosis; Rigidity; Trombone tongue

Chromesthesia

- see SYNESTHESIA

Chvostek's Sign

Chvostek's sign is contraction of facial muscles provoked by lightly tapping over the facial nerve as it crosses the zygomatic arch. Chvostek's sign is observed in hypocalcemic states, such as hypoparathyroidism and the respiratory alkalosis associated with hyperventilation. There may be concurrent posturing of the hand, known as *main d'accoucheur* for its resemblance to the posture adopted for manual delivery of a baby. The pathophysiology of this mechanosensitivity of nerve fibers is uncertain, but is probably related to increased discharges in central pathways. Although hypocalcemia might be expected to impair neuromuscular junction transmission and excitation-contraction coupling (since Ca^{2+} ions are required for these processes) this does not in fact occur.

Cross References

Main d'accoucheur; Spasm

Ciliospinal Response

The ciliospinal response consists of rapid bilateral pupillary dilatation and palpebral elevation in response to a painful stimulus in the mantle area, for example pinching the skin of the neck.

References

Reeves AG, Posner JB. The ciliospinal response in man. *Neurology* 1969; **19**: 1145-1152

Cross References

Pupillary reflexes

Circumlocution

Circumlocution refers to:

- A discourse that wanders from the point, only eventually to return to the original subject matter, as seen in fluent aphasias;
- A response to word-finding difficulties, as in early Alzheimer's disease or nonfluent aphasias: in response to familiar pictures, patients may comment that the name is on the tip-of-the-tongue but they cannot access it, and therefore give alternatives (*e.g.,* "gardener's friend" or "beetle" for ladybird).

References

Astell AJ, Harley TA. Tip of the tongue states in lexical access in dementia. *Brain and Language* 1996; **54**: 196-215

Cross References

Anomia; Aphasia; Dementia

Clasp-Knife Phenomenon

Clasp-knife phenomenon is the name sometimes applied to the sudden "give" encountered when passively moving a markedly spastic limb. Since the clasp-knife phenomenon is a feature of spasticity, the term "clasp-knife rigidity" is probably best eschewed to avoid possible confusion.

Cross References

Rigidity; Spasticity

Claudication

Claudication (literally limping, Latin *claudicatio*) refers to intermittent symptoms of pain secondary to ischemia. Claudication of the legs on walking is a symptom of peripheral vascular disease. Claudication of the jaw, tongue, and limbs (especially upper) may be a feature of giant cell arteritis.

References

Caselli RJ, Hunder GG, Whisnant JP. Neurologic disease in biopsy-proven giant cell (temporal) arteritis. *Neurology* 1988; **38**: 352-359

Claw Foot

Claw foot, or *pied en griffe*, is an abnormal posture of the foot, occurring when weakness and atrophy of the intrinsic foot muscles allows the long flexors and extensors to act unopposed, producing shortening of the foot, heightening of the arch, flexion of the distal phalanges and dorsiflexion of the proximal phalanges (*cf*. pes cavus). This may occur in chronic neuropathies of early onset which involve motor fibers, such as hereditary motor and sensory neuropathies types I and II.

Cross References

Pes cavus

Claw Hand

Claw hand, or *main en griffe*, is an abnormal posture of the hand with hyperextension at the metacarpophalangeal joints (5th, 4th, and, to a lesser extent, 3rd finger) and flexion at the interphalangeal joints. This results from ulnar nerve lesions above the elbow, or injury to the lower part of the brachial plexus (Dejerine-Klumpke type), producing wasting and weakness of hypothenar muscles, interossei, and ulnar (medial) lumbricals, allowing the long finger extensors and flexors to act unopposed.

Cross References

Benediction hand; Camptodactyly

Clonus

Clonus is rhythmic, involuntary, and repetitive muscular contraction and relaxation. It may be induced by sudden passive stretching of a muscle or tendon, most usually the Achilles tendon (ankle clonus) or patella (patellar clonus). Ankle clonus is best elicited by holding the relaxed leg underneath the moderately flexed knee, then quickly dorsiflexing the ankle and holding it dorsiflexed. A few beats of clonus is within normal limits but sustained clonus is pathological.

Clonus reflects hyperactivity of muscle stretch reflexes and may result from self reexcitation. It is a feature of upper motor neurone disorders affecting the corticospinal (pyramidal) system. Patients with disease of the corticospinal tracts may describe clonus as a rhythmic jerking of the foot, for example when using the foot pedals of a car. Clonus may also be observed as part of a generalized (primary or secondary) epileptic seizure, either in isolation (clonic seizure) or much more commonly following a tonic phase (tonic-clonic seizure). The clonic movements

usually involve all four limbs and decrease in frequency and increase in amplitude over about 30-60 seconds as the attack progresses. Rather different "clonic" movements may occur in nonepileptic seizures. A few clonic jerks may also be observed in syncopal attacks, leading the uninitiated to diagnose "seizure" or "convulsion."

Cross References

Myoclonus; Seizure; Upper motor neurone (UMN) syndrome

"Closing-in" Sign

Copying of drawings that are close to or superimposed on the original has been referred to as the "closing-in" sign. It may be seen in patients with Alzheimer's disease with deficits in visuospatial function. This has sometimes been characterized as one aspect of the "constructional apraxia" of Alzheimer's disease; it may be useful in differentiating AD from subcortical vascular dementia.

References

Kwak YT. "Closing-in" phenomenon in Alzheimer's disease and subcortical vascular dementia. *European Journal of Neurology* 2004; **11(suppl2)**: 47 (P1037)

Mayer-Gross W. Some observations on apraxia. *Proceedings of the Royal Society of Medicine* 1935; **28**: 1203-1212

Coactivation Sign

This sign is said to be characteristic of psychogenic tremors, namely increased tremor amplitude with loading (*cf*. reduced amplitude of organic tremor with loading), perhaps due to muscle coactivation to maintain oscillation.

References

Deuschl G, Koster B, Lucking CH, Scheidt C. Diagnostic and pathophysiological aspects of psychogenic tremors. *Movement Disorders* 1998; **13**: 294-302

Cross References

Tremor

Cock Walking

- see TOE WALKING

Cogan's (Lid Twitch) Sign

Cogan's sign is a twitching of the upper eyelid seen a moment after the eyes are moved from downgaze to the primary position. Twitches may also be seen with eye closure after sustained upgaze. These phenomena are said to be characteristic signs of ocular myasthenia gravis, and were found in 60% of myasthenics in one study. They may also occur occasionally in other oculomotor brainstem disorders, such as Miller Fisher syndrome, but are not seen in normals.

Cogan's sign should not be confused with either Cogan's syndrome, an autoimmune disorder of episodic vertigo, tinnitus, hearing loss and interstitial keratitis; or the oculomotor apraxia of Cogan, a congenital lack of lateral gaze.

References

Cogan DG. Myasthenia gravis: a review of the disease and a description of lid twitch as a characteristic sign. *Archives of Ophthalmology* 1965; **74**: 217-221

Whye Onn H, Cleary M, Metcalfe R. Cogan's lid twitch revisited. *Journal of Neurology, Neurosurgery and Psychiatry* 2004; **75**: 805 (abstract 082)

Cross References

Fatigue; Ice pack test; Ocular apraxia

Cogwheeling, Cogwheel Phenomenon, Cogwheel Rigidity

- see RIGIDITY; SACCADIC INTRUSION; SACCADIC PURSUIT

"Cold Hands Sign"

In multiple system atrophy (MSA), the hands may be cold, dusky, and violaceous with poor circulatory return after blanching by pressure, suggesting defective neurovascular control of the distal extremities as one feature of the autonomic dysfunction in MSA. The findings are not present in idiopathic Parkinson's disease.

References

Klein C, Brown R, Wenning G, Quinn N. The "cold hands sign" in multiple system atrophy. *Movement Disorders* 1997; **12**: 514-518

Collapsing Weakness

Collapsing weakness, or "give-way" weakness, suggesting intermittent voluntary effort, is often taken as a sign of functional weakness. Although sometimes labeled as "volitional weakness," it is not clear that such weakness is in any conscious sense willed, and it is therefore probably better to use a noncommittal term, such as "apparent weakness." Such collapsing weakness has also been recorded following acute brain lesions, such as stroke.

References

Gould R, Miller BL, Goldberg MA, Benson DF. The validity of hysterical signs and symptoms. *Journal of Nervous and Mental Disease* 1986; **174**: 593-597

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

Cross References

Functional weakness and sensory disturbance; Spasticity; Weakness; "Wrestler's sign"

Collier's Sign

Collier's sign ("posterior fossa stare," "tucked lid" sign) is elevation and retraction of the upper eyelids, baring the sclera above the cornea, with the eyes in the primary position or looking upward. This may be seen with upper dorsal midbrain supranuclear lesions (*e.g.,* "top of the basilar syndrome," Parinaud's syndrome). There may be accompanying paralysis of vertical gaze (especially upgaze) and light-near pupil-

lary dissociation. The sign is thought to reflect damage to the posterior commissure levator inhibitory fibers.

References

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Cross References

Lid retraction; Light-near pupillary dissociation; Parinaud's syndrome

Color Anomia

- see ACHROMATOPSIA; ANOMIA

Coma

Coma is a state of unresponsiveness, with eyes closed, from which a patient cannot be roused by verbal or mechanical stimuli. It represents a greater degree of impairment of consciousness than stupor or obtundation, all three forming part of a continuum, rather than discrete stages, ranging from alert and comatose. This lack of precision prompts some authorities to prefer the description of the individual aspects of neurological function in unconscious patients, such as eye movements, limb movements, vocalization, and response to stimuli, since this conveys more information than the use of terms, such as coma, stupor or obtundation, or the use of a lumped "score," such as the Glasgow Coma Scale.

These signs should be documented serially to assess any progression of coma. Assessment of the depth of coma may be made by observing changes in eye movements and response to central noxious stimuli: roving eye movements are lost before oculocephalic responses; caloric responses are last to go. The switch from flexor to extensor posturing (decorticate *vs*. decerebrate rigidity) also indicates increasing depth of coma.

There are many causes of coma, which may be broadly categorized as structural or toxic-metabolic; the latter are generally more slowly progressive and produce symmetrical signs, whereas structural lesions more often have an abrupt onset and some focal asymmetric findings on examination, but these distinctions are not absolute. Recognized causes of coma include:

- Structural:
  - Vascular insults (subarachnoid hemorrhage, cerebral infarction or hemorrhage)
  - Trauma
  - Tumor
  - Hydrocephalus
  - Vasculitides, leukodystrophies, leukoencephalopathies.
- Toxic-metabolic:
  - Drugs/toxins
  - Metabolic causes: for example, hypoxia, hypercapnia, hypoglycemia

Infections: for example, meningitis, encephalitis, sepsis
Epilepsy.

Unrousability which results from psychiatric disease, or which is being feigned ("pseudocoma"), also needs to be differentiated.

A number of neurobehavioral states may be mistaken for coma, including abulia, akinetic mutism, catatonia, and the locked-in syndrome.

EEG features may assist in differential diagnosis: prominent rhythmic beta activity raises the possibility of drug intoxication.

References

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

Rubino FA. Approach to the comatose patient. In: Biller J (ed.). *Practical neurology* (2nd edition). Philadelphia: Lippincott Williams & Wilkins, 2002: 54-65

Teasdale G, Jennett B. Assessment of coma and impaired consciousness: a practical scale. *Lancet* 1974; **2**: 81-84

Young GB, Ropper AH, Bolton CF (eds.). *Coma and impaired consciousness: a clinical perspective*. New York: McGraw-Hill, 1998

Cross References

Abulia; Akinetic mutism; Caloric testing; Catatonia; Decerebrate rigidity; Decorticate rigidity; Locked-in syndrome; Obtundation; Oculocephalic response; Roving eye movements; Stupor; Vegetative states; Vestibulo-ocular reflexes

"Compulsive Grasping Hand"

This name has been given to involuntary left hand grasping related to all right hand movements in a patient with a callosal hemorrhage. This has been interpreted as a motor grasp response to contralateral hand movements, and a variant of anarchic or alien hand. The description does seem to differ from that of behaviors labeled as forced groping and the alien grasp reflex (*q.v.*).

References

Kumral E. Compulsive grasping hand syndrome: a variant of anarchic hand. *Neurology* 2001; **57**: 2143-2144

Cross References

Alien hand, Alien limb; Intermanual conflict

Conduction Aphasia

Conduction aphasia is defined as a fluent aphasia with paraphasic errors (especially phonemic/literal) during speech, repetition and naming. In its "pure" form, there is a dissociation between relatively preserved auditory and reading comprehension of language and impaired repetition (in which the phenomenon of *conduit d'approche* may occur) and naming. Reading comprehension is good or normal, and is better than reading aloud which is impaired by paraphasic errors.

Conduction aphasia was traditionally explained as due to a disconnection between sensory (Wernicke) and motor (Broca) areas for

language, involving the arcuate fasciculus in the supramarginal gyrus. Certainly the brain damage (usually infarction) associated with conduction aphasia most commonly involves the left parietal lobe (lower postcentral and supramarginal gyri) and the insula, but it is variable, and the cortical injury may be responsible for the clinical picture.

Conduction aphasia is most often seen during recovery from Wernicke's aphasia, and clinically there is often evidence of some impairment of comprehension. If isolated, the prognosis for conduction aphasia is good.

References

Benson DF, Sheremata WA, Bouchard R, Segarra JM, Price D, Geschwind N. Conduction aphasia. A clinicopathological study. *Archives of Neurology* 1973; **28**: 339-346

Damasio H, Damasio AR. The anatomical basis of conduction aphasia. *Brain* 1980; **103**: 337-350

Cross References

Anomia; Aphasia; Broca's aphasia; *Conduit D'approche*; Paraphasia; Transcortical aphasias; Wernicke's aphasia

Conduit d'approche

Conduit d'approche, or "homing-in" behavior, is a verbal output phenomenon applied to patients with conduction aphasia attempting to repeat a target word, in which multiple phonemic approximations of the word are presented, with gradual improvement until the target word is achieved. This phenomenon suggests that an acoustic image of the target word is preserved in this condition. A similar phenomenon may be observed in patients with optic aphasia attempting to name a visual stimulus.

A similar behavior is seen in so-called speech apraxia, in which patients repeatedly approximate to the desired output before reaching it.

Cross References

Aphasia; Conduction aphasia; Optic aphasia; Speech apraxia

Confabulation

Confabulation is the falsification of episodic memory occurring in clear consciousness in association with amnesia; in other words, paramnesias are related as true events. However, most amnesic patients, suffering from medial temporal lobe/hippocampal lesions do not confabulate, and poor memory alone cannot explain confabulation. Concurrent hypothalamic/diencephalic and basal forebrain/frontal cortex lesions may be required to develop this syndrome: a functional imaging study of an amnesic patient found a correlation between the presence of orbital and medial frontal hypoperfusion and confabulation.

Confabulating patients often give a fairly coherent and entirely plausible account of events or experiences, sometimes in response to the examiner's suggestion.

Confabulations may be classified as:

- Momentary; or
- Fantastic: these may be of almost delusional intensity.

Confabulation is a classic feature of the Wernicke-Korsakoff syndrome, but is in fact rarely seen. It may also occur in cortical blindness (Anton's syndrome).

References

Benson DF, Djenderedjian A, Miller BL *et al*. Neural basis of confabulation. *Neurology* 1996; **46**: 1239-1243.
Berlyne N. Confabulation. *British Journal of Psychiatry* 1972; **120**: 31-39.
Downes JJ, Mayes AR. How bad memories can sometimes lead to fantastic beliefs and strange visions. In: Campbell R, Conway MA (eds.). *Broken memories: case studies in memory impairment*. Oxford: Blackwell, 1995: 115-123.

Cross References

Amnesia; Asomatognosia; Cortical blindness; Delusion; Paramnesia

Consensual Light Reflex

- see PUPILLARY REFLEXES

Constructional Apraxia

-see APRAXIA

Contracture

The term contracture may be used in various contexts:

- Clinically, to describe an acquired restriction of joint mobility (prenatally acquired restriction of joint mobility is called arthrogryposis). This may be due to a variety of factors, including prolonged muscle spasticity with or without muscle fibrosis (*i.e.,* without pathological muscle shortening), and ligamentous restrictions. This often occurs in the context of limb immobilization or inactivity, for example in a flexed posture. Injections of botulinum toxin to abolish muscle spasticity may be required to assess whether there is concurrent ligamentous restriction, and thus to plan optimum treatment, which may involve surgery. Contractures of muscular origin may be seen in conditions, such as Emery-Dreifuss disease (especially elbow, Achilles tendon, posterior part of neck) and Duchenne muscular dystrophy.
- Clinically, to describe a hard, contracted muscle, painful to straighten, and lasting several hours following exercise in a metabolic myopathy, such as McArdle's disease (myophosphorylase deficiency, glycogen storage disease type V); this may be associated with EMG silence.
- Physiologically, to describe a prolonged painful muscle spasm with EMG silence, as observed in myotonia and paramyotonia.

Cross References

Myotonia; Paramyotonia; Paraplegia; Spasm; Spasticity

Convergence-Retraction Nystagmus
- see NYSTAMGUS; PARINAUD'S SYNDROME

Coprolalia
Coprolalia is the use of expletives or other obscene language. This may be

- Vocal: involuntary utterance of obscenities
- Mental: compulsion to think obscenities

The former is a complex vocal tic most characteristically seen in Gilles de la Tourette syndrome although it actually occurs in less than half of affected individuals. Other disease associations are:

Lesch-Nyhan syndrome
Postencephalitic parkinsonism
Neuroacanthocytosis
Cingulate cortical seizures

The pathophysiology of coprolalia is unknown but may be related to frontal (cingulate and orbitofrontal) dysfunction, for which there is some evidence in Gilles de la Tourette syndrome.
Cross References
Tic

Copropraxia
Copropraxia is a complex motor tic comprising obscene gesturing, sometimes seen in Gilles de la Tourette syndrome.
Cross References
Coprolalia; Tic

Corectopia
Corectopia is pupillary displacement, which may be seen with midbrain lesions, including transtentorial herniation and top-of-the-basilar syndrome, peripheral oculomotor nerve palsies, and focal pathology in the iris.
References
Selhorst JB, Hoyt WF, Feinsod M, Hosobuch Y. Midbrain corectopia. *Archives of Neurology* 1976; **33**: 193-195

Corneal Reflex
The corneal reflex consists of a bilateral blink response elicited by touching the cornea lightly, for example, with a piece of cotton wool. As well as observing whether the patient blinks, the examiner should also ask whether the stimulus was felt: a difference in corneal sensitivity may be the earliest abnormality in this reflex. Synkinetic jaw movement may also be observed (see Corneomandibular Reflex).

The afferent limb of the corneal reflex is via the trigeminal (V) nerve, the efferent limb via the facial (VII) nerve to orbicularis oculi. The fibers subserving the corneal reflex seem to be the most sensitive to trigeminal nerve compression or distortion: an intact corneal reflex

with a complaint of facial numbness leads to suspicion of a nonorganic cause. Reflex impairment may be an early sign of a cerebellopontine angle lesion, which may also cause ipsilateral lower motor neurone type facial (VII) weakness and ipsilateral sensorineural hearing impairment (VIII). Trigeminal nerve lesions cause both ipsilateral and contralateral corneal reflex loss.

Cerebral hemisphere (but not thalamic) lesions causing hemiparesis and hemisensory loss may also be associated with a decreased corneal reflex.

The corneal reflex has a high threshold in comatose patients, and is usually preserved until late (unless coma is due to drug overdose), in which case its loss is a poor prognostic sign.

Cross References

Blink Reflex; Coma; Cerebellopontine angle syndrome; Corneomandibular reflex; Facial paresis

Corneomandibular Reflex

The corneomandibular reflex, also known as the corneopterygoid reflex or Wartenberg's reflex or sign, consists of anterolateral jaw movement following corneal stimulation. In one study, the corneomandibular reflex was observed in about three-quarters of patients with motor neurone disease (MND) who displayed no other pathological reflexes, a frequency much higher than that seen in patients with stroke causing hemiparesis or pseudobulbar palsy. It was therefore suggested to be a sensitive indicator of upper motor neurone involvement in MND.

References

Okuda B, Kodama N, Kawabata K, Tachibana H, Sugita M. Corneomandibular reflex in ALS. *Neurology* 1999; **52**: 1699-1701

Schott JM, Rossor MN. The grasp and other primitive reflexes. *Journal of Neurology, Neurosurgery and Psychiatry* 2003; **74**: 558-560

Cross References

Corneal reflex; Pseudobulbar palsy

Corneopterygoid Reflex

-see CORNEOMANDIBULAR REFLEX

Cortical Blindness

Cortical blindness is loss of vision due to bilateral visual cortical damage (usually hypoxic-ischemic in origin), or bilateral subcortical lesions affecting the optic radiations. A small central field around the fixation point may be spared (macula sparing). Pupillary reflexes are preserved but optokinetic nystagmus cannot be elicited.

Cortical blindness may result from:

Bilateral (sequential or simultaneous) posterior cerebral artery occlusion

"Top of the basilar syndrome"

Migraine

Cerebral anoxia

Bacterial endocarditis
Wegener's granulomatosis
Coronary or cerebral angiography (may be transient)
Epilepsy (transient)
Cyclosporin therapy, *e.g.,* following organ transplantation.

If acute in onset (*i.e.,* vascular), cortical blindness may ultimately evolve to prosopagnosia via visual object agnosia.

Patients with cortical blindness may deny their visual defect (Anton's syndrome, visual anosognosia) and may confabulate about what they "see."

Cross References

Anosognosia; Confabulation; Macula sparing, macula splitting; Optokinetic nystagmus, Optokinetic response; Prosopagnosia; pupillary reflexes; Visual agnosia

Coup de Sabre

Coup de sabre is a localized form of scleroderma manifest as a linear, atrophic lesion on the forehead which may be mistaken for a scar. This lesion may be associated with hemifacial atrophy and epilepsy, and neuroimaging may show hemiatrophy and intracranial calcification. Whether these changes reflect inflammation or a neurocutaneous syndrome is not known.

References

Duyff RF, Vos J. A "scar" and epilepsy: coup de sabre. *Journal of Neurology, Neurosurgery and Psychiatry* 1998; **65**: 568

Cross References

Hemifacial atrophy

Cover Tests

The simple cover and cover-uncover tests may be used to demonstrate manifest and latent strabismus (heterotropia and heterophoria) respectively.

The cover test demonstrates tropias: the uncovered eye is forced to adopt fixation; any movement therefore represents a manifest strabismus (heterotropia).

The cover-uncover test demonstrates phorias: any movement of the covered eye to reestablish fixation as it is uncovered represents a latent strabismus (heterophoria). The alternate cover or cross cover test, in which the hand or occluder moves back and forth between the eyes, repeatedly breaking and reestablishing fixation, is more dissociating, preventing binocular viewing, and therefore helpful in demonstrating whether or not there is strabismus. It should be performed in the nine cardinal positions of gaze to determine the direction that elicits maximal deviation. However, it does not distinguish between tropias and phorias, for which the cover and cover-uncover tests are required.

Cross References

Heterophoria; Heterotropia

Cramp
- see FASCICULATION; SPASM; STIFFNESS

Cremasteric Reflex
The cremasteric reflex is a superficial or cutaneous reflex consisting of contraction of the cremaster muscle causing elevation of the testicle, following stimulation of the skin of the upper inner aspect of the thigh from above downwards (*i.e.,* the L1, L2 dermatomes, via the ilioinguinal and genitofemoral nerves).

The cremasteric reflex is lost when the corticospinal pathways are damaged above T12, or following lesions of the genitofemoral nerve. It may also be absent in elderly men, or with local pathology, such as hydrocele, varicocele, orchitis or epididymitis.

Cross References
Abdominal reflexes; Reflexes

Crocodile Tears
Crocodile tears, or Bogorad's syndrome, reflect inappropriate unilateral lacrimation during eating, such that tears may spill down the face (epiphora). This autonomic synkinesis is a striking but rare consequence of aberrant reinnervation of the facial (VII) nerve, usually after a Bell's palsy, when fibers originally supplying the salivary glands are re-routed to the lacrimal gland via the greater superficial petrosal nerve.

Cross References
Bell's palsy; Epiphora; Synkinesia, Synkinesis

Crossed Adductor Reflex
Contralateral adductor muscle contraction in response to a tap on the adductor tendon may be found with a pyramidal lesion above L2, although it is a normal finding in infants.

Cross References
Reflexes

Crossed Aphasia
Aphasia from a right-sided lesion in a right-handed patient, crossed aphasia, is rare, presumably a reflection of crossed or mixed cerebral dominance.

References
Bakar M, Kirshner HS, Wertz RT. Crossed aphasia: functional brain imaging with PET or SPECT. *Archives of Neurology* 1996; **53**: 1026-1032

Cross References
Aphasia

Crossed Apraxia
A name given to apraxia in right-handed patients with right-sided lesions; apraxia is more commonly associated with left-sided brain injury.

References

Raymer AM, Merians AS, Adair JC *et al*. Crossed apraxia: implications for handedness. *Cortex* 1999; **35**: 183-199

Cross References

Apraxia

Crossed Straight Leg Raising

- see LASÈGUE'S SIGN

Crying

- see AUTOMATISM; PATHOLOGICAL CRYING, PATHOLOGICAL LAUGHTER; SEIZURES

Cuirasse

- see SUSPENDED SENSORY LOSS

Czarnecki's Sign

Aberrant regeneration of the oculomotor (III) nerve to the iris sphincter may lead to gaze-evoked segmental constriction of the pupil, which may be visible only with slit-lamp examination.

D

Dalrymple's Sign
Dalrymple's sign is increased width of the palpebral fissure, often seen in hyperthyroidism.
Cross References
Lid retraction

Dazzle
Dazzle is 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 References
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/ischemia, 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.
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/ischemia, trauma, and drugs.

Cross References

Coma; Decerebrate rigidity

Déjà Entendu

A sensation of familiarity akin to *déjà vu* but referring to auditory 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. Recurrent hallucinations or vivid dream-like imagery may also enter the differential diagnosis.

Epileptic *déjà vu* may last longer and be more frequent, and may be associated with other features, such as depersonalization and derealization, strong emotion, such as fear, epigastric aura, or olfactory hallucinations. 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 lateralizing significance (right). *Déjà vécu* ("already lived") has been used to denote a broader experience than *déjà vu* but the clinical implications are similar.

Déjà vu may also occur with psychiatric illness, such as anxiety, depression, and schizophrenia.

References

Warren-Gash C, Zeman A. Déjà vu. *Practical Neurology* 2003; **3**: 106-109

Cross References

Aura; Hallucination; *Jamais vu*

Delirium

Delirium, also sometimes known as acute confusional state, acute organic reaction, acute brain syndrome, or toxic-metabolic encephalopathy, is a neurobehavioral 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
Behavioral disturbances: agitation, restlessness, aggression, wandering, which may occur as a consequence of perceptual problems;
Language: rambling incoherent speech, logorrhea
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 characterized 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 in the elderly delirium is often superimposed on dementia, which 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 categorized as either predisposing or precipitating.

- *Predisposing factors* include:
  - Age: frailty, physiological age rather than chronological
  - Sex: men > women
  - Neurological illness: dementia
  - Burden of comorbidity; 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, septicemia)
    - Metabolic: hypoxia, hypo-/hyperglycemia, hepatic failure, uremia, 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, orthopedic)

These precipitating factors merit treatment in their own right, and investigations should be tailored to identify these etiological factors. The EEG may show nonspecific 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 haloperidol may be used, probably in preference to atypical neuroleptics, benzodiazepines (lorazepam), or cholinesterase inhibitors.

References

Ashton H. Delirium and hallucinations. In: Perry E, Ashton H, Young A (eds.). *Neurochemistry of consciousness: neurotransmitters in mind*. Amsterdam: John Benjamins, 2002: 181-203

Burns A, Gallagley A, Byrne J. Delirium. *Journal of Neurology, Neurosurgery and Psychiatry* 2004; **75**: 362-367

Larner AJ. Delirium: diagnosis, aetiopathogenesis and treatment. *Advances in Clinical Neuroscience & Rehabilitation* 2004; **4(2)**: 28-29

Lindesay J, Rockwood K, Macdonald A (eds.). *Delirium in old age*. Oxford: OUP, 2003

Nayeem K, O'Keeffe ST. Delirium. *Clinical Medicine* 2003; **3**: 412-415

Cross References

Agraphia; Attention; Coma; Delusion; Dementia; Hallucination; Illusion; Logorrhea; 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 patients thoughts, ideas

Grandiose/expansive: occur particularly in mania

Guilt/worthlessness: occur 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

Tekin S, Cummings JL. Hallucinations and related conditions. In: Heilman KM, Valenstein E (eds.). *Clinical neuropsychology* (4th edition). Oxford: OUP, 2003: 479-494

Cross References

Delirium; Dementia; Hallucination; Illusion; Intermetamorphosis; Misidentification syndromes; Reduplicative paramnesia]

Dementia

Dementia is a syndrome characterized 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. Although commoner 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. Multiple neuropsychological tests are available to test different areas of cognition. 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.,* Huntington's disease, progressive supranuclear palsy). 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 labeled 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). Behavioral abnormalities are common in dementias due to degenerative brain disease, and may require treatment in their own right.

Recognized causes of a dementia syndrome include:

- Neurodegenerative diseases:
  Alzheimer's disease, frontotemporal lobar degenerations (frontotemporal dementia, encompassing Pick's disease; semantic dementia; primary progressive aphasia), 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, tumors.
- Infection: HIV dementia, neurosyphilis, Whipple's disease.
- Metabolic causes: Wernicke-Korsakoff syndrome, vitamin B_{12} deficiency, hypothyroidism, hyperparathyroidism/hypercalcemia, leukodystrophies, 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 rare. Specific treatments for 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 dementia with Lewy bodies and vascular dementia, for behavioral as well as mnestic features.

References

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Snowden JS, Neary D, Mann DMA. *Fronto-temporal lobar degeneration: fronto-temporal dementia, progressive aphasia, semantic dementia*. New York: Churchill Livingstone, 1996

Cross References

Agnosia; Amnesia; Aphasia; Apraxia; Aprosexia; Attention; Delirium; Dysmentia; Pseudodementia; Psychomotor retardation

De Musset's Sign

- see HEAD TREMOR

Developmental Signs

- see FRONTAL RELEASE SIGNS; PRIMITIVE REFLEXES

Diagonistic Dyspraxia

A dissociative phenomenon observed after callosotomy, probably identical to intermanual conflict.

References

Akelaitis AJ. Studies on the corpus callosum, IV: Diagonistic dyspraxia in epileptics following partial and complete section of the corpus callosum. *American Journal of Psychiatry* 1944-1945; **101**: 594-599

Cross References

Alien hand, Alien limb; Intermanual conflict

Diaphoresis

Diaphoresis is sweating, either physiological as in sympathetic activation (*e.g.,* during hypotension, hypoglycemia), or pathological (hyperhidrosis, *q.v.*). Diaphoresis may be seen 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 References

Hyperhidrosis

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 maximum. 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 nonfixing 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 nonfixing 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 ptosis (unilateral: oculomotor (III) nerve palsy; bilateral: myasthenia gravis), and head tilt or turn (*e.g.,* 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 long-standing. 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 (tumor, pseudotumor).

Divergence of the visual axes or ophthalmoplegia without diplopia suggests a long-standing 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

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Yee RD. Approach to the patient with diplopia. In: Biller J (ed.). *Practical neurology* (2nd edition). Philadelphia: Lippincott Williams & Wilkins, 2002: 147-161

Cross References

Abducens (vi) nerve palsy; Amblyopia; Bielschowsky's sign, Bielschowsky's test; Cover tests; Heterophoria; Heterotropia; Internuclear ophthalmoplegia (ino); Oculomotor (iii) nerve palsy

Disc Swelling

Swelling or edema 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 hemorrhages and loss of spontaneous retinal venous pulsation. Disc swelling due to edema must be distinguished from pseudopapilledema, elevation of the optic disc not due to edema, in which the nerve fibre layer is clearly seen.

Disc swelling may be due to raised intracranial pressure (papilledema, *q.v.*), or local inflammation of the optic nerve (papillitis), and may be associated with marked impairment of vision, for example in optic neuritis, or be without specific visual complaint (as may be the case in papilledema). The clinical history, visual acuity and visual fields may help determine the cause of disc swelling.

Recognized causes of disc swelling include:

- *Unilateral*:
  - Optic neuritis
  - Acute ischemic optic neuropathy (arteritic, nonarteritic)
  - Orbital compressive lesions, for example, 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 (papilledema; more usually bilateral)
- *Bilateral*:
  - Raised intracranial pressure (papilledema)
  - Malignant hypertension

Hypercapnia
High CSF protein, as in Guillain-Barré syndrome
Any of the unilateral causes

Cross References
Foster kennedy syndrome; Papilledema; Pseudopapilledema; Retinal venous pulsation; Visual field defects

Disinhibition

Disinhibited behavior is impulsive, showing poor judgment and insight; it may transgress normal cultural or social bounds. There is a loss of normal emotional and/or behavioral 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 (frontotemporal dementia, Alzheimer's disease), mass lesions, or be a feature of epileptic seizures.

Cross References
Attention; Emotionalism, Emotional lability; Frontal lobe syndromes; 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 fibers 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), while 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, while 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 fibers 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

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 muscular groups allows diagnosis of a divisional palsy and suggests pathology at the superior orbital fissure or anterior cavernous sinus. However, occasionally this division may occur more proximally, at the fascicular level (*i.e.,* within the midbrain) or within the subarachnoid space, giving a false-localizing divisional palsy. This may reflect the topographic arrangement of axons within the oculomotor nerve.

References

Larner AJ. Proximal superior division oculomotor nerve palsy from metastatic subarachnoid infiltration *Journal of Neurology* 2002; **249**: 343-344

Cross References

"False-localizing signs"; Oculomotor (iii) nerve palsy

Dix-Hallpike Positioning Test

- see HALLPIKE MANEUVER, HALLPIKE TEST

Doll's Eye Maneuver, Doll's Head Maneuver

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 maneuver 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 maneuver may be used in conscious patients to assess vestibulo-ocular reflexes. While directly observing the eyes, "catch up" saccades may be seen in the absence of VOR. Measuring visual acuity (dynamic visual acuity, or illegible E test) two to three lines may be dropped on visual acuity with head movement compared to visual acuity with the head still if VOR is impaired. On ophthalmoscopy, the disc moves with the head if VOR is lost.

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 tumors at the foramen magnum may also produce this picture (remote atrophy, a "false-localizing sign"). In many elderly people the extensor tendons are prominent in the absence of significant muscle wasting.

Cross References

Wasting

"Double Elevator Palsy"

This name has been given to monocular elevation paresis. It may occur in association with pretectal supranuclear lesions 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.

References

Thömke F, Hopf HC. Acquired monocular elevation paresis. An asymmetric up-gaze palsy. *Brain* 1992; **115**: 1901-1910

Cross References

Bell's phenomenon, Bell's sign

Downbeat Nystagmus

- see NYSTAGMUS

Dressing Apraxia

- see APRAXIA

Drooling

- see SIALORRHEA

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 dementia with motor neurone disease, FTD/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 etiology but which may on occasion be steroid-responsive ("bent spine syndrome" or camptocormia may be a related form 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**: 917-921

Nicholas RS, Lecky BRF. Dropped head syndrome: the differential diagnosis. *Journal of Neurology, Neurosurgery and 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**: 115-118

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

Cross References

Antecollis; Camptocormia; Myopathy

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 no paraphasias, minimal anomia, preserved repetition and automatic speech. "Incorporational echolalia," when the patient uses the examiner's question to help form an answer, may be observed. Dynamic aphasia may be conceptualized as a variant of transcortical motor aphasia, and may be seen with lesions of dorsolateral prefrontal cortex ("frontal aphasia").

References

Alexander MP. Transcortical motor aphasia: a disorder of language production. In: D'Esposito M (ed.). *Neurological foundations of cognitive neuroscience*. Cambridge: MIT Press, 2003: 165-174

Esmonde T, Giles E, Xuereb J, Hodges J. Progressive supranuclear palsy presenting with dynamic aphasia. *Journal of Neurology, Neurosurgery and Psychiatry* 1996; **60**: 403-410

Robinson G, Blair J, Cipolotti L. Dynamic aphasia: an inability to select between competing verbal responses. *Brain* 1998; **121**: 77-89

Cross References

Echolalia; Transcortical aphasias

Dysarthria

Dysarthria is a motor speech disorder of neurological origin (*cf*. dysphonia due to primary laryngeal pathology), causing impaired motor control (articulation) of the speech musculature. There is no language disturbance (*cf*. aphasia, although the two may coexist).

There are various syndromes of dysarthria, which have been classified as follows:

- *Flaccid or nasal dysarthria*:
  hypernasal, breathy, whining output, as in bulbar palsy, 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 Gilles de la Tourette syndrome.
- *Mixed dysarthria*:
  combination of any of above.

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 (ed.). *Practical neurology* (2nd edition). Philadelphia: Lippincott Williams & Wilkins, 2002: 236-243

Murdoch BE (ed.). *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; Upper motor neurone (UMN) syndrome

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 or basal ganglia.

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

Dysesthesia
Dysesthesia is 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 (paresthesia). Dysesthesia differs from paresthesia in its unpleasant quality, but may overlap in some respects with allodynia, hyperalgesia and hyperpathia (the latter phenomena are provoked by stimuli, either nonnoxious or noxious).

There are many causes of dysesthesia, both peripheral (including small fibre neuropathies, neuroma, nerve trauma) and central (*e.g.*, spinal multiple sclerosis). Dysesthetic sensations may be helped by agents, such as carbamazepine, amitriptyline, gabapentin and pregabalin.

Cross References
Allodynia; Hyperalgesia; Hyperpathia; Paresthesia

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 behavior. 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.

References
Knight RT, D'Esposito M. Lateral prefrontal syndrome: a disorder of executive control. In: D'Esposito M (ed.). *Neurological foundations of cognitive neuroscience*. Cambridge: MIT Press, 2003: 259-279

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.

- Henkin RI, Patten BM, Pe RK, Bronzert DA. A syndrome of acute zinc loss. Cerebellar dysfunction, mental changes, anorexia and taste and smell dysfunction. *Archives of Neurology* 1975; **32**: 745-751

Cross References

Ageusia; Anosmia

Dysgraphesthesia

- see AGRAPHOGNOSIA; GRAPHESTHESIA

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) and occasionally trunk and arms; usually persists after withdrawal of causative therapy; clonazepam, baclofen, and tetrabenazine may help.
- *Paroxysmal dyskinesias*:
  Paroxysmal kinesigenic choreoathetosis/dystonia (PKC; usually responds to carbamazepine), and paroxysmal nonkinesigenic dystonia/choreoathetosis (PDC; does not respond to carbamazepine).
- *Focal dyskinesias*:
  Orofacial dyskinesia, belly-dancer's dyskinesia.

References
Fahn S. The paroxysmal dyskinesias. In: Marsden CD, Fahn S (eds.). *Movement disorders 3*. Oxford: Butterworth-Heinemann, 1994: 310-345
Wojcieszek J. Drug-induced movement disorders. In: Biller J (ed.). *Iatrogenic neurology*. Boston: Butterworth-Heinemann, 1998: 215-231
Cross References
Athetosis; Chorea, Choreoathetosis; Dystonia; Hyperekplexia; 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 References
Alexia

Dysmentia

The term dysmentia has been suggested as an alternative to dementia, to emphasize the possibility of treating and preventing cognitive decline.
References
Chiu E. What's in a name: dementia or dysmentia? *International Journal of Geriatric Psychiatry* 1994; **9**: 1-4
Cross References
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 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 coordinated muscular contraction.
References
Bötzel K, Rottach K, Büttner U. Normal and pathological saccadic dysmetria. *Brain* 1993; **116**: 337-353
Büttner U, Straube A, Spuler A. Saccadic dysmetria and "intact" smooth pursuit eye movements after bilateral deep cerebellar nuclei lesions. *Journal of Neurology, Neurosurgery and Psychiatry* 1994; **57**: 832-834
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, color 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.

References

Milner AD, Perrett DI, Johnston RS, *et al*. Perception and action in "visual form agnosia." *Brain* 1991; **114**: 405-428

Cross References

Agnosia; Visual agnosia

Dysnomia

- see ANOMIA

Dysphagia

Dysphagia is difficulty swallowing. This may have local mechanical causes which are usually gastroenterological in origin (tumor; peptic ulceration/stricture, in which case there may be additional pain on swallowing -odynophagia) but sometimes vascular (aberrant right subclavian artery – dysphagia lusoria). 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.
Recognized neurological causes of dysphagia include:

- Upper motor neurone pathology: pseudobulbar palsy, *e.g.*, motor neurone disease, bilateral cerebrovascular disease, multiple sclerosis
- Lower motor neurone pathology: bulbar palsy, isolated vagus (X) nerve palsy, jugular foramen syndrome
- Autonomic neuropathy, *e.g.*, Chagas' disease, Riley Day syndrome
- Neuromuscular junction pathology: myasthenia gravis
- Muscular pathology: polymyositis, oculopharyngeal muscular dystrophy.

Difficulty swallowing may on occasion be functional in origin (globus hystericus).

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.

References
Hamdy S, Aziz Q, Rothwell JC, Singh KD, Barlow J, Hughes DG, Tallis RC, Thompson DG. The cortical topography of human swallowing musculature in health and disease. *Nature Medicine* 1996; **2**: 1217-1224
Logemann JA. Approach to the patient with dysphagia. In: Biller J (ed.). *Practical neurology* (2nd edition). Philadelphia: Lippincott Williams & Wilkins, 2002: 227-235
Cross References
Bulbar palsy; Dysphonia; Gag reflex; Jugular foramen syndrome; Pseudobulbar palsy

Dysphasia
- see 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 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).

Recognized 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.

References
Whurr R, Lorch M, Fontana H, Brookes G, Lees A, Marsden CD. The use of botulinum toxin in the treatment of adductor spasmodic dysphonia. *Journal of Neurology, Neurosurgery and Psychiatry* 1993; **56**: 526-530
Cross References
Aphonia; Bulbar palsy; Diplophonia; Dysarthria; Dystonia; Hypophonia; Vocal tremor, Voice tremor

Dyspraxia
Dyspraxia is difficulty or impairment in the performance of a 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 References

Apraxia

Dysprosody

- see APROSODIA, APROSODY

Dyssynergia

- see ASYNERGIA

Dystaxia

- see ATAXIA

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, generalized, hemidystonia;

Etiology: 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 (more than 40 known causes), including:

Heredodegenerative disorders: Wilson's disease, Huntington's disease, Hallervorden-Spatz disease, mitochondrial disorders, X-linked dystonia-parkinsonism (lubag)

Paroxysmal dystonias/dyskinesias: paroxysmal kinesigenic choreoathetosis/dystonia (PKC; usually responds to carbamazepine), and paroxysmal nonkinesigenic 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 poorly understood. Different mechanism 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 characterization of various dystonic syndromes may facilitate understanding of pathogenesis.

From a 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 minutes 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

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Németh AH. The genetics of primary dystonias and related disorders. *Brain* 2002; **125**: 695-721

Van Harten PN, Hoek HW, Kahn RS. Acute dystonia induced by drug treatment. *BMJ* 1999; **319**: 623-626

Cross References

Anismus; Blepharospasm; Dysphonia; Eyelid apraxia; Fatigue; Gaping; Geste antagoniste; Hemidystonia; Torticollis; Writer's cramp

E

Ear Click
- see PALATAL MYOCLONUS; TINNITUS

Echolalia
Echolalia is the involuntary automatic repetition of an interviewer's speech. This may be observed in a variety of clinical situations:

- Transcortical sensory aphasia:
  In the context of a fluent aphasia with repetition often well or normally preserved, usually as a result of a vascular lesion of the left hemisphere although an analogous situation may be encountered in Alzheimer's disease; "incorporational echolalia," when the patient uses the examiner's question to help form an answer, may be observed as a feature of "dynamic aphasia" which bears resemblance to transcortical motor aphasia, but may result from a frontal lesion.
- Transcortical motor aphasia:
  "Effortful echolalia" has been reported in the context of infarction of the left medial frontal lobe, including the supplementary motor area, showing that neither the ability to repeat nor fluent speech is required for echolalia.
- Gilles de la Tourette syndrome:
  As a complex vocal tic, along with coprolalia.
- Alzheimer's disease, Pick's disease:
  As a symptom of dementia.
- Schizophrenia:
  As a catatonic symptom.
- Early infantile autism, mental retardation:
  As a reflection of pathological mental development.
- Frontal lobe lesions:
  As a feature of imitation behavior.
- Normal children:
  At a particular stage of language acquisition.

References
Hadano K, Nakamura H, Hamanaka T. Effortful echolalia. *Cortex* 1998; **34**: 67-82
Cross References
Aphasia; Coprolalia; Dynamic aphasia; Imitation behavior; Jargon aphasia; Logorrhea; Palilalia; Transcortical aphasias

Echophenomena
- see IMITATION BEHAVIOR

Echopraxia
Echopraxia is the involuntary, automatic, imitation of an interviewer's movements. This may be observed as a feature of apraxic syndromes, such as corticobasal degeneration, as a complex motor tic in Gilles de la Tourette syndrome, and in frontal lobe disorders (imitation behavior).
Cross References
Copropraxia; Imitation behavior; Tic

Écriture en Double Miroir
- see MIRROR WRITING

Ectropion
- see LID RETRACTION

Eidetic Memory
Photographic, or eidetic, memory is an enhancement of memory to prodigious capacity, beyond hypermnesia. Synesthesia may be linked to eidetic memory, synesthesia being used as a mnemonic aid.
References
Luria AR. *The mind of a mnemonist*. New York: Basic Books, 1968
Cross References
Synesthesia

Eight-and-a-Half Syndrome
The combination of a facial (VII) nerve palsy with a one-and-a-half syndrome due to a pontine lesion has been labeled the eight-and-a-half syndrome. Patients may develop oculopalatal myoclonus months to years after the onset of the ocular motility problem.
References
Eggenberger EJ. Eight-and-a-half syndrome: One-and-a-half syndrome plus cranial nerve VII palsy. *Journal of Neuro-ophthalmology* 1998; **18**: 114-116
Wolin MJ, Trent RG, Lavin PJM, Cornblath WT. Oculopalatal myoclonus after the one-and-a-half syndrome with facial nerve palsy. *Ophthalmology* 1996; **103**: 177-180
Cross References
Facial paresis; Myoclonus; One-and-a-half syndrome; Palatal myoclonus

Emotionalism, Emotional Lability
Emotionalism or emotional lability, or emotional incontinence, implies both frequent and unpredictable changes in emotional expression, for example tearfulness followed shortly by elation, and an inappropriate expression of emotion, for example uncontrollable ("uninhibited" or disinhibited) laughter or crying.

A distinction may be drawn between the occurrence of these phenomena spontaneously or without motivation, or in situations which although funny or sad are not particularly so. Also, a distinction may be made between such phenomena when there is congruence of mood

and affect, sometimes labeled with terms, such as moria or *witzelsucht* (*e.g.*, laughing when feeling happy or elated), and when there is no such congruence (*e.g.*, laughing when not feeling happy or elated), sometimes labeled as pathological, forced, or inappropriate laughter and crying (*q.v.*).

The neurobehavioral state of emotional lability reflects frontal lobe (especially orbitofrontal) lesions, often vascular in origin, and may coexist with disinhibited behavior. It is commoner in vascular dementia than Alzheimer's disease. It may also be seen in delirium and in psychiatric disorders (mania). Pathological laughter and crying may occur as one component of pseudobulbar palsy ("pseudobulbar affect").

References

Heilman KM, Blonder LX, Bowers D, Valenstein E. Emotional disorders associated with neurological diseases. In: Heilman KM, Valenstein E (eds.). *Clinical neuropsychology* (4th edition). Oxford: OUP, 2003: 447-478

Cross References

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

Emposthotonos

Emposthotonos is an abnormal posture consisting of flexion of the head on the trunk and the trunk on the knees, sometimes with flexion of the limbs (*cf*. opisthotonos). Such attacks of "bowing" may be seen in infantile epilepsy syndromes, such as West's syndrome, sometimes called salaam seizures or jack-knife spasms.

Cross References

Opisthotonos; Seizures; Spasm

Encephalopathy

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

As with terms, such as coma and stupor, it is probably better to give a description of the patients clinical state rather than use a term that is open to variable interpretation. Although the term is sometimes reserved for metabolic causes of diffuse brain dysfunction, this usage is not universal. Conditions which may be described as an encephalopathy include:

Metabolic disorders: hypoxia/ischemia, hypoglycemia; organ failure, electrolyte disturbances, hypertension

Drug/toxin ingestion
Brain inflammation/infection (*e.g.*, encephalitis)
Miscellaneous conditions, *e.g.*, Alzheimer's disease, Creutzfeldt-Jakob disease.

Cross References
Asterixis; Coma; Delirium; Myoclonus; Stupor; Tremor

En Garde Position
- see FENCER'S POSTURE; FENCING POSTURE

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

Entomopia
Entomopia (literally "insect eye") is the name given to a grid-like pattern of multiple copies of the same visual image; hence, this is a type of polyopia. This phenomenon has been reported in migraine; its pathogenesis is uncertain.
References
Lopez JR, Adornato BT, Hoyt WF. "Entomopia": a remarkable case of cerebral polyopia. *Neurology* 1993; **43**: 2145-2146
Cross References
Polyopia

Environmental Dependency Syndrome
- see IMITATION BEHAVIOR; UTILIZATION BEHAVIOR

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

Lateral medullary syndrome of Wallenberg
Transient ischemic attacks in basilar artery territory

Demyelinating disease
Head injury
Encephalitis
Following third ventriculostomy for hydrocephalus

Cross References
Lateral medullary syndrome; Vertigo; Vestibulo-ocular reflexes

Epiphora

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

Cross References
Bell's palsy; Crocodile tears

Epley Maneuver

- see HALLPIKE MANEUVER, HALLPIKE TEST; VERTIGO

Erythropsia

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

Cross References
Illusion; "Monochromatopsia"; Phantom chromatopsia

Esophoria

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

Esophoria may occur in individuals with hyperopia (long-sightedness).

Cross References
Cover tests; Exophoria; Heterophoria

Esotropia

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

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

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

Cross References

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

Ewart Phenomenon

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

Cross References

Ptosis; Synkinesia, Synkinesis

Exophoria

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

Cross References

Cover tests; Esophoria; Heterophoria

Exophthalmos

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

Cross References

Lid retraction; Proptosis

Exotropia

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

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

Cross References

Cover tests; Esotropia; Heterotropia

Extensor Posturing

- see DECEREBRATE RIGIDITY

External Malleolar Sign

- see CHADDOCK'S SIGN

External Ophthalmoplegia

- see OPHTHALMOPARESIS, OPHTHALMOPLEGIA

Extinction

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

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

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

The presence of extinction is one of the behavioral manifestations of neglect, and most usually follows nondominant (right) hemisphere lesions.

There is evidence for physiological interhemispheric rivalry or competition in detecting stimuli from both hemifields, which may account for the emergence of extinction following brain injury.

References

Fink GR, Driver J, Rorden C, Baldeweg T, Dolan RJ. Neural consequences of competing stimuli in both visual hemifields: a physiological basis for visual extinction. *Annals of Neurology* 2000; **47**: 440-446

Cross References

Akinesia; Hemiakinesia; Neglect; Visual extinction

Extrapyramidal Signs

- see PARKINSONISM

Eyelid Apraxia

Eyelid apraxia is an inability to open the eyelids at will, although they may open spontaneously at other times (*i.e.*, voluntary-automatic dissociation). Eyelids may be opened manually or by a backward head thrust.

The term has been criticized on the grounds that this may not always be a true "apraxia," in which case the term "levator inhibition" may be preferred since the open eyelid position is normally maintained by tonic activity of the levator palpebrae superioris. Clinically there is no visible contraction of orbicularis oculi, which distinguishes eyelid apraxia from blepharospasm (however, perhaps paradoxically, the majority of cases of eyelid apraxia occur in association with blepharospasm). Electrophysiological studies do in fact show abnormal muscle contraction in the pre-tarsal portion of orbicularis oculi, which

has prompted the suggestion that "focal eyelid dystonia" may be a more appropriate term.

Although the phenomenon may occur in isolation, associations have been reported with:

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

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

References

Boghen D. Apraxia of eyelid opening: a review. *Neurology* 1997; **48**: 1491-1494

Cross References

Apraxia; Blepharospasm; Dystonia

F

"Face-Hand Test"
- see "Arm Drop"

Facial Paresis
Facial paresis, or prosopoplegia, may result from:

- central (upper motor neurone) lesions
- peripheral (lower motor neurone; facial (VII) nerve) lesions
- neuromuscular junction transmission disorders
- primary disease of muscle (*i.e.*, myogenic)

Facial paresis is clinically heterogeneous which may be helpful with lesion localization.

- *Upper motor neurone facial weakness* ("central facial palsy"):
  The ability to raise the eyebrow is preserved due to bilateral supranuclear connections to the frontalis muscle. A dissociation between volitional and emotional facial movements may also occur. Emotional facial palsy refers to the absence of emotional facial movement but with preserved volitional movements, as may be seen with frontal lobe (especially non-dominant hemisphere) precentral lesions (as in abulia, Fisher's sign) and in medial temporal lobe epilepsy with contralateral mesial temporal sclerosis. Volitional paresis without emotional paresis may occur when corticobulbar fibers are interrupted (precentral gyrus, internal capsule, cerebral peduncle, upper pons).

  Causes of upper motor neurone facial paresis include:

  Unilateral:

  Hemisphere infarct (with hemiparesis)
  Lacunar infarct (facio-brachial weakness, +/– dysphasia)
  Space occupying lesions: intrinsic tumor, metastasis, abscess

  Bilateral:

  Motor neurone disease
  Diffuse cerebrovascular disease
  Pontine infarct (locked-in syndrome)

- *Lower motor neurone facial weakness* (peripheral origin):

If this is due to facial (VII) nerve palsy, it results in ipsilateral weakness of frontalis (*cf.* upper motor neurone facial paresis), orbicularis oculi, buccinator, orbicularis oris and platysma. Clinically this produces:

Drooping of the side of the face with loss of the nasolabial fold

Widening of the palpebral fissure with failure of lid closure (lagophthalmos)
Eversion of the lower lid (ectropion) with excessive tearing (epiphora)
Inability to raise the eyebrow, close the eye, frown, blow out the cheek, show the teeth, laugh, and whistle
+/– dribbling of saliva from the paretic side of the mouth
Depression of the corneal reflex (efferent limb of reflex arc affected)
Speech alterations: softening of labials (p, b).

Depending on the precise location of the facial nerve injury, there may also be paralysis of the stapedius muscle in the middle ear, causing sounds to seem abnormally loud (especially low tones: hyperacusis), and impairment of taste sensation on the anterior two-thirds of the tongue if the chorda tympani is affected (ageusia, hypogeusia). Lesions within the facial canal distal to the meatal segment cause both hyperacusis and ageusia; lesions in the facial canal between the nerve to stapedius and the chorda tympani cause ageusia but no hyperacusis; lesions distal to the chorda tympani cause neither ageusia nor hyperacusis (*i.e.*, facial motor paralysis only). Lesions of the cerebellopontine angle cause ipsilateral hearing impairment and corneal reflex depression (afferent limb of reflex arc affected) in addition to facial weakness. There is also a sensory branch to the posterior wall of the external auditory canal which may be affected resulting in local hypoesthesia (Hitselberg sign).

Causes of lower motor neurone facial paresis include:

Bell's palsy: idiopathic lower motor neurone facial weakness, assumed to result from a viral neuritis
Herpes zoster (Ramsey Hunt syndrome);
Diabetes mellitus
Lyme disease (borreliosis, Bannwarth's disease)
Sarcoidosis
Leukemic infiltration, lymphoma
HIV seroconversion
Neoplastic compression (*e.g.*, cerebellopontine angle tumor; rare)
Facial nerve neuroma.

These latter conditions may need to be differentiated from Bell's palsy.

Causes of recurrent facial paresis of lower motor neurone type include:

Diabetes mellitus
Lyme disease (borreliosis, Bannwarth's disease)
Sarcoidosis
Leukemia, lymphoma.

In myasthenia gravis, a disorder of neuromuscular transmission at the neuromuscular junction, there may be concurrent ptosis, diplopia, bulbar palsy and limb weakness, and evidence of fatigable weakness.

Myogenic facial paresis may be seen in facioscapulohumeral (FSH) dystrophy, myotonic dystrophy, mitochondrial disorders. In primary

disorders of muscle the pattern of weakness and family history may suggest the diagnosis.

References

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Hopf HC, Muller-Forell W, Hopf NJ. Localization of emotional and volitional facial paresis. *Neurology* 1992; **42**: 1918-1923

Jacob A, Cherian PJ, Radhakrishnan K, Sankara SP. Emotional facial paresis in temporal lobe epilepsy: its prevalence and lateralizing value. *Seizure* 2003; **12**: 60-64

Cross References

Abulia; Ageusia; Bell's palsy; Bell's phenomenon, Bell's sign; *Bouche de tapir*; Cerebellopontine angle syndrome; Corneal reflex; Eight-and-a-half syndrome; Epiphora; Fisher's sign; Hitselberg sign; Hyperacusis; Lagophthalmos; Locked-in syndrome; Lower motor neurone (LMN) syndrome; Pseudobulbar palsy; Upper motor neurone (UMN) syndrome

Facilitation

Facilitation is an increase in muscle strength following repeated contraction. Clinically, facilitation may be demonstrated by the appearance of tendon-reflexes after prolonged (*ca*. 30 seconds) forced maximal contractions against resistance, *e.g*., the biceps jerk after elbow flexion, knee jerk after knee extension; and by Lambert's sign (increased force grip with sustained contraction).

This phenomenon of post-tetanic potentiation is most commonly seen in the Lambert-Eaton myasthenic syndrome (LEMS), a disorder of neuromuscular junction transmission associated with the presence of autoantibodies directed against presynaptic voltage-gated calcium ion (Ca^{2+}) channels (VGCC). The mechanism is thought to be related to an increased build up of Ca^{2+} ions within the presynaptic terminal with the repetitive firing of axonal action potentials, partially overcoming the VGCC antibody-mediated ion channel blockade, and leading to release of increasing quanta of acetylcholine.

Cross References

Fatigue; Lambert's sign

"False-Localizing Signs"

Neurological signs may be described as "false-localizing" when their appearance reflects pathology distant from the expected anatomical locus. The classic example, and probably the most frequently observed, is abducens nerve palsy (unilateral or bilateral) in the context of raised intracranial pressure, presumed to result from stretching of the nerve over the ridge of the petrous temporal bone. Many false-localizing signs occur in the clinical context of raised intracranial pressure, either idiopathic (idiopathic intracranial hypertension [IIH]) or symptomatic (secondary to tumor, hematoma, abscess).

A brief topographical overview of false-localizing signs (more details may be found in specific entries) includes:

- Motor system:
  - Kernohan's notch syndrome: false-localizing hemiparesis
  - Cerebellar syndrome with anterior cerebral artery territory infarction damaging frontocerebellar pathways
  - Brainstem compression causing diaphragm paralysis
- Cranial nerves:
  - Proptosis with middle cranial fossa tumor
  - Oculomotor (III) nerve palsy with contralateral supratentorial lesion
  - Divisional oculomotor nerve palsy with brainstem or subarachnoid space pathology
  - Trochlear nerve palsy with IIH
  - Trigeminal nerve palsy with IIH
  - Abducens nerve palsy with IIH
  - Facial nerve palsy with IIH
  - Vestibulocochlear nerve dysfunction with IIH
- Spinal cord and roots:
  - Foramen magnum/upper cervical cord lesion causing hand muscle wasting ("remote atrophy")
  - Lower cervical/upper thoracic myelopathy producing mid-thoracic girdle sensation
  - Urinary retention with rostral spinal cord compression
  - Radiculopathy with IIH, may even mimic Guillain-Barré syndrome

References

Larner AJ. False localizing signs. *Journal of Neurology, Neurosurgery and Psychiatry* 2003; **74**: 415-418

Larner AJ. A topographical anatomy of false-localizing signs. *Advances in Clinical Neuroscience & Rehabilitation* 2005; **5(1)**: 20-21

Cross References

Abducens (vi) Nerve palsy; Divisional palsy; Girdle sensation; Kernohan's notch syndrome; Oculomotor (III) nerve palsy; Proptosis; Urinary retention

Fan Sign *(Signe de l'éventail)*

- see BABINSKI'S SIGN (1)

Fasciculation

Fasciculations are rapid, flickering, twitching, involuntary movements within a muscle belly resulting from spontaneous activation of a bundle, or fasciculus, of muscle fibers (*i.e.*, a motor unit), insufficient to move the joint. Fasciculations may also be induced by lightly tapping over a partially denervated muscle belly. The term was formerly used synonymously with fibrillation, but the latter term is now reserved for contraction of a single muscle fibre, or a group of fibers smaller than a motor unit.

Brief and localized fasciculations can be a normal finding (*e.g.*, in the intrinsic foot muscles, especially abductor hallucis, and gastrocnemius, but not tibialis anterior), particularly if unaccompanied by other neurological symptoms and signs (wasting, weakness, sensory disturbance, sphincter dysfunction). Persistent fasciculations most usually reflect a pathological process involving the lower motor neurones in the anterior (ventral) horn of the spinal cord and/or in brainstem motor nuclei, typically motor neurone disease (in which cramps are an early associated symptom). Facial and perioral fasciculations are highly characteristic of Kennedy's disease (X-linked bulbospinal neuronopathy). However, fasciculations are not pathognomonic of lower motor neurone pathology since they can on rare occasions be seen with upper motor neurone pathology.

The pathophysiological mechanism of fasciculations is thought to be spontaneous discharge from motor nerves, but the site of origin of this discharge is uncertain. Although ectopic neural discharge from anywhere along the lower motor neurone from cell body to nerve terminal could produce fasciculation, the commonly encountered assumption that it originates in the anterior horn cell body is not supported by the available evidence, which points to a more distal origin in the intramuscular nerve terminals. In addition, denervation of muscle fibers may lead to nerve fibre sprouting (axonal and collateral) and enlargement of motor units which makes fasciculations more obvious clinically.

Fasciculations may be seen in:

- Motor neurone disease with lower motor neurone involvement (*i.e.*, progressive muscular atrophy, progressive bulbar atrophy variants)
- Spinal muscular atrophy
- Cervical radiculopathy (restricted to myotomal distribution)
- Multifocal motor neuropathy with conduction block
- Benign fasciculation syndrome: typically seen only after exercise and without associated muscle atrophy or weakness
- Cramp fasciculation syndrome
- Kennedy's disease (X-linked bulbospinal neuronopathy; especially perioral)
- Almost any lower motor neurone disease, especially compression
- Metabolic causes: thyrotoxicosis, tetany, after acetylcholinesterase inhibitors, anesthetic muscle relaxants.

Fasciculations may need to be distinguish from myokymia or neuromyotonia.

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Desai J, Swash M. Fasciculations: what do we know of their significance? *Journal of the Neurological Sciences* 1997; **152 (suppl1)**: S43-S48

Layzer RB. The origin of muscle fasciculations and cramps. *Muscle Nerve* 1994; **17**: 1243-1249

Cross References
Calf hypertrophy; Cramp; Fibrillation; Lower motor neurone (LMN) syndrome; Myokymia; Neuromyotonia

Fast Micrographia

In "fast" micrographia, written letters are microscopic from the outset, sometimes approximating to a straight line, though produced at normal speed without fatigue. This pattern has been observed in progressive supranuclear palsy and with globus pallidus lesions, and contrasts with the "slow" micrographia, writing becoming progressively slower and smaller, seen in idiopathic Parkinson's disease.

References
Quinn NP. Fast micrographia and pallidal pathology. *Journal of Neurology, Neurosurgery and Psychiatry* 2002; **72**: 135 (abstract)

Cross References
Micrographia

Fatigue

The term fatigue may be used in different contexts to refer to both a sign and a symptom.

The sign of fatigue, also known as peripheral fatigue, consists of a reduction in muscle strength with repeated muscular contraction. This most characteristically occurs in disorders of neuromuscular junction transmission (*e.g.*, myasthenia gravis), but it may also be observed in disorders of muscle (*e.g.*, myopathy, polymyositis) and neurogenic atrophy (*e.g.*, motor neurone disease). In myasthenia gravis, fatigue may be elicited in the extraocular muscles by prolonged upgaze causing eyelid drooping; in bulbar muscles by prolonged counting or speech causing hypophonia; and in limb muscles by repeated contraction, especially of proximal muscles (*e.g.*, shoulder abduction) leading to weakness in previously strong muscles. Fatigue in myasthenia gravis is understood as a decline in the amount of acetylcholine released from motor nerve terminals with successive neural impulses, along with a reduced number of functional acetylcholine receptors (AChR) at the motor end-plates, due to binding of AChR antibodies and/or complement mediated destruction of the postsynaptic folds.

(A gradual decline in the amplitude and speed of initiation of voluntary movements, hypometria and hypokinesia, as seen in disorders of the basal ganglia, especially Parkinson's disease, may also be described as fatigue, *e.g.*, "slow" micrographia may be ascribed to "fatigue." Progressive supranuclear palsy is notable for lack of fatigue.)

Fatigue as a symptom, or central fatigue, is an enhanced perception of effort and limited endurance in sustained physical and mental activities. This may occur in multiple sclerosis (MS), post-polio syndrome, post-stroke syndromes, and chronic fatigue syndrome (CFS). In MS and CFS, fatigue may be a prominent and disabling complaint even though neurological examination reveals little or no clinical deficit. This type of fatigue is ill-understood: in MS, frequency-

dependent conduction block in demyelinated axons has been suggested, as has hypothalamic pathology. Current treatment is symptomatic (amantadine, modafinil, 3,4-diaminopyridine) and rehabilitative (graded exercise).

Fatigue may be evaluated with various instruments, such as the Krupp Fatigue Severity Score.

References

Chaudhuri A, Behan PO. Fatigue in neurological disorders. *Lancet* 2004; **363**: 978-988

Zifko UA. Management of fatigue in patients with multiple sclerosis. *Drugs* 2004; **64**: 1295-1304

Cross References

Dystonia; Hypokinesia; Hypometria; Micrographia; Weakness

Femoral Stretch Test

The femoral stretch test, or reverse straight leg raising, consists of extension of the hip with the knee straight with the patient lying prone, a maneuver which puts traction on the femoral nerve or L3 root and may exacerbate pain in a femoral neuropathy or L3 radiculopathy, perhaps due to a retroperitoneal hemorrhage.

Cross References

Lasègue's sign

Fencer's Posture, Fencing Posture

Epileptic seizures arising in or involving the supplementary motor area may lead to adversial head and eye deviation, abduction and external rotation of the contralateral arm, flexion at the elbows, and posturing of the legs, with maintained consciousness, a phenomenon christened by Penfield the "fencing posture" because of its resemblance to the *en garde* position. These may also be known as "salutatory seizures."

Cross References

Seizures

Festinant Gait, Festination

Festinant gait or festination is a gait disorder characterized by rapid short steps (Latin: *festinare*, to hurry, hasten, accelerate) due to inadequate maintenance of the body's centre of gravity over the legs. To avoid falling and to maintain balance the patient must "chase" the centre of gravity, leading to an increasing speed of gait and a tendency to fall forward when walking (propulsion). A similar phenomenon may be observed if the patient is pulled backward (retropulsion). Festination may be associated with freezing of gait.

Festination is common in idiopathic Parkinson's disease; it is associated with longer duration of disease and higher Hoehn & Yahr stage. Festination may be related to the flexed posture and impaired postural reflexes commonly seen in these patients. It is less common in symptomatic causes of parkinsonism, but has been reported, for example in aqueduct stenosis.

References

Leheta O, Boschert J, Krauss JK, Whittle IR. Festination as the leading symptom of late onset idiopathic aqueduct stenosis. *Journal of Neurology, Neurosurgery and Psychiatry* 2002; **73**: 599-600

Cross References

Freezing; Parkinsonism; Postural reflexes

Fibrillation

Fibrillation was previously synonymous with fasciculation, but the term is now reserved for the spontaneous contraction of a single muscle fibre, or a group of fibers smaller than a motor unit, hence this is more appropriately regarded as an electrophysiological sign without clinical correlate.

Cross References

Fasciculation

Finger Agnosia

Finger agnosia is a type of tactile agnosia, in which there is inability to identify which finger has been touched when the eyes are closed, despite knowing that a finger has been touched; or inability to point to or move a finger when it is named; or inability to name the fingers (patient's own fingers or those of another person). This is a disorder of body schema, and may be regarded as a partial form of autotopagnosia.

Finger agnosia is most commonly observed with lesions of the dominant parietal lobe. It may occur in association with acalculia, agraphia, and right-left disorientation, with or without alexia and difficulty spelling words, hence as one feature of Gerstmann syndrome. Isolated cases of finger agnosia in association with left corticosubcortical posterior parietal infarction have been reported. Since this causes no functional deficit, it may be commoner than reported.

References

Della Sala S, Spinnler H. Finger agnosia: fiction or reality? *Archives of Neurology* 1994; **51**: 448-450

Cross References

Agnosia; Autotopagnosia; Gerstmann syndrome

Finger Drop

- see WRIST DROP

Finger-Floor Distance

In patients with leg (+/– back) pain suspected of having lumbosacral nerve root compression, a finger-floor distance of > 25 cm when the patient bends forward and attempts to touch the floor with the fingers has been found an independent predictor of radiological (MR imaging) compression. This was not the case for the straight leg raising test.

References

Vroomen PCAJ, de Krom MCTFM, Wilmink JT, Kester ADM, Knottnerus JA. Diagnostic value of history and physical examination in patients suspected of lumbosacral nerve root compression. *Journal of Neurology, Neurosurgery and Psychiatry* 2002; **72**: 630-634

Cross References

Lasègue's sign

"Finger-Nose Test"

- see ATAXIA; CEREBELLAR SYNDROMES

Fisher's Sign

Fisher's sign is the paucity of facial expression conveying emotional states or attitudes (emotional facial paresis). It follows nondominant (right) hemisphere lesions and may accompany emotional dysprosody of speech.

Cross References

Abulia; Aprosodia, aprosody; Facial paresis

Flaccidity

Flaccidity is a floppiness which implies a loss of normal muscular tone (hypotonia). This may occur transiently after acute lesions of the corticospinal tracts (flaccid paraparesis), before the development of spasticity, or as a result of lower motor neurone syndromes. It is difficult to separate the change in tone from weakness.

Cross References

Hypotonia, Hypotonus; Lower motor neurone (LMN) syndrome

Flail Arm

Flail arm refers to a severe and symmetric wasting and weakness of the arms without significant functional involvement of other regions, seen in one variant of motor neurone disease, the "flail arm syndrome," also known as Vulpian-Bernhart's form. Men are reported to be much more frequently affected than women, and this group may show improved survival compared to other MND patients. Alternative designations for this syndrome include amyotrophic brachial diplegia, dangling arm syndrome, and neurogenic man-in-a-barrel syndrome.

References

Hu MTM, Ellis CM, Al-Chalabi A, Leigh PN, Shaw CE. Flail arm syndrome: a distinctive variant of amyotrophic lateral sclerosis. *Journal of Neurology, Neurosurgery and Psychiatry* 1998; **65**: 950-951

Cross References

Amyotrophy; "man-in-a-barrel"

Flail Foot

- see CAUDA EQUINA SYNDROME; FOOT DROP

Flap, Flapping Tremor

- see ASTERIXIS

Flexibilitas Cerea (Waxy Flexibility)
- see CATATONIA

Flexion-Adduction Sign
Neuralgic amyotrophy (Parsonage-Turner syndrome) may cause arm pain, which may be prevented by holding the arm flexed at the elbow and adducted at the shoulder.

- Waxman SG. The flexion-adduction sign in neuralgic amyotrophy. *Neurology* 1979; **29**: 1301-1304

Flexor Posturing
- see DECORTICATE RIGIDITY

Flick Sign
A flicking, shaking movement of the hands made by patients with carpal tunnel syndrome to try to relieve the paresthesia and pain caused by the condition, typically noted on waking at night.

Cross References

Phalen's sign; Tinel's sign

Floccillation
- see CARPHOLOGIA

Flycatcher Tongue
- see TROMBONE TONGUE

Flynn Phenomenon
Flynn phenomenon is paradoxical constriction of the pupils in darkness. This has been documented in various conditions including congenital achromatopsia, following optic neuritis, and in autosomal dominant optic atrophy.

References

Frank JW, Kushner BJ, France TD. Paradoxical pupillary phenomena: a review of patients with pupillary constriction to darkness. *Archives of Ophthalmology* 1988; **106**: 1564-1566

Cross References

Pupillary reflexes

Foot Drop
Foot drop, often manifest as the foot dragging during the swing phase of the gait, causing tripping and/or falls, may be due to upper or lower motor neurone lesions, which may be distinguished clinically.

- *Stiff foot drop*, with upper motor neurone lesions:
  leads to a circumducting gait; it may be possible to see or hear the foot dragging or scuffing along the floor, and this may cause excessive wear on the point of the shoe. There will be other upper motor neurone signs (hemiparesis; spasticity, clonus, hyperreflexia, Babinski's sign).
- *Floppy foot drop*, with lower motor neurone lesions:

leads to a stepping gait (steppage) to try to lift the foot clear of the floor, and a slapping sound on planting the foot. At worst, there is a flail foot in which both the dorsiflexors and the plantar flexors of the foot are weak (*e.g.*, in high sciatic nerve or sacral plexus lesions). Other lower motor neurone signs may be present (hypotonia, areflexia or hyporeflexia).

Causes of floppy foot drop include:

Common peroneal nerve palsy
Sciatic neuropathy
Lumbosacral plexopathy
L4/L5 radiculopathy
Motor or sensorimotor polyneuropathy (*e.g.*, hereditary motor and sensory neuropathy)
Motor neuronopathy (anterior horn cell disease)
Mononeuropathy multiplex

These may be distinguished on clinical and/or neurophysiological grounds

References

McNamara B. Foot drop. *Advances in Clinical Neuroscience & Rehabilitation* 2003; **3(1)**: 24-25

Cross References

Cauda equina syndrome; Hemiparesis; Lower motor neurone (LMN) syndrome; Steppage, stepping gait; Upper motor neurone (UMN) syndrome

Foot Grasping

- see GRASP REFLEX

Forced Ductions

Forced ductions, performed by grasping the anesthetized sclera with forceps and then moving the eye through its range of motions, may be used to determine whether restricted eye movement is mechanical, due to a lesion within the orbit, such as thyroid ophthalmopathy or superior oblique tendon sheath (Brown's) syndrome.

Forced Grasping

- see GRASP REFLEX

Forced Groping

Forced groping describes involuntary movements of a hand, as if searching for an object or item which has touched or brushed against it; the hand may follow the object around if it moves (magnetic movements). There may be an accompanying grasp reflex. This type of behavior may be displayed by an alien hand, most usually in the context of corticobasal degeneration. Forced groping may be conceptualized as an exploratory reflex which is "released" from frontal lobe control by a pathological process, as in utilization behavior.

References

Adie WS, Critchley M. Forced grasping and groping. *Brain* 1927; **50**: 142-170

Cross References

Alien hand, alien limb; Grasp reflex; Magnetic movements; Utilization behavior

Forced Laughter and Crying

- see EMOTIONALISM, EMOTIONAL LIABILITY; PATHOLOGICAL CRYING, PATHOLOGICAL LAUGHTER

Forced Upgaze

Tonic upward gaze deviation, forced upgaze, may be seen in coma after diffuse hypoxic-ischemic brain injury with relative sparing of the brainstem. Forced upgaze may also be psychogenic, in which case it is overcome by cold caloric stimulation of the ear drums. Forced upgaze must be differentiated from oculogyric crisis.

Cross References

Oculogyric crisis

Foreign Accent Syndrome

- see APHEMIA

Formication

- see PARESTHESIA; TINEL'S SIGN

Fortification Spectra

Fortification spectra, also known as teichopsia, are visual hallucinations which occur as an aura, either in isolation (migraine without headache) or prior to an attack of migraine (migraine with aura; "classical migraine"). The appearance is a radial array likened to the design of medieval castles, not simply of battlements. Hence these are more complex visual phenomena than simple flashes of light (photopsia) or scintillations. They are thought to result from spreading depression, of possible ischemic origin, in the occipital cortex.

Cross References

Aura; Hallucination; Photopsia

Foster Kennedy Syndrome

The Foster Kennedy syndrome consists of optic atrophy in one eye with optic disc edema in the other eye, due to a tumor compressing one optic nerve (to produce atrophy) and causing raised intracranial pressure (to produce contralateral papilledema). A pseudo-Foster Kennedy syndrome is described in consecutive acute ischemic optic neuropathy.

References

Kennedy F. Retrobulbar neuritis as an exact diagnostic sign of certain tumors and abscesses in the frontal lobe. *American Journal of Medical Science* 1911; **142**: 355-368.

Cross References
Optic atrophy; Papilledema

Freezing

Freezing is the sudden inability in a patient with parkinsonism to move or to walk, *i.e.*, gait failure, as though the patient were turned to ice or the feet were nailed to the floor. This is one of the unpredictable motor fluctuations in late Parkinson's disease (associated with longer duration of disease and treatment) which may lead to falls, usually forward onto the knees, and injury. It may occur in confined spaces (*e.g.*, doorways), when trying to turn, or when trying to do two things at once. It is not seen in the early years of levodopa therapy.

Two variants are encountered, occurring either during an off period or wearing off period, or randomly, *i.e.*, unrelated to drug dosage or timing.

Treatment strategies include use of dopaminergic agents and, anecdotally, L-threodops, but these agents are not reliably helpful, particularly in random freezing. Use of visual targets (real or imagined) may help, *e.g.*, stepping over a line.

Freezing may also occur in multiple system atrophy, and has also been reported as an isolated phenomenon.

Cross References
Parkinsonism

Fregoli Syndrome

- see DELUSION

Froment's Sign

Froment has two eponymous signs:

- Activated rigidity or synkinesis (*q.v.*).
- In an ulnar nerve lesion, flexion of the distal phalanx of the thumb (flexor pollicis longus, innervated by the median nerve) is seen when attempting to squeeze a sheet of paper between the thumb and the index finger, as a compensation for the weakness of thumb adduction (adductor pollicis, innervated by the ulnar nerve), also known as Froment's prehensile thumb sign or the *signe du journal*. The term is also sometimes used for weakness of little finger adduction, evident when trying to grip a piece of paper between the ring and little finger.

Cross References
Rigidity; Synkinesia, synkinesis

Frontal Ataxia

- see ATAXIA

Frontal Lobe Syndromes

The frontal lobes of the brain have enlarged greatly during phylogeny; their diverse connections with the basal ganglia, basal forebrain, and

cerebellum, as well as other cortical areas, reflect their multiple motor and behavioral functions. Damage to the frontal lobes may produce a variety of clinical signs, most frequently changes in behavior. Such changes may easily be overlooked with the traditional neurological examination, although complained of by patient's relatives, and hence specific bedside tests of frontal lobe function should be utilized, for example:

- Verbal fluency: *e.g.*, letter/phonemic (F, A, S) probably a more specific test than category/semantic (animals, foods).
- Proverb interpretation: *e.g.*, "Make hay while the sun shines"; "Too many cooks spoil the broth"; interpretation tends to be concrete in frontal lobe disorders.
- Cognitive estimates: *e.g.*, height of the Post Office Tower, length of a man's spine, distance from London to Edinburgh; may be grossly abnormal or inappropriate.
- Copying motor sequences, to assess motor programming ability: *e.g.*, Luria fist-edge-palm test (three step motor sequence with hand).
- Alternating sequence tests: *e.g.*, alternating finger flexion/extension out of phase in two hands, or repeatedly writing m n m n m n (also used as tests of praxis, which may be affected with frontal lobe pathology); swapping a coin from hand to hand behind back in a predictable pattern and asking the patient which hand the coin is in.
- Set-shifting or go/no go tests, in which an alternating pattern is suddenly changed, *e.g.*, changing the previously predictable (left/right) pattern of coin hidden in clenched hand swapped over behind back; rhythmic tapping with pen on a surface (I tap once, you tap twice; I tap twice, you tap once); tests of response inhibition (ask patient to clap three times, s/he does so multiple times).

A useful clinico-anatomical classification of frontal lobe syndromes which reflects the functional subdivisions of the frontal lobes is as follows:

- *Orbitofrontal Syndrome* ("disinhibited"):
  Disinhibited behavior (including sexual disinhibition), impulsivity
  Inappropriate affect, *witzelsucht*, euphoria
  Emotional lability (moria)
  Lack of judgment, insight
  Distractibility, lack of sustained attention; hypermetamorphosis
  Motor perseverations are not a striking feature
- *Frontal Convexity Syndrome* ("apathetic"):
  Apathy; abulia, indifference
  Motor perseveration
  Difficulty set-shifting, stimulus boundedness
  Reduced verbal fluency
  Deficient motor programming, *e.g.*, three step hand sequence, rhythmical tapping (go/no-go test)
- *Medial Frontal Syndrome* ("akinetic"):
  Little spontaneous movement, bradykinesia, hypokinesia
  Sparse verbal output (akinetic mutism)

Urinary incontinence
Sensorimotor signs in lower limbs
Indifference to pain

Overlap between these regional syndromes may occur.

A "*dysexecutive syndrome*" has also been defined, consisting of difficulty planning, adapting to changing environmental demands (impaired cognitive flexibility, *e.g.*, in set-shifting tests), and directing attentional resources. This may be seen with dorsolateral (prefrontal) damage.

These frontal lobe syndromes may be accompanied by various neurological signs (frontal release signs or primitive reflexes). Other phenomena associated with frontal lobe pathology include imitation behaviors (echophenomena) and, less frequently, utilization behavior, features of the environmental dependency syndrome.

References

Larner AJ, Leach JP. Phineas Gage and the beginnings of neuropsychology. *Advances in Clinical Neuroscience & Rehabilitation* 2002; **2(3)**: 26

Parkin AJ. *Explorations in cognitive neuropsychology*. Hove: Psychology Press, 1996: 220-242

Trimble MR. *Biological psychiatry* (2nd edition). Chichester: Wiley, 1996: 147-156

Cross References

Abulia; Akinesia; Akinetic mutism; Apathy; Attention; Disinhibition; Dysexecutive syndrome; Emotionalism, Emotional lability; Frontal release signs; Hypermetamorphosis; Hyperorality; Hyperphagia; Hypersexuality; Incontinence; Perseveration; Utilization behavior; *Witzelsucht*

Frontal Release Signs

Frontal release signs are so named because of the belief that they are released from frontal inhibition by diffuse pathology within the frontal lobes (usually vascular or degenerative) with which they are often associated, although they may be a feature of normal ageing. Some of these responses are present during infancy but disappear during childhood, hence the terms "primitive reflexes" or "developmental signs" are also used (Babinski's sign may therefore fall into this category). The term "psychomotor signs" has also been used since there is often accompanying change in mental status.

The frontal release signs may be categorized as:

- *Prehensile*:
  - Sucking reflex (tactile, visual)
  - Grasp reflex: hand, foot
  - Rooting reflex (turning of the head toward a tactile stimulus on the face)
- *Nociceptive*:
  - Snout reflex
  - Pout reflex
  - Glabellar (blink) reflex
  - Palmomental reflex

The corneomandibular and nuchocephalic reflexes may also be categorized as "frontal release" signs. Some are of little clinical value

(*e.g.*, palmomental reflex). Concurrent clinical findings may include dementia, gait disorder (frontal gait, *marche à petit pas*), urinary incontinence, akinetic mutism and *gegenhalten*.

Common causes of these findings are diffuse cerebrovascular disease and motor neurone disease.

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

Cross References

Age-related signs; Babinski's sign (1); Corneomandibular reflex; *Gegenhalten*; Grasp reflex; *Marche à petit pas;* Palmomental reflex; Pout reflex; Rooting reflex; Sucking reflex

Fugue

Fugue, and fugue-like state, are used to refer to a syndrome characterized by loss of personal memory (hence the alternative name of "twilight state"), automatic and sometimes repetitive behaviors, and wandering or driving away from normal surroundings.

Fugue may be:

Psychogenic: associated with depression (sometimes with suicide); alcoholism, amnesia; "hysteria";

Epileptic: complex partial seizures

Narcoleptic

Some patients with frontotemporal dementia may spend the day walking long distances, and may be found a long way from home, unable to give an account of themselves, and aggressive if challenged; generally they are able to find their way home (spared topographical memory) despite their other cognitive deficits.

Cross References

Amnesia; Automatism; Dementia; Poriomania; Seizures

Functional Weakness and Sensory Disturbance

Various signs have been deemed useful indicators of functional or "nonorganic" neurological illness, including:

Collapsing or "give way" weakness
Hoover's sign
Babinski's trunk-thigh test
"Arm drop"
Belle indifférence
Sternocleidomastoid sign
Midline splitting sensory loss
Functional postures, gaits:
- Monoplegic "dragging"
- Fluctuation of impairment
- Excessive slowness, hesitation
- "Psychogenic Romberg" sign
- "Walking on ice"

Uneconomic posture, waste of muscle energy
Sudden knee buckling

Although such signs may be suggestive, their diagnostic utility has never been formally investigated in prospective studies, and many, if not all, have been reported with "organic" illness. Hence it is unwise to rely on them as diagnostic indicators.

References

Lempert T, Brandt T, Dieterich M *et al*. How to identify psychogenic disorders of stance and gait: a video study in 37 patients. *Journal of Neurology* 1991; **238**: 140-146

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

Cross References

"Arm drop"; Babinski's trunk-thigh test; *Belle indifférence*; Collapsing weakness; Hoover's sign; Sternocleidomastoid test

Funnel Vision

- see "TUNNEL VISION"

G

Gag Reflex

The gag reflex is elicited by touching the posterior pharyngeal wall, tonsillar area, or the base of the tongue, with the tip of a thin wooden ("orange") stick. Depressing the tongue with a wooden spatula, and the use of a torch for illumination of the posterior pharynx, may be required to get a good view. There is a palatal response (palatal reflex), consisting of upward movement of the soft palate with ipsilateral deviation of the uvula; and a pharyngeal response (pharyngeal reflex or gag reflex) consisting of visible contraction of the pharyngeal wall. Lesser responses include medial movement, tensing, or corrugation of the pharyngeal wall. In addition there may be head withdrawal, eye watering, coughing, and retching. Hence there is variability of response in different individuals. Some studies claim the reflex is absent in many normal individuals, especially with increasing age, without evident functional impairment; whereas others find it in all healthy individuals, although variable stimulus intensity is required to elicit it.

The afferent limb of the reflex arc is the glossopharyngeal (IX) nerve, the efferent limb in the glossopharyngeal and vagus (X) nerves. Hence individual or combined lesions of the glossopharyngeal and vagus nerves depress the gag reflex, as in neurogenic bulbar palsy.

Dysphagia is common after a stroke, and the gag reflex is often performed to assess the integrity of swallowing. Some argue that absence of the reflex does not predict aspiration and is of little diagnostic value, since this may be a normal finding in elderly individuals, whereas pharyngeal sensation (feeling the stimulus at the back of the pharynx) is rarely absent in normals and is a better predictor of the absence of aspiration. Others find that even a brisk pharyngeal response in motor neurone disease may be associated with impaired swallowing. Hence the value of the gag reflex remains debatable. A video swallow may be a better technique to assess the integrity of swallowing.

References

Davies AE, Kidd D, Stone SP, MacMahon J. Pharyngeal sensation and gag reflex in healthy subjects. *Lancet* 1995; **345**: 487-488

Hughes TAT, Wiles CM. Palatal and pharyngeal reflexes in health and motor neuron disease. *Journal of Neurology, Neurosurgery and Psychiatry* 1996; **61**: 96-98

Cross References

Bulbar palsy; Dysphagia

Gait Apraxia

Gait apraxia is a name given to an inability to walk despite intact motor systems and sensorium. Patients with gait apraxia are often

hesitant, seemingly unable to lift their feet from the floor ("magnetic gait") or put one foot in front of the other. Arms may be held out at the sides to balance for fear of falling; fear may be so great that the patient sits in a chair gripping its sides. These phenomena may be observed with lesions of the frontal lobe and white matter connections, with or without basal ganglia involvement, for example in diffuse cerebrovascular disease and normal pressure hydrocephalus. A syndrome of isolated gait apraxia has been described with focal degeneration of the medial frontal lobes. In modern classifications of gait disorders, gait apraxia is subsumed into the categories of frontal gait disorder, frontal disequilibrium, and isolated gait ignition failure.

Gait apraxia is an important diagnosis to establish since those afflicted generally respond poorly, if at all, to physiotherapy; moreover, because both patient and therapist often become frustrated because of lack of progress, this form of treatment is often best avoided.

References

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

Rossor MN, Tyrrell PJ, Warrington EK, Thompson PD, Marsden CD, Lantos P. Progressive frontal gait disturbance with atypical Alzheimer's disease and corticobasal degeneration. *Journal of Neurology, Neurosurgery and Psychiatry* 1999; **67**: 345-352

Cross References

Apraxia

Ganglionopathy

- see NEUROPATHY

Ganser Phenomenon

The Ganser phenomenon consists of giving approximate answers to questions which can at times verge on the absurd (Q: "How many legs does a cow have?"; A: "Three"), also known as paralogia or *vorbeireden*. This may occur in psychiatric disease, such as depression, schizophrenia, and malingering, and sometimes in neurological disease (head injury, epilepsy). A Ganser syndrome of hallucinations, conversion disorder, cognitive disorientation and approximate answers is also described but of uncertain nosology.

References

Carney MW. Ganser syndrome and its management. *British Journal of Psychiatry* 1987; **151**: 697-700

Enoch MD, Ball HN. *Uncommon psychiatric syndromes* (4th edition). London: Arnold, 2001: 74-94

Gaping

Gaping, or involuntary opening of the mouth, may occur as a focal dystonia of the motor trigeminal nerve, also known as Brueghel syndrome after that artist's painting *De Gaper* ("Yawning man," *ca*. 1558) which is said to illustrate a typical case. Afflicted individuals may also

demonstrate paroxysmal hyperpnea and upbeating nystagmus, suggesting a brainstem (possibly pontine) localization of pathology. The condition should be distinguished from other cranial dystonias with blepharospasm (Meige syndrome).

References

Gilbert GJ. Brueghel syndrome: its distinction from Meige syndrome. *Neurology* 1996; **46**: 1767-1769

Cross References

Blepharospasm; Dystonia; Nystagmus

Gaze-Evoked Phenomena

A variety of symptoms have been reported to be evoked, on occasion, by alteration of the direction of gaze:

- Amaurosis: lesion, usually intraorbital, compressing central retinal artery
- Laughter
- Nystagmus: usually indicative of cerebellar lesion; may occur as a side-effect of medications; also convergence-retraction nystagmus on upgaze in dorsal midbrain (Parinaud's) syndrome
- Phosphenes: increased mechanosensitivity in demyelinated optic nerve
- Segmental constriction of the pupil (Czarnecki's sign) following aberrant regeneration of the oculomotor (III) nerve to the iris sphincter
- Tinnitus: may develop after resection of cerebellopontine angle tumors, may be due to abnormal interaction between vestibular and cochlear nuclei
- Vertigo

Cross References

Leopold NA. Gaze-induced laughter. *Journal of Neurology, Neurosurgery and Psychiatry* 1977; **40**: 815-817

Gaze Palsy

Gaze palsy is a general term for any impairment or limitation in conjugate (yoked) eye movements. This may be supranuclear, nuclear, or infranuclear in origin. Preservation of the vestibulo-ocular reflexes may help differentiate supranuclear gaze palsies from nuclear/ infranuclear causes.

Cross References

Locked-in syndrome; Supranuclear gaze palsy; Vestibulo-ocular reflexes

Gegenhalten

Gegenhalten, or paratonia, or paratonic rigidity, is a resistance to passive movement of a limb when changing its posture or position, which is evident in both flexor and extensor muscles (as in rigidity, but not spasticity), which seems to increase further with attempts to get the patient to relax, such that there is a resistance to any applied movement

(German: to counter, stand ones ground). However, this is not a form of impaired muscle relaxation akin to myotonia and paramyotonia. For instance, when lifting the legs by placing the hands under the knees, the legs may be held extended at the knees despite encouragement on the part of the examiner for the patient to flex the knees.

Gegenhalten is a sign of bilateral frontal lobe dysfunction, especially mesial cortex and superior convexity (premotor cortex, area 6). It is not uncommon in elderly individuals with diffuse frontal lobe cerebrovascular disease.

Cross References

Frontal release signs; Myotonia; Paramyotonia; Rigidity; Spasticity

Gerstmann Syndrome

The Gerstmann syndrome, or angular gyrus syndrome, consists of acalculia, agraphia (of central type), finger agnosia, and right-left disorientation; there may in addition be alexia and difficulty spelling words but these are not necessary parts of the syndrome. Gerstmann syndrome occurs with lesions of the angular gyrus and supramarginal gyrus in the posterior parietotemporal region of the dominant (usually left) hemisphere, for example infarction in the territory of the middle cerebral artery.

All the signs comprising Gerstmann syndrome do fractionate or dissociate, *i.e.*, they are not causally related, or representative of a unitary neuropsychological function, as was once suggested. Nonetheless the Gerstmann syndrome remains useful for the purposes of clinical localization.

References

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Mayer E, Martory M-D, Pegna AJ *et al*. A pure case of Gerstmann's syndrome with a subangular lesion. *Brain* 1999; **122**: 1107-1120

Pearce JMS. Gerstmann's syndrome. In: Pearce JMS. *Fragments of neurological history*. London: Imperial College Press, 2003: 92-94

Cross References

Acalculia; Agraphia; Alexia; Finger agnosia; Right-left disorientation

Geste Antagoniste

Geste antagoniste is a sensory "trick" which alleviates, and is characteristic of, dystonia. *Geste antagoniste* consists of a tactile or proprioceptive stimulus, which is learned by the patient, which reduces or eliminates the dystonic posture. For example, touching the chin, face or neck may overcome torticollis (cervical dystonia), and singing may inhibit blepharospasm. *Gestes* may also modify tremor. They are almost ubiquitous in sufferers of cervical dystonia and have remarkable efficacy.

The mechanism is unknown: although afferent feedback from the periphery may be relevant, it is also possible that concurrent motor output to generate the trick movement may be the key element, in which case the term "sensory trick" is a misnomer.

References
Filipovic SR, Jahanshahi M, Viswanathan R, Heywood P, Bhatia KP, Rogers D. Clinical features of the geste antagoniste in cervical dystonia. *Journal of Neurology, Neurosurgery and Psychiatry* 2002; **73**: 215 (abstract 10)
Cross References
Dystonia; Torticollis

Gibbus

Angulation of the spine due to vertebral collapse may be due to osteoporosis, metastatic disease, or spinal tuberculosis. There may be associated myelopathy. Camptocormia (bent spine syndrome) enters the differential diagnosis.
Cross References
Camptocormia; Myelopathy

Girdle Sensation

Compressive lower cervical or upper thoracic myelopathy may produce spastic paraparesis with a false-localizing mid-thoracic sensory level or "girdle sensation" (*cf*. cuirasse). The pathophysiology is uncertain, but ischemia of the thoracic watershed zone of the anterior spinal artery from compression at the cervical level has been suggested.
References
Ochiai H, Yamakawa Y, Minato S, Nakahara K, Nakano S, Wakisaka S. Clinical features of the localized girdle sensation of mid-trunk (false localizing sign) appeared [*sic*] in cervical compressive myelopathy patients. *Journal of Neurology* 2002; **249**: 549-553
Cross References
"false-localizing signs"; Paraparesis; Suspended sensory loss

"Give-Way" Weakness

- see COLLAPSING WEAKNESS; FUNCTIONAL WEAKNESS AND SENSORY DISTURBANCE

Glabellar Tap Reflex

The glabellar tap reflex, also known as Myerson's sign or the nasopalpebral reflex, is elicited by repeated gentle tapping with a finger on the forehead, preferably with irregular cadence and so that the patient cannot see the finger (to avoid blinking due to the threat or menace reflex), while observing the eyelids blink (*i.e*., blink reflex). Usually, reflexive blinking in response to tapping habituates quickly, but in extrapyramidal disorders it may not do so. This sign was once thought useful for the diagnosis of idiopathic Parkinson's disease but in fact it is fairly nonspecific, occurring in many akinetic-rigid disorders.
References
Schott JM, Rossor MN. The grasp and other primitive reflexes. *Journal of Neurology, Neurosurgery and Psychiatry* 2003; **74**: 558-560
Cross References
Blink reflex; Parkinsonism

Glossolalia

Glossolalia, or speaking in tongues, may be considered a normal phenomenon in certain Christian denominations, as divinely inspired, since it is mentioned in the Bible (1 Corinthians, 14:27-33, although St. Paul speaks of the importance of an interpreter, since "God is not the author of confusion"), but it is not confined to Christianity or even overtly religious environments. Others conceptualize glossolalia as a form of automatic speech, usually of a pseudo-language which may be mistaken for a foreign tongue. Such happenings may occur in trance-like states, or in pathological states, such as schizophrenia.

References

Enoch MD, Ball HN. *Uncommon psychiatric syndromes* (4th edition). London: Arnold, 2001: 237-240

"Glove and Stocking" Sensory Loss

Sensory loss, to all or selected modalities, confined to the distal parts of the limbs ("glove and stocking") implies the presence of a peripheral sensory neuropathy. If the neuropathy involves both sensory and motor fibers, motor signs (distal weakness, reflex diminution or loss) may also be present.

Cross References

Neuropathy

Goosebumps

- see ANSERINA

Gordon's Sign

Gordon's sign is an extensor plantar response in response to squeezing the calf muscles, also called the paradoxical flexor response. As with Chaddock's sign and Oppenheim's sign, this reflects an expansion of the receptive field of the reflex.

Cross References

Babinski's sign (1); Plantar response

Gowers' Sign

Gowers' sign is a characteristic maneuver used by patients with proximal lower limb and trunk weakness to rise from the ground. From the lying position, the patient rolls to the kneeling position, pushes on the ground with extended forearms to lift the hips and straighten the legs, so forming a triangle with the hips at the apex with hands and feet on the floor forming the base (known in North America as the "butt-first maneuver"). Then the hands are used to push on the knees and so lift up the trunk ("climbing up oneself"). This sign was originally described by Gowers in the context of Duchenne muscular dystrophy but may be seen in other causes of proximal leg and trunk weakness, *e.g.*, Becker muscular dystrophy, spinal muscular atrophy.

Gowers was not the first to describe the sign; Bell had reported it almost 50 years before Gowers' account.

References

Pearce JMS. Gowers' sign. In: Pearce JMS. *Fragments of neurological history*. London: Imperial College Press, 2003: 378-380

Graefe's Sign

- see VON GRAEFE'S SIGN

Graphanesthesia

- see AGRAPHESTHESIA

Graphesthesia

Graphesthesia is the ability to identify numbers or letters written or traced on the skin, first described by Head in 1920. Loss of this ability (agraphesthesia, dysgraphesthesia, or graphanesthesia; sometimes referred to as agraphognosia) is typically observed with parietal lobe lesions, for example in conditions such as corticobasal degeneration. Such a cortical sensory syndrome may also cause astereognosis and impaired two-point discrimination.

Cross References

Agraphesthesia; Astereognosis; Two-point discrimination

Graphospasm

- see WRITER'S CRAMP

Grasp Reflex

The grasp reflex consists of progressive forced closure of the hand (contraction of flexor and adductor muscles) when tactile stimulation (*e.g.*, the examiner's hand) is moved slowly, exerting pressure, across the patient's palm in an upward direction. Once established, the patient is unable to release the grip (forced grasping), allowing the examiner to draw the arm away from the patient's body. There may also be accompanying groping movements of the hand, once touched, in search of the examiner's hand or clothing (forced groping, magnetic movement). Although categorized a reflex, it may sometimes be accessible to modification by will (so-called alien grasp reflex). It is usually bilateral, even with unilateral pathology. Foot grasping (*i.e.*, flexion and adduction of the toes and curling of the sole in response to pressure on the sole), may coexist, as may other frontal release signs (*e.g.*, pout reflex, palmomental reflex, *gegenhalten*).

The grasp reflex may be categorized as a frontal release sign (or primitive reflex) of prehensile type, since it is most commonly associated with lesion(s) in the frontal lobes or deep nuclei and subcortical white matter. Clinicoradiological correlations suggest the cingulate gyrus is the structure most commonly involved, followed by the supplementary motor area.

References

De Renzi E, Barbieri C. The incidence of the grasp reflex following hemispheric lesion and its relation to frontal damage. *Brain* 1992; **115**: 293-313

Schott JM, Rossor MN. The grasp and other primitive reflexes. *Journal of Neurology, Neurosurgery and Psychiatry* 2003; **74**: 558-560

Cross References

Akinetic mutism; Alien grasp reflex; Frontal release signs

Guttmann's Sign

Guttmann's sign is autonomic overactivity occurring as a feature of the acute phase of high spinal cord lesions, which may manifest with facial vasodilatation associated with nasal congestion, hypertension, bradycardia, sweating, mydriasis and piloerection.

Gynecomastia

Gynecomastia is inappropriate breast development in males. It may be observed in chronic liver disease and in certain neurological diseases:

Excessive pituitary prolactin release secondary to impaired dopamine release from the hypothalamus due to local tumor or treatment with dopaminergic antagonist drugs (*e.g.*, antipsychotic medications)
Kennedy's syndrome (X-linked bulbospinal neuronopathy)
Klinefelter's syndrome
POEMS syndrome.

H

Habit Spasm
- see SPASM; TIC

Hallpike Maneuver, Hallpike Test
The Hallpike maneuver (Nylen-Bárány maneuver, positioning maneuver, Dix-Hallpike positioning test) is a test used in the investigation of vertigo to induce (or to modify) nystagmus by stimulating the otolith organs of the inner ear. It most usually consists of briskly tilting the patient's head backward to 30-45° below the horizontal ("head hanging position") and turning it 45° to one side or the other, thus stimulating the posterior semicircular canal. Prior to performing the maneuver, the examiner should warn the patient that s/he may feel "giddy" or vertiginous, and to keep their eyes open throughout, since the development of nystagmus with the symptoms of vertigo is the observation of interest to the examiner. With a peripheral lesion (*e.g.*, benign paroxysmal positional vertigo, diseases of the labyrinth), nausea, vomiting and rotational-vertical nystagmus occur several seconds after the maneuver and then rapidly fatigue (usually < 30 seconds), only to recur when the patient is returned to the upright position, with the nystagmus now in the opposite direction. Repetition of the maneuver (if the patient can be persuaded to undergo it) causes less severe symptoms (habituation). This is the diagnostic test for benign paroxysmal positional vertigo (BPPV). Central lesions (disorders of the vestibular connections) tend to produce isolated nystagmus which does not fatigue or habituate with repetition.

Variants of the Hallpike maneuver are described for BPPV of anterior or horizontal semicircular canal origin. Caloric testing may be required to elicit the causes of dizziness if the Hallpike maneuver is uninformative.

References

Bronstein AM. Vestibular reflexes and positional manoeuvres. *Journal of Neurology, Neurosurgery and Psychiatry* 2003; **74**: 289-293

Dix MR, Hallpike CS. The pathology, symptomatology and diagnosis of certain common disorders of the vestibular system. *Proceedings of the Royal Society of Medicine* 1952; **45**: 341-354

Lanska DJ, Remler B. Benign paroxysmal positioning vertigo: classic descriptions, origins of the provocative positioning technique, and conceptual developments. *Neurology* 1997; **48**: 1167-1177

Cross References

Caloric testing; Nystagmus; Vertigo; Vestibulo-ocular reflexes

Hallucination

An hallucination is a perception in the absence of adequate peripheral stimulus (*cf*. illusion). Such perceptions are substantial, constant, occur in objective space, and are usually not accompanied by insight. They most usually occur in the visual and auditory domains.

Visual hallucinations may range in complexity. They may be "simple," spots or flashes of light (photopsia, photism, scintillation), or "complex," ranging from patterns (fortification spectra, epileptic aura) to fully formed objects or individuals. They may be transient, such as brief visions of a person or animal (passage hallucinations, for example in Parkinson's disease) or long lasting. Visual hallucinations may be normal, especially when falling asleep or waking (hypnogogic, hypnopompic). There are many other associations including both psychiatric and neurological disease, including:

Delirium: especially hyperalert subtype
Withdrawal states: *e.g*., delirium tremens; hypnotics, anxiolytics
Drug overdose: *e.g*., anticholinergic drugs
Neurodegenerative disorders: dementia with Lewy bodies (a diagnostic criterion) more often than Alzheimer's disease: these may be associated with cholinergic depletion, and improved with cholinesterase inhibitor drugs; idiopathic Parkinson's disease (with or without treatment).
Narcolepsy-cataplexy
Peduncular hallucinosis
Migraine aura
Charles Bonnet syndrome (visual hallucinations of the visually impaired)
Schizophrenia
Epilepsy: complex partial seizures
"Alice in Wonderland" syndrome

Different mechanisms may account for visual hallucinations in different conditions: defective visual input and processing may occur in visual pathway lesions, whereas epilepsy may have a direct irritative effect on brain function; visual hallucinations associated with brainstem lesions may result from neurotransmitter abnormalities (cholinergic, serotonergic).

Auditory hallucinations may be simple (tinnitus) or complex (voices, music) and may be associated with focal pathology in the temporal cortex. Third person hallucinations, commenting on a person's actions, are one of the first rank symptoms of schizophrenia.

References

Ashton H. Delirium and hallucinations. In: Perry E, Ashton H, Young A (eds.). *Neurochemistry of consciousness: neurotransmitters in mind*. Amsterdam: John Benjamins, 2002: 181-203

Barodawala S, Mulley GP. Visual hallucinations. *Journal of the Royal College of Physicians of London* 1997; **31**: 42-48

Manford M, Andermann F. Complex visual hallucinations. Clinical and neurobiological insights. *Brain* 1998; **121**: 1819-1840

Tekin S, Cummings JL. Hallucinations and related conditions. In: Heilman KM, Valenstein E (eds.). *Clinical neuropsychology* (4th edition). Oxford: OUP, 2003: 479-494

Cross References

"Alice in wonderland" syndrome; *Anwesenheit*; Charles bonnet syndrome; Delirium; Fortification spectra; Illusion; *Narcolepsy*; Photism; Photopsia

Hammer Toes

Hammer toes are a feature of hereditary neuropathies, *e.g.*, Charcot-Marie-Tooth disease type I, some cases of hereditary neuropathy with liability to pressure palsies, and Friedreich's ataxia. There may be associated pes cavus.

Cross References

Pes cavus

Harlequin Sign

The harlequin sign is asymmetrical facial flushing with sweating after exercise. That it reflects localized autonomic dysfunction may be indicated by its associations with congenital Horner's syndrome, and as one element in the spectrum of Holmes-Adie syndrome and Ross's syndrome. Harlequin sign has on occasion been described in association with multiple sclerosis and superior mediastinal neurinoma.

References

Carroll CB, Zajicek JP. The "harlequin" sign in association with multiple sclerosis. *Journal of Neurology* 2004; **251**: 1145-1146

Lance JW, Drummond PD, Gandevia SC, Morris JGL. Harlequin syndrome: the sudden onset of unilateral flushing and sweating. *Journal of Neurology, Neurosurgery and Psychiatry* 1988; **51**: 635-642

Cross References

Holmes-adie pupil, Holmes-adie syndrome; Horner's syndrome

Head Droop, Head Drop

- see DROPPED HEAD SYNDROME

Head Impulse Test

The head impulse test, also known as the head thrust test, assesses the vestibulo-ocular reflex. It consists of a rapid turning of the head to one side by about 15 degrees, sufficiently rapid to ensure that smooth pursuit eye movements do not compensate for head turning. The examiner observes the ability of the subject to maintain fixation on a distant target; if the vestibulo-ocular reflex is intact fixation is maintained. If the vestibulo-ocular reflex is impaired, then an easily visible saccade back to the target occurs at the end of the movement. Tilting the head down by 20 degrees and moving the head unpredictably may optimize testing. This test is recommended in patients suffering a first attack of acute spontaneous vertigo. Sensitivity and specificity of around 80% for detecting a peripheral vestibular lesion, such as acute unilateral vestibular neuritis has been reported.

References
Halmagyi GM, Curthoys IS. A clinical sign of canal paresis. *Archives of Neurology* 1988; **45**: 737-739
Schubert MC, Das VE, Tusa RJ, Herdman SJ. Optimizing the sensitivity of the head thrust test for diagnosing vestibular hypofunction. *Neurology* 2002; **58 (suppl3)**: A439 (abstract P06.031)
Cross References
Vertigo; Vestibulo-ocular reflexes

Head Thrust
- see EYELID APRAXIA; OCULAR APRAXIA

Head Tilt
Head tilt may be observed with:

Diplopia, cranial nerve palsies (IV, VI); skew deviation
Neck dystonia (laterocollis)
Incipient tonsillar herniation with cerebellar tumors, sometimes associated with neck stiffness and limitation of neck movement.

Cross References
Bielschowsky's sign, Bielschowsky's test; Diplopia; Laterocollis; Ocular tilt reaction

Head Tremor
Head tremor may be characterized as "yes-yes" (nodding, *tremblement affirmatif*) when predominantly in the vertical plane, or "no-no" (side-to-side, *tremblement negatif*) when predominantly in the horizontal plane.

Head tremor may occur in isolation or with evidence of tremor elsewhere (*e.g.*, postural limb tremor, vocal tremor, in essential tremor), or dystonia (*e.g.*, torticollis). In essential tremor the head movements are often intermittent, "yes-yes," and of frequency about 7 Hz. Dystonic head tremor is often jerky and disorganized, with a frequency of less than 5 Hz. Cerebellum and brainstem disease, such as multiple sclerosis, can also produce head tremor (or titubation). Head tremor is an exceptionally rare symptom of Parkinson's disease. It may also be seen as a consequence of aortic valve regurgitation (De Musset's sign).

Treatment of head tremor varies with cause. Possible treatments, of variable efficacy, include:

- Essential tremor: propranolol, primidone, nicardipine, gabapentin, topiramate
- Dystonic tremor: anticholinergics, propranolol, botulinum toxin injections
- Cerebellar tremor: isoniazid, carbamazepine, ondansetron

Cross References
Dystonia; Tremor

"Head Turning Sign"

It is often observed that patients who are cognitively impaired turn their head toward their spouse, partner, or caregiver to seek assistance when asked to give a history of their problems, or during tests of neuropsychological function. It is a nonspecific sign of cognitive impairment.

Cross References

Dementia

Heautoscopy

This term was coined to denote seeing oneself, encountering ones alter ego or *doppelgänger*. Hence unlike the situation in autoscopy, there are two selves, a reduplicated body rather than a mirror image; egocentric and body-centered perspectives do not coincide.

References

Brugger P. Reflective mirrors: perspective taking in autoscopic phenomena. *Cognitive Neuropsychiatry* 2002; **7**: 179-194

Cross References

Autoscopy; Hallucination

Heel-Knee-Shin Test, Heel-Shin Test

A frequently used test of coordination in which the patient, sitting on the examination couch, is asked to lift the heel onto the contralateral knee, then run it smoothly down the shin bone toward the foot. Jerky performance, or a tendency for the heel to slide off the shin, may be seen in an ataxic limb.

Cross References

Ataxia; Cerebellar syndromes; Shin-tapping

Heel-Toe Walking

- see TANDEM WALKING

Hemeralopia

Hemeralopia, or day blindness, is worsening of vision in bright light (*cf*. nyctalopia). This phenomenon may reflect severe impairment of blood flow to the eye, such that photostressing the macula by exposure to bright light is followed by only slow regeneration of the bleached photopigments.

If due to retinal ischemia, hemeralopia may be accompanied by neovascularization of the retina. Impoverished perfusion pressure may be demonstrated by pressing on the eyeball (*e.g*., with the thumb) during ophthalmoscopy ("digital ophthalmodynamometry") and observing the collapse of retinal arteries: thumb pressure greater than diastolic retinal artery pressure causes intermittent collapse; thumb pressure greater than systolic pressure leads to a cessation of pulsation.

Hemeralopia may also occur in retinal diseases, such as cone dystrophies.

References

Furlan AJ, Whisnant JP, Kearns TP. Unilateral visual loss in bright light: an unusual symptom of carotid artery occlusive disease. *Archives of Neurology* 1979; **36**: 675-676

Cross References

Nyctalopia

Hemiachromatopsia

- see ACHROMATOPSIA; ALEXIA

Hemiakinesia

Hemiakinesia is akinesia or hypokinesia (inability or difficulty initiating movement) confined to one side of the body. Although hemiakinesia is the norm at the onset of idiopathic Parkinson's disease ("hemiparkinsonism"), persistent hemiakinesia should prompt a re-evaluation of this diagnosis. Corticobasal degeneration often remains unilateral; a search for structural lesions of the basal ganglia should also be undertaken. Hemiakinesia may also indicate motor neglect, usually with right-sided lesions. Lesions of the basal ganglia, ventral ("motor") thalamus, limbic system, and frontal lobes may cause hemiakinesia.

Cross References

Akinesia; Extinction; Hemiparkinsonism; Hypokinesia; Neglect; Parkinsonism

Hemialexia

This is the inability to read words in the visual left half-field in the absence of hemianopia. It may occur after callosotomy (complete, or partial involving only the splenium), and represents a visual disconnection syndrome.

References

Zaidel E, Iacobini M, Zaidel DW, Bogen JE. The callosal syndromes. In: Heilman KM, Valenstein E (eds.). *Clinical neuropsychology* (4th edition). Oxford: OUP, 2003: 347-403

Cross References

Alexia; Hemianomia

Hemianomia

This is the absence of verbal report of stimuli presented in the visual left half-field in the absence of hemianopia. It may occur after callosotomy (complete, or partial involving only the splenium), and represents a visual disconnection syndrome.

References

Zaidel E, Iacobini M, Zaidel DW, Bogen JE. The callosal syndromes. In: Heilman KM, Valenstein E (eds.). *Clinical neuropsychology* (4th edition). Oxford: OUP, 2003: 347-403

Cross References

Anomia; Hemialexia

Hemianopia

Hemianopia (hemianopsia) is a defect of one half of the visual field: this may be vertical or horizontal (= altitudinal field defect).

Hemianopic defects may be congruent (homonymous) or noncongruent (heteronymous), and may be detected by standard confrontational testing of the visual fields or automatically (*e.g.*, Goldman perimetry). These tests of the visual fields are an extension of the tests for visual acuity which assess areas away from the fovea. Because of the strict topographic arrangement of neural pathways within the visual system, particular abnormalities of the visual fields give a very precise indication of the likely site of pathology.

- Homonymous hemianopia:
  Reflects a post-chiasmal lesion. It is important to assess whether the vertical meridian of a homonymous hemianopia cuts through the macula (macula splitting), implying a lesion of the optic radiation; or spares the macula (macula sparing), suggesting an occipital cortical lesion. Incongruous defects may be found with lesions of the optic tract. Commonly, homonymous hemianopias result from cerebrovascular disease causing occipital lobe infarction, or intraparenchymal tumor, but they may be "false-localizing" due to raised intracranial pressure if temporal lobe herniation causes posterior cerebral artery compromise.
- Heteronymous hemianopia:
  Reflects a chiasmal lesion. The most common of these is a bitemporal hemianopia due to chiasmal compression, for example by a pituitary lesion or craniopharyngioma. Tilted optic discs may also be associated with bitemporal field loss but this extends to the blind spot and not the vertical meridian as in chiasmal pathology ("pseudobitemporal hemianopia"). Binasal defects are rare, suggesting lateral compression of the chiasm, for example from bilateral carotid artery aneurysms; binasal hemianopia is also described with optic nerve head lesions. Unilateral (monocular) temporal hemianopia may result from a lesion anterior to the chiasm which selectively affects only the ipsilateral crossing nasal fibers (junctional scotoma of Traquair).

Unawareness of visual field loss, anosognosic hemianopia, occurs principally with right-sided brain lesions.

Bilateral homonymous hemianopia or double hemianopia may result in cortical blindness.

Cross References

Alexia; Altitudinal field defect; Anosognosia; Cortical blindness; "False-localizing signs"; Macula sparing, Macula splitting; Quadrantanopia; Scotoma; Visual field defects

Hemiataxia

Hemiataxia is ataxia confined to one half of the body. The vast majority of isolated hemiataxic syndromes reflect a lesion of the ipsilateral

cerebellar hemisphere, but on occasion supratentorial lesions may cause hemiataxia (posterior limb of the internal capsule, thalamus). However, in almost all of these cases hemiataxia coexists with ipsilateral hemiparesis (ataxic hemiparesis, *q.v.*), hemisensory disturbance (hemiataxia-hypesthesia), or both.

References

Luijckx G-J, Boiten J, Lodder J, Heurs-van Raak L, Wilmink J. Isolated hemiataxia after supratentorial brain infarction. *Journal of Neurology, Neurosurgery and Psychiatry* 1994; **57**: 742-744

Cross References

Ataxia; Ataxic hemiparesis; Cerebellar syndromes; Cerebellopontine angle syndrome; Lateral medullary syndrome

Hemiballismus

Hemiballismus is unilateral ballismus, an involuntary hyperkinetic movement disorder in which there are large amplitude, vigorous ("flinging") irregular movements. Hemiballismus overlaps clinically with hemichorea ("violent chorea"); the term *hemiballismus-hemichorea* is sometimes used to reflect this overlap. Hemiballismic limbs may show a loss of normal muscular tone (hypotonia).

Anatomically, hemiballismus is most often associated with lesions of the contralateral subthalamic nucleus of Luys or its efferent pathways, although there are occasional reports of its occurrence with lesions of the caudate nucleus, putamen, globus pallidus, lentiform nucleus, thalamus, and precentral gyrus; and even with ipsilateral lesions. Pathologically, vascular events (ischemia, hemorrhage) are the most common association but hemiballismus has also been reported with space-occupying lesions (tumor, arteriovenous malformation), inflammation (encephalitis, systemic lupus erythematosus, post-streptococcal infection), demyelination, metabolic causes (hyperosmolal nonketotic hyperglycemia), infection (toxoplasmosis in AIDS), drugs (oral contraceptives, phenytoin, levodopa, neuroleptics) and head trauma.

Pathophysiologically, hemiballismus is thought to result from reduced conduction through the direct pathway within the basal ganglia-thalamo-cortical motor circuit (as are other hyperkinetic involuntary movements, such as choreoathetosis). Removal of excitation from the globus pallidus following damage to the efferent subthalamic-pallidal pathways disinhibits the ventral anterior and ventral lateral thalamic nuclei which receive pallidal projections and which in turn project to the motor cortex.

Hemiballismus of vascular origin usually improves spontaneously, but drug treatment with neuroleptics (haloperidol, pimozide, sulpiride) may be helpful. Other drugs which are sometimes helpful include tetrabenazine, reserpine, clonazepam, clozapine, and sodium valproate.

References

Albin RL, Young AB, Penney JB. The functional anatomy of basal ganglia disorders. *Trends in Neurosciences* 1989; **12**: 366-375

Lee MS, Marsden CD. Movement disorders following lesions of the thalamus or subthalamic region. *Movement Disorders* 1994; **9**: 493-507

Martin JP. Hemichorea resulting from a local lesion of the brain. (The syndrome of the body of Luys.) *Brain* 1927; **50**: 637-651

Cross References

Ballism, Ballismus; Chorea, Choreoathetosis; Hemichorea; Hypotonia, Hypotonus

Hemichorea

Hemichorea is unilateral chorea, an involuntary movement disorder which overlaps with hemiballismus, and with which it shares a similar pathophysiology and etiology. It may replace hemiballismus during recovery from a contralateral subthalamic lesion.

Cross References

Chorea, Choreoathetosis; Hemiballismus

Hemidystonia

Hemidystonia is dystonia affecting the whole of one side of the body, a pattern which mandates structural brain imaging because of the chance of finding a causative structural lesion (vascular, neoplastic), which is greater than with other patterns of dystonia (focal, segmental, multifocal, generalized). Such a lesion most often affects the contralateral putamen or its afferent or efferent connections.

References

Marsden CD, Obeso JA, Zaranz JJ, Lang AE. The anatomical basis of symptomatic hemidystonia. *Brain* 1985; **108**: 461-483

Cross References

Dystonia

Hemifacial Atrophy

Hemifacial atrophy is thinning of subcutaneous tissues on one side of the face; it may also involve muscle and bone (causing enophthalmos), and sometimes brain, in which case neurological features (hemiparesis, hemianopia, focal seizures, cognitive impairment) may also be present.

The clinical heterogeneity of hemifacial atrophy probably reflects pathogenetic heterogeneity. The syndrome, sometimes referred to as Parry-Romberg syndrome, may result from maldevelopment of autonomic innervation or vascular supply, or as an acquired feature following trauma, or a consequence of linear scleroderma (morphea), in which case a *coup de sabre* may be seen.

References

Larner AJ. Neurological contributions of Caleb Hillier Parry. *Advances in Clinical Neuroscience & Rehabilitation* 2004; **4(3)**: 38-39

Larner AJ, Bennison DP. Some observations on the aetiology of hemifacial atrophy ("Parry-Romberg syndrome"). *Journal of Neurology, Neurosurgery and Psychiatry* 1993; **56**: 1035-1036

Cross References

Coup de sabre; Enophthalmos; Hemianopia; Hemiparesis

Hemifacial Spasm

Hemifacial spasm is an involuntary dyskinetic (not dystonic) movement disorder consisting of painless contractions of muscles on one

side of the face, sometimes triggered by eating or speaking, and exacerbated by fatigue or emotion. The movements give a twitching appearance to the eye or side of the mouth, sometimes described as a pulling sensation. Patients often find this embarrassing because it attracts the attention of others. The movements may continue during sleep. Paradoxical elevation of the eyebrow as orbicularis oris contracts and the eye closes may be seen (Babinski's "other sign"). Very rarely, movements may be bilateral.

Hemifacial spasm may be idiopathic, or associated with neurovascular compression of the facial (VII) nerve, usually at the root entry zone, often by a tortuous anterior or posterior inferior cerebellar artery. Other causes include intrapontine lesions (*e.g.*, demyelination), following a Bell's palsy, and mass lesions (tumor, arteriovenous malformation) located anywhere from the facial nucleus to the stylomastoid foramen. Very rarely, contralateral (false-localizing) posterior fossa lesions have been associated with hemifacial spasm, suggesting that kinking or distortion of the nerve, rather than direct compression, may be of pathogenetic importance.

Structural lesions may be amenable to surgical resection. For idiopathic hemifacial spasm, or patients declining surgery, botulinum toxin injections are the treatment of choice.

References

Evidente VGH, Adler Ch H. Hemifacial spasm and other craniofacial movement disorders. *Mayo Clinic Proceedings* 1998; **73**: 67-71

Cross References

Babinski's sign (2); Bell's palsy; Dyskinesia; "False-localizng signs"

Hemiinattention

- see NEGLECT

Hemimicropsia

- see MICROPSIA

Hemineglect

- see NEGLECT

Hemiparesis

Hemiparesis is a weakness affecting one side of the body, less severe than a hemiplegia. Characteristically this affects the extensor muscles of the upper limb more than flexors, and the flexors of the leg more than extensors ("pyramidal" distribution of weakness), producing the classic hemiparetic/hemiplegic posture with flexed arm and extended leg, the latter permitting standing and a circumducting gait.

Hemiparesis results from damage (most usually vascular) to the corticospinal pathways anywhere from motor cortex to the cervical spine. Accompanying signs may give clues as to localization, the main possibilities being hemisphere, brainstem, or cervical cord. Hemisphere lesions may also cause hemisensory impairment, hemi-

anopia, aphasia, agnosia or apraxia; headache, and incomplete unilateral ptosis, may sometimes feature. Spatial neglect, with or without anosognosia, may also occur, particularly with right-sided lesions producing a left hemiparesis. Pure motor hemiparesis may be seen with lesions of the internal capsule, corona radiata, and basal pons (lacunar/small deep infarct), in which case the face and arm are affected more than the leg; such facio-brachial predominance may also be seen with cortico-subcortical lesions laterally placed on the contralateral hemisphere. Crural predominance suggests a contralateral paracentral cortical lesion or one of the lacunar syndromes.

Brainstem lesions may produce diplopia, ophthalmoplegia, nystagmus, ataxia, and crossed facial sensory loss or weakness in addition to hemiparesis ("alternating hemiplegia").

Spinal lesions are more likely to show bilateral long tract signs (*e.g.*, bilateral Babinksi's sign) and may have accompanying spinal or root pain, sphincter disturbance, and a sensory or motor level.

Hemiparesis is most usually a consequence of a vascular event (cerebral infarction). Tumor may cause a progressive hemiparesis (although meningiomas may produce transient "stroke-like" events). Hemiparetic multiple sclerosis is rare but well described. Transient hemiparesis may be observed as an ictal phenomenon (Todd's paresis), or in familial hemiplegic migraine which is associated with mutations in a voltage-gated Ca^{2+} ion channel gene.

Cross References

Agnosia; Anosognosia; Aphasia; Apraxia; Babinski's sign (1); "False-localizing signs"; Hemianopia; Hemiplegia; Neglect; Ptosis; Upper motor neurone (UMN) syndrome; Weakness

Hemiparkinsonism

Hemiparkinsonism describes the finding of parkinsonian signs restricted to one side of the body, most usually akinesia, in which case the term hemiakinesia may be used. Idiopathic Parkinson's disease may present with exclusively or predominantly unilateral features (indeed, lack of asymmetry at onset may argue against this diagnosis) but persistent hemiparkinsonism, particularly if unresponsive to adequate doses of levodopa, should alert the clinician to other possible diagnoses, including corticobasal degeneration or structural lesions.

Cross References

Hemiakinesia; Parkinsonism

Hemiplegia

Hemiplegia is a complete weakness affecting one side of the body, *i.e.*, clinically a more severe picture than hemiparesis.

Cross References

Hemiparesis; Weakness

Hemiplegia Cruciata

Cervicomedullary junction lesions where the pyramidal tract decussates may result in paresis of the contralateral upper extremity and

ipsilateral lower extremity. There may be concurrent facial sensory loss with onion skin pattern, respiratory insufficiency, bladder dysfunction and cranial nerve palsies. Such cases are very rare.

Hennebert's Sign

Hennebert's sign is the induction of vertigo and nystagmus by pressure changes in the external auditory canal, such as when using pneumatic otoscopy or simply with tragal pressure. These findings are highly suggestive of the presence of a bony labyrinthine fistula. There may be a history of chronic otitis media.

Cross References

Nystagmus; Vertigo

Hertwig-Magendie Sign

- see SKEW DEVIATION

Heterochromia Iridis

Different color of the irides may be seen in congenital Horner's syndrome, and in Waardenburg syndrome of nerve deafness, white forelock, abnormal skin pigmentation, and synophrys.

Cross References

Horner's syndrome

Heterophoria

Heterophoria is a generic term for a latent tendency to imbalance of the ocular axes (latent strabismus; *cf*. heterotropia). This may be clinically demonstrated using the cover-uncover test: if there is movement of the covered eye as it is uncovered and takes up fixation, this reflects a phoria. Phorias may be in the horizontal (esophoria, exophoria) or vertical plane (hyperphoria, hypophoria).

References

Shaunak S, O'Sullivan E, Kennard C. Eye movements. In: Hughes RAC (ed.). *Neurological investigations*. London: BMJ Publishing, 1997: 253-282

Cross References

Cover tests; Esophoria; Exophoria; Heterotropia; Hyperphoria; Hypophoria

Heterotropia

Heterotropia is a generic term for manifest deviation of the eyes (manifest strabismus; *cf*. heterophoria), synonymous with squint. This may be obvious; an amblyopic eye, with poor visual acuity and fixation, may become deviated. Sometimes it may be more subtle, coming to attention only with the patient's complaint of diplopia.

Using the alternate cover (cross cover) test, in which binocular fixation is not permitted, an imbalance in the visual axes may be demonstrated, but this will not distinguish between heterotropia and heterophoria. To make this distinction the cover test is required: if the uncovered eye moves to adopt fixation then heterotropia is confirmed.

Tropias may be in the horizontal (esotropia, exotropia) or vertical plane (hypertropia, hypotropia).

References

Shaunak S, O'Sullivan E, Kennard C. Eye movements. In: Hughes RAC (ed.). *Neurological Investigations*. London: BMJ Publishing, 1997: 253-282

Cross References

Amblyopia; Cover tests; Esotropia; Exotropia; Heterophoria; Hypertropia; Hypotropia

Hiccups

A hiccup (hiccough) is a brief burst of inspiratory activity involving the diaphragm and the inspiratory intercostal muscles with reciprocal inhibition of expiratory intercostal muscles. The sound ("hic") and discomfort result from glottic closure immediately after the onset of diaphragmatic contraction, *i.e*., the latter is insufficient or asynchronous. Hiccups may be characterized as a physiological form of myoclonus (or singultus).

Most episodes of hiccups are self-limited, but prolonged or intractable hiccuping (*hocquet diabolique*) should prompt a search for a structural or functional cause, either gastroenterological or neurological. Hiccuping is seldom the only abnormality if the cause is neurological since it usually reflects pathology within the medulla or affecting the afferent and efferent nerves of the respiratory muscles. Medullary causes include:

Infarction (posterior inferior cerebellar artery territory; lateral medullary syndrome, especially middle level and dorsolateral lesion locations)
Tumor
Abscess
Tuberculoma
Syrinx
Hematoma
Demyelination
CNS infection, *e.g*., viral encephalitis

Treatment should be aimed at the underlying cause. If none is identified, physical measures to stop the hiccups, such as rebreathing, may then be tried. Of the many various pharmacotherapies tried, the best are probably baclofen and chlorpromazine.

References

Davis JW. An experimental study of hiccup. *Brain* 1970; **93**: 851-872
Fetter M, Kennard C. Hiccup. In: Brandt T, Caplan LR, Dichgans J, Diener HC, Kennard C (eds.). *Neurological disorders: course and treatment*. San Diego: Academic Press, 1996: 145-148
Howard RS. Persistent hiccups. *BMJ* 1992; **305**: 1237-1238
Park MH, Kim BJ, Koh SB, Park MK, Park KW, Lee DH. Lesional location of lateral medullary infarction presenting hiccups (singultus). *Journal of Neurology, Neurosurgery and Psychiatry* 2005; **76**: 95-98

Cross References
Lateral medullary syndrome; myoclonus

Hip Abduction Sign

The hip abduction sign refers to abduction of the thighs when attempting to rise from the ground, due to relative weakness of hip adductors with preserved strength in hip abductors. The sign was first described in patients with sarcoglycanopathies, a group of autosomal recessive limb-girdle muscular dystrophies, and is reported to have a sensitivity of 76% and a specificity of 98% for this diagnosis. It may perhaps be envisaged as the equivalent to Gowers' sign but with hip adductor, rather than gluteal, weakness.

References

Khadilkar SV, Singh RK. Hip abduction sign: a new clinical sign in sarcoglycanopathies. *Journal of Clinical Neuromuscular Disease* 2001; **3**: 13-15

Cross References
Gowers' sign

Hippus

Hippus is excessive pupillary unrest, *i.e.*, rhythmic, oscillatory, contraction and dilatation of the pupil. It may reflect an imbalance between afferent pupillary sympathetic and parasympathetic autonomic activity. Hippus may be a normal phenomenon; it may be observed during recovery from an oculomotor (III) nerve palsy, but otherwise is of no localizing significance.

Hitselberg Sign

Hypoesthesia of the posterior wall of the external auditory canal may be seen in facial paresis since the facial nerve sends a sensory branch to innervate this territory.

Cross References
Facial paresis

Hocquet Diabolique

- see HICCUPS

Hoffmann's Sign

Hoffmann's sign or reflex is a digital reflex consisting of flexion of the thumb and index finger in response to snapping or flicking the distal phalanx of the middle finger, causing a sudden extension of the joint. Although sometimes a normal finding, for example in the presence of generalized hyperreflexia (anxiety, hyperthyroidism), it may be indicative of a corticospinal tract lesion above C5 or C6, particularly if present unilaterally.

Cross References
Trömner's sign; Upper motor neurone (UMN) Syndrome

Hoffmann-Tinel Sign
- see TINEL'S SIGN

Holmes-Adie Pupil, Holmes-Adie Syndrome
The Holmes-Adie, or tonic, pupil is an enlarged pupil which, in a darkened environment, is unresponsive to a phasic light stimulus, but may respond slowly to a tonic light stimulus. Reaction to accommodation is preserved (partial iridoplegia), hence this is one of the causes of light-near pupillary dissociation (*q.v.*). A Holmes-Adie pupil is usually unilateral, and hence a cause of anisocoria.

Holmes-Adie pupil may be associated with other neurological features (Holmes-Adie syndrome). These include loss of lower limb tendon reflexes (especially ankle jerks); impaired corneal sensation; chronic cough; and localized or generalized anhidrosis, sometimes with hyperhidrosis (Ross's syndrome). Holmes-Adie syndrome is much commoner in women than men.

Pathophysiologically Holmes-Adie pupil results from a peripheral lesion of the parasympathetic autonomic nervous system and shows denervation supersensitivity, constricting with application of dilute (0.2%) pilocarpine (*cf*. pseudo-Argyll Robertson pupil).

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

Martinelli P. Holmes-Adie syndrome. *Lancet* 2000; **356**: 1760-1761

Pearce JMS. The Holmes-Adie tonic pupil and Hughlings Jackson. In: Pearce JMS. *Fragments of neurological history*. London: Imperial College Press, 2003: 249-251

Cross References

Anhidrosis; Anisocoria; Hyperhidrosis; Light-near pupillary dissociation; Pseudo-argyll robertson pupil

Holmes's Tremor
Holmes's tremor, also known as rubral tremor, or midbrain tremor, has been defined as a rest and intention tremor, of frequency < 4.5 Hz. The rest tremor may resemble parkinsonian tremor, and is exacerbated by sustained postures and voluntary movements. Hence there are features of rest, postural and kinetic (intention) tremor. Once attributed to lesions of the red nucleus (hence "rubral"), the anatomical substrate is now thought to be interruption of fibers of the superior cerebellar peduncle (hence "midbrain") carrying cerebellothalamic and/or cerebello-olivary projections; lesions of the ipsilateral cerebellar dentate nucleus may produce a similar clinical picture. Recognized causes include multiple sclerosis, head injury and stroke. If a causative lesion is defined, there is typically a delay before tremor appearance (4 weeks to 2 years).

References

Alusi SH, Worthington J, Glickman S, Bain PG. A study of tremor in multiple sclerosis. *Brain* 2001; **124**: 720-730

Deuschl G, Bain P, Brin M and an Ad Hoc Scientific Committee. Consensus statement of the Movement Disorder Society on tremor. *Movement Disorders* 1998; **13(suppl3)**: 2-23

Cross References

Tremor

Hoover's Sign

Hoover's sign may be used to help differentiate organic from functional hemiplegia or monoplegia. It is based on the fact that when a recumbent patient attempts to lift one leg, downward pressure is felt under the heel of the other leg, hip extension being a normal synergistic or synkinetic movement. The finding of this synkinetic movement, detected when the heel of the supposedly paralyzed leg presses down on the examiner's palm, constitutes Hoover's sign: no increase in pressure is felt beneath the heel of a paralyzed leg in an organic hemiplegia.

In addition, the synkinetic hip extension movement is accentuated when attempting to raise a contralateral paretic leg, whereas in functional weakness it is abolished.

References

Stone J, Sharpe M. Hoover's sign. *Practical Neurology* 2001; **1**: 50-53

Cross References

"Arm drop"; Babinski's trunk-thigh test; Functional weakness and sensory disturbance; Synkinesia, Synkinesis

Horner's Syndrome

Horner's syndrome is defined by a constellation of clinical findings, most usually occurring unilaterally, *viz*.:

- Partial ptosis, due to weakness of Müller's muscle
- Miosis, due to the unopposed action of the sphincter pupillae muscle, innervated by the parasympathetic nervous system
- Anhidrosis, a loss of sweating (if the lesion is distal to the superior cervical ganglion)
- Enophthalmos, retraction of the eyeball (though this is seldom measured).

The first two mentioned signs are usually the most evident and bring the patient to medical attention; the latter two are usually less evident or absent.

Additional features which may be seen include:

- Heterochromia iridis, different color of the iris (if the lesion is congenital)
- Elevation of the inferior eyelid due to a weak inferior tarsal muscle ("reverse ptosis," or "upside-down ptosis").

Horner's syndrome results from impairment of ocular sympathetic innervation. The sympathetic innervation of the eye consists of a long, three neurone, pathway, extending from the diencephalon down

to the cervicothoracic spinal cord, then back up to the eye via the superior cervical ganglion and the internal carotid artery, and the ophthalmic division of the trigeminal (V) nerve. A wide variety of pathological processes, spread across a large area, may cause a Horner's syndrome, although many examples remain idiopathic despite intensive investigation. Recognized causes include:

- Brainstem/cervical cord disease (vascular, demyelination, syringomyelia)
- Pancoast tumor
- Malignant cervical lymph nodes
- Carotid aneurysm, carotid artery dissection
- Involvement of T1 fibers, *e.g* in T1 radiculopathy, or lower trunk brachial plexopathy
- Cluster headache

References

Pearce JMS. Claude Bernard-Horner's syndrome and Edward Selleck Hare. In: Pearce JMS. *Fragments of neurological history*. London: Imperial College Press, 2003: 252-255

Cross References

Anhidrosis; Anisocoria; Enophthalmos; Miosis; Plexopathy; Ptosis; Radiculopathy

Hoyt-Spencer Sign

This name is given to the triad of findings characteristic of chronic optic nerve compression, especially due to spheno-orbital optic nerve sheath meningiomas:

- Optociliary shunt vessels
- Disc pallor
- Visual loss

"Hung-Up" Reflexes

- see WOLTMAN'S SIGN

Hutchinson's Pupil

Hutchinson's pupil is unilateral pupillary dilatation ipsilateral to a supratentorial (usually extrinsic) space-occupying lesion, which may be the earliest sign of raised intracranial pressure. It reflects involvement of peripheral pupilloconstrictor fibers in the oculomotor (III) nerve, perhaps due to compression on the margin of the tentorium.

Cross References

Anisocoria; Mydriasis; Oculomotor (III) nerve palsy

Hyperacusis

Hyperacusis is an abnormal loudness of sounds, especially low tones, due to paralysis of the stapedius muscle, whose normal reflex function is to damp conduction across the ossicular chain of the middle ear. This most commonly occurs with lower motor neurone facial (VII) nerve (Bell's) palsy, located proximal to the nerve to stapedius. Ageusia

may also be present if the chorda tympani branch of the facial nerve is involved. Hyperacusis may occasionally occur with central (brainstem) lesions.

Reduction or absence of the stapedius reflex may be tested using the stethoscope loudness imbalance test: with a stethoscope placed in the patients ears, a vibrating tuning fork is placed on the bell. Normally the perception of sound is symmetrical, but sound lateralizes to the side of facial paresis if the attenuating effect of the stapedius reflex is lost.

Cross References

Ageusia; Bell's palsy; Facial paresis

Hyperalgesia

Hyperalgesia is the exaggerated perception of pain from a stimulus which is normally painful (*cf*. allodynia). This may result from sensitization of nociceptors (paradoxically this may sometimes be induced by morphine) or abnormal ephaptic cross-excitation between primary afferent fibers.

Cross References

Allodynia; Dysesthesia; Hyperpathia

Hyperekplexia

Hyperekplexia (literally, to jump excessively) is an involuntary movement disorder in which there is a pathologically exaggerated startle response, usually to sudden unexpected auditory stimuli, but sometimes also to tactile (especially trigeminal) and visual stimuli. The startle response is a sudden shock-like movement which consists of eye blink, grimace, abduction of the arms, and flexion of the neck, trunk, elbows, hips, and knees. The muscular jerk of startle satisfies the definition of myoclonus.

Ideally for hyperekplexia to be diagnosed there should be a physiological demonstration of exaggerated startle response, but this criterion is seldom adequately fulfilled.

Hyperekplexia syndromes may be classified as:

- *Idiopathic*: the majority
- *Hereditary/familial*:
  An autosomal dominant disorder with muscular hypertonia in infancy, leg jerks and gait disorder. Familial cases have been associated with mutations in the α_1 subunit of the inhibitory glycine receptor gene
- *Symptomatic*:
  perinatal ischemic-hypoxic encephalopathy
  brainstem lesions (encephalitis, hemorrhage)
  thalamic lesions (inflammation, vascular)
  drugs (cocaine, amphetamines)
  Gilles de la Tourette syndrome

Attacks may respond to the GABA agonist clonazepam.

References

Matsumoto J, Hallett M. Startle syndromes. In: Marsden CD, Fahn S (eds.) *Movement disorders 3*. Boston: Butterworth, 1994: 418-433

Shiang R, Ryan SG, Zhu Y-Z, *et al*. Mutational analysis of familial and sporadic hyperekplexia. *Annals of Neurology* 1995; **38**: 85-91

Cross References

Incontinence; Myoclonus

Hyperesthesia

Hyperesthesia is increased sensitivity to sensory stimulation of any modality, *e.g*., pain (hyperalgesia), touch.

Cross References

Anesthesia; Hyperalgesia

Hypergraphia

Hypergraphia is a form of increased writing activity. It has been suggested that it should refer specifically to all transient increased writing activity with a noniterative appearance at the syntactic or lexicographemic level (*cf*. automatic writing behavior). Hypergraphia may be seen as part of the interictal psychosis which sometimes develops in patients with complex partial seizures from a temporal lobe (especially nondominant hemisphere) focus, or with other nondominant temporal lobe lesions (vascular, neoplastic, demyelinative, neurodegenerative), or psychiatric disorders (schizophrenia). Hypergraphia is a feature of Geschwind's syndrome, along with hyperreligiosity and hyposexuality.

References

Benson DF. The Geschwind syndrome. *Advances in Neurology* 1991; **55**: 411-421

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

Automatic writing behavior; Hyperreligiosity; Hyposexuality

Hyperhidrosis

Hyperhidrosis is excessive (unphysiological) sweating. This may be "essential" (*i.e*., without obvious cause), or seen as a feature of acromegaly, Parkinson's disease, or occurring in a band above a spinal cord injury. Localized hyperhidrosis caused by food (gustatory sweating) may result from aberrant connections between nerve fibers supplying sweat glands and salivary glands. Other causes of hyperhidrosis include mercury poisoning, phaeochromocytoma, and tetanus. Transient hyperhidrosis contralateral to a large cerebral infarct in the absence of autonomic dysfunction has also been described. Regional syndromes of hyperhidrosis (hands, feet, axillae) are also described.

Treatment is difficult. Symptoms may be helped (but not abolished) by low dose anticholinergic drugs, clonidine or propantheline. For focal syndromes, botulinum toxin injections or sympathectomy may be helpful.

References

Collin J, Whatling P. Treating hyperhidrosis. *BMJ* 2000; **320**: 1221-1222

Labar DR, Mohr JP, Nichols FT, Tatemichi TK. Unilateral hyperhidrosis after cerebral infarction. *Neurology* 1988; **38**: 1679-1682
Naumann M, Flachenecker P, Brocker EB, Toyka KV, Reiners K. Botulinum toxin for palmar hyperhidrosis. *Lancet* 1997; **349**: 252

Cross References

Anhidrosis; Diaphoresis; Holmes-adie pupil, Holmes-adie syndrome

Hyperkinesia

Hyperkinesia indicates an involuntary movement disorder characterized by excessive amplitude of movement, such as ballism, or chorea, or the speech disorders occurring with them.

Cross References

Ballism, ballismus; Chorea, choreoathetosis; Dysarthria

Hypermetamorphosis

Hypermetamorphosis is an overattention to external stimuli. Patients with hypermetamorphosis may explore compulsively and touch everything in their environment. This is one element of the environmental dependency syndrome and may be associated with other forms of utilization behavior, imitation behavior (echolalia, echopraxia) and frontal release signs, such as the grasp reflex. It occurs with severe frontal lobe damage and may be observed following recovery from herpes simplex encephalitis and in frontal lobe dementias including Pick's disease. Bitemporal lobectomy may also result in hypermetamorphosis, as a feature of the Klüver-Bucy syndrome.

Cross References

Attention; Echolalia; Echopraxia; Frontal release signs; Grasp reflex; imitation behavior; Klüver-bucy syndrome; Utilization behavior

Hypermetria

- see DYSMETRIA

Hypermnesia

- see EIDETIC MEMORY; SYNESTHESIA

Hyperorality

Hyperorality is a neurobehavioral abnormality consisting of drinking more than usual, eating excessively, eating anything in sight, and putting objects inappropriately into the mouth. It is a feature of frontal lobe pathology. It is one element of the Klüver-Bucy syndrome, along with hypersexuality.

Cross References

Klüver-bucy syndrome

Hyperpathia

Hyperpathia is an unpleasant sensation, often a burning pain, associated with elevated threshold for cutaneous sensory stimuli, such as light touch or hot and cold stimuli, especially repetitive stimuli. Even light stimuli may produce pain. Clinical features of hyperpathia may

include summation (pain perception increases with repeated stimulation) and aftersensations (pain continues after stimulation has ceased).

The term thus overlaps to some extent with hyperalgesia (although the initial stimulus need not be painful itself) and dysesthesia. There is an accompanying diminution of sensibility due to raising of the sensory threshold (*cf.* allodynia), and the pain is not stimulus-bound (*i.e.*, spreads beyond the area of stimulation).

Hyperpathia is a feature of thalamic lesions, and hence tends to involve the whole of one side of the body following a unilateral lesion, such as a cerebral hemorrhage or thrombosis. Generalized hyperpathia may also be seen in variant Creutzfeldt-Jakob disease, in which posterior thalamic (pulvinar) lesions are said to be a characteristic neuroradiological finding.

Cross References

Allodynia; Dysesthesia; Hyperalgesia

Hyperphagia

Hyperphagia is increased or excessive eating. Binge eating, particularly of sweet things, is one of the neurobehavioral disturbances seen in certain of the frontotemporal dementias. Hyperphagia may be one feature of a more general tendency to put things in the mouth (hyperorality), for example in the Klüver-Bucy syndrome.

Cross References

Hyperorality; Klüver-bucy syndrome

Hyperphoria

Hyperphoria is a variety of heterophoria in which there is a latent upward deviation of the visual axis of one eye. Using the cover-uncover test, this may be observed clinically as the downward movement of the eye as it is uncovered.

Cross References

Cover tests; Heterophoria; Hypophoria

Hyperpilaphesie

The name given to the augmentation of tactile faculties in response to other sensory deprivation, for example touch sensation in the blind.

Hyperpronation

- see CHOREA, CHOREOATHETOSIS; DECEREBRATE RIGIDITY

Hyperreflexia

Hyperreflexia is an exaggerated briskness of the tendon reflexes. This may be physiological in an anxious patient (reflexes often denoted ++), or pathological in the context of corticospinal pathway pathology (upper motor neurone syndrome, often denoted +++). It is sometimes difficult to distinguish normally brisk reflexes from pathologically brisk reflexes. Hyperreflexia (including a jaw jerk) in isolation cannot be used to diagnose an upper motor neurone syndrome, and asymmetry of reflexes is a soft sign. On the other hand, upgoing plantar

responses are a hard sign of upper motor neurone pathology; other accompanying signs (weakness, sustained clonus, absent abdominal reflexes) also indicate abnormality.

Hyperreflexia reflects an increased gain in the stretch reflex. This may be due to impaired descending inhibitory inputs to the monosynaptic reflex arc. Rarely pathological hyperreflexia may occur in the absence of spasticity, suggesting different neuroanatomical substrates underlying these phenomena.

"Hyperreflexia" of the bladder detrusor muscle may be a cause of urinary urge incontinence.

References

Sherman SJ, Koshland GF, Laguna JF. Hyper-reflexia without spasticity after unilateral infarct of the medullary pyramid. *Journal of the Neurological Sciences* 2000; **175**: 145-155

Cross References

Abdominal reflexes; Clonus; Incontinence; Jaw jerk; Reflexes; Spasticity; Upper motor neurone (UMN) syndrome; Weakness

Hyperreligiosity

Hyperreligiosity is a neurobehavioral symptom, manifest as sudden religious conversion, or increased and unswerving orthodoxy in devotion to religious rituals. It may be encountered along with hypergraphia and hyposexuality as a feature of Geschwind's syndrome. It has also been observed in some patients with frontotemporal dementia; the finding is cross-cultural, having been described in Christians, Moslems, and Sikhs. In the context of refractory epilepsy, it has been associated with reduced volume of the right hippocampus, but not right amygdala.

References

Benson DF. The Geschwind syndrome. *Advances in Neurology* 1991; **55**: 411-421

Wuerfel J, Krishnamoorthy ES, Brown RJ *et al*. Religiosity is associated with hippocampal but not amygdala volumes in patients with refractory epilepsy. *Journal of Neurology, Neurosurgery and Psychiatry* 2004; **75**: 640-642

Cross References

Hypergraphia; Hyposexuality

Hypersexuality

Hypersexuality is a pathological increase in sexual drive and activity. Recognized causes include bilateral temporal lobe damage, as in the Klüver-Bucy syndrome, septal damage, hypothalamic disease (rare) with or without subjective increase in libido, and drug-treatment in Parkinson's disease. Hypersexuality is also a feature of the Kleine-Levin syndrome. Sexual disinhibition may be a feature of frontal lobe syndromes, particularly of the orbitofrontal cortex.

Cross References

Disinhibition; Frontal lobe syndromes; Klüver-bucy syndrome

Hypersomnolence

Hypersomnolence is characterized by excessive daytime sleepiness, with a tendency to fall asleep at inappropriate times and places, for example during meals, telephone conversations, at the wheel of a car.

Causes of hypersomnolence include:

- Narcolepsy or the narcoleptic syndrome: may be accompanied by other features such as sleep paralysis, hypnagogic hallucinations, cataplexy
- Midbrain lesions
- Idiopathic CNS hypersomnia
- Kleine-Levin syndrome
- Nocturnal hypoventilation, due to:
- Obstructive sleep apnea-hypopnea syndrome (OSAHS; Pickwickian syndrome)
- Chest wall anomalies
- Neuromuscular and myopathic disorders affecting the respiratory muscles, especially the diaphragm, for example:
  - Motor neurone disease
  - Myotonic dystrophy
  - Metabolic myopathies, for example, acid maltase deficiency
  - Mitochondrial disorders
- Drugs: benzodiazepines, ergot-derivative dopamine agonists
- Post-stroke sleep-related disorders.

Nocturnal hypoventilation as a consequence of obstructed breathing, often manifest as snoring, causes arterial oxygen desaturation as a consequence of hypopnea/apnea which may lead to disturbed sleep, repeated arousals associated with tachycardia and hypertension. Clinical signs may include a bounding hyperdynamic circulation and sometimes papilledema, as well as features of any underlying neuromuscular disease. OSAHS may present in the neurology clinics with loss of consciousness (sleep secondary to hypersomnolence), stroke, morning headaches, and cognitive impairment (slowing). Investigations may reveal a raised hematocrit and early morning hypoxia. Sleep studies confirm nocturnal hypoventilation with dips in arterial oxygen saturation. Treatment is with nocturnal intermittent positive pressure ventilation. Modafinil is also licensed for this indication.

Cross References

Asterixis; Cataplexy; Papilledema; Paradoxical breathing; Snoring

Hyperthermia

Body temperature is usually regulated within narrow limits through the coordinating actions of a centre for temperature control ("thermostat"), located in the hypothalamus (anterior-preoptic area), and effector mechanisms (shivering, sweating, panting, vasoconstriction, vasodilation), controlled by pathways located in or running through the posterior hypothalamus and peripherally in the autonomic nervous

system. Lesions of the anterior hypothalamus (*e.g.*, trauma, ischemia, inflammation, tumor) may result in hyperthermia (*cf.* hypothermia). Other recognized causes of hyperthermia include:

Infection: bacteria, viruses (pyrogens, *e.g.*, interleukin-1)
Malignant hyperthermia
Neuroleptic malignant syndrome (hyperthermia, rigidity, autonomic dysfunction)
Heatstroke
Hyperthyroidism
Phaeochromocytoma crisis.

Cross References
Hypothermia

Hypertonia, Hypertonus

Hypertonia or hypertonus is an exaggeration of normal muscular tone, manifest as resistance to passive movement. It usually implies spasticity of corticospinal (pyramidal) pathway origin, rather than extrapyramidal (lead-pipe) rigidity.
Cross References
Clasp-knife phenomenon; Hyperreflexia; Paratonia; Rigidity; Spasticity; Upper motor neurone (UMN) syndrome

Hypertrophy

- see MUSCLE HYPERTROPHY

Hypertropia

Hypertropia is a variety of heterotropia in which there is manifest upward vertical deviation of the visual axis of one eye. Using the cover test, this manifests as downward movement of the uncovered eye. Depending on the affected eye, this finding is often described as a "left-over right" or "right-over left."

Asymptomatic hypertropia on lateral gaze is often congenital or physiological.
Cross References
Bielschowsky's sign, Bielschowsky's test; Cover tests; Heterotropia; Hypotropia

Hypoalgesia

Hypoalgesia is a decreased sensitivity to, or diminution of, pain perception in response to a normally painful stimulus.
Cross References
Analgesia

Hypoesthesia

Hypoesthesia (hypoaesthesia, hypesthesia) is decreased sensitivity to, or diminution of, sensory perception in any modality, most frequently used to describe pain (hypoalgesia) or touch.

Cross References
Anesthesia

Hypogeusia
- see AGEUSIA

Hypohidrosis
- see ANHIDROSIS

Hypokinesia
Hypokinesia is a reduction in the speed of voluntary movements, which at worst may progress to an inability to initiate voluntary movement (akinesia). Repeated apposition of finger and thumb or foot tapping may be useful in demonstrated hypokinesia of gradual onset ("fatigue").

It may often coexist with bradykinesia and hypometria, and is a feature of disorders of the basal ganglia (akinetic-rigid or parkinsonian syndromes), for example:

Parkinson's disease
Multiple system atrophy
Progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome)
Some variants of prion disease.

Cross References
Akinesia; Bradykinesia; Fatigue; Parkinsonism

Hypometria
Hypometria is a reduction in the amplitude of voluntary movements. It may be demonstrated by asking a patient to make repeated, large amplitude, opposition movements of thumb and forefinger, or tapping movements of the foot on the floor. A gradual decline in amplitude (which may be referred to as fatigability; *cf*. fatigue) denotes hypometria. Voluntary saccadic eye movements may also show a "step," as a correcting additional saccade compensates for the undershoot (hypometria) of the original movement.

Hypometria is a feature of parkinsonian syndromes, such as idiopathic Parkinson's disease.

Cross References
Akinesia; Bradykinesia; Dysmetria; Fatigue; Hypokinesia; Parkinsonism; Saccades

Hypomimia
Hypomimia, or amimia, is a deficit or absence of expression by gesture or mimicry. This is usually most obvious as a lack of facial expressive mobility ("mask-like facies"). This is a feature of frontal-subcortical disease, *e.g*., basal ganglia disease producing akinetic-rigid or parkinsonian syndromes, and frontal lobe lesions (especially of the nondominant hemisphere).

Cross References
Facial paresis; Fisher's sign; Parkinsonism

Hypophonia
Hypophonia is a quiet voice, as in hypokinetic dysarthria. It is often a feature of parkinsonian syndromes (*e.g.*, idiopathic Parkinson's disease, multiple system atrophy), and may occur early in progressive supranuclear palsy. In isolation, other causes of dysphonia may need to be considered.
Cross References
Dysarthria; Dysphonia; Parkinsonism

Hypophoria
Hypophoria is a variety of heterophoria in which there is a latent downward deviation of the visual axis of one eye. Using the cover-uncover test, this may be observed clinically as the upward movement of the eye as it is uncovered.
Cross References
Cover tests; Heterophoria; Hypophoria

Hyporeflexia
Hyporeflexia is a diminution of tendon reflexes, short of their total absence (areflexia). This may be physiological, as with the diminution of the ankle jerks with normal ageing; or pathological, most usually as a feature of peripheral lesions, such as radiculopathy or neuropathy. The latter may be axonal or demyelinating, in the latter the blunting of the reflex may be out of proportion to associated weakness or sensory loss. Although frequently characterized as a feature of the lower motor neurone syndrome, the pathology underlying hyporeflexia may occur anywhere along the monosynaptic reflex arc, including the sensory afferent fibre and dorsal root ganglion as well as the motor efferent fibre, and/or the spinal cord synapse.

Hyporeflexia may also accompany central lesions, particularly with involvement of the mesencephalic and upper pontine reticular formation. Hyporeflexia is an accompaniment of hemiballismus, and may also be noted in brainstem encephalitis (Bickerstaff's encephalitis), in which the presence of a peripheral nerve disorder is debated. Hyporeflexia is not a feature of myasthenia gravis but may occur in Lambert-Eaton myasthenic syndrome (*cf.* facilitation); it is not seen in most muscle diseases unless they are advanced.
Cross References
Age-related signs; Areflexia; Facilitation; Lower motor neurone (LMN) syndrome; Reflexes

Hyposexuality
Hyposexuality is a lack of sexual drive, interest, or activity. It may be associated with many diseases, physical or psychiatric, and/or medications which affect the central nervous system. Along with hyper-

graphia and hyperreligiosity, hyposexuality is one of the defining features of the Geschwind syndrome.

References

Benson DF. The Geschwind syndrome. *Advances in Neurology* 1991; **55**: 411-421

Pritchard PB. Hyposexuality: a complication of complex partial epilepsy. *Transactions of the American Neurological Association* 1980; **105**: 193-195

Cross References

Hypergraphia; Hyperreligiosity

Hypothermia

Hypothalamic damage, particularly in the posterior region, can lead to hypothermia (*cf.* hyperthermia) or poikilothermia (body temperature varying with ambient temperature, as in reptiles). There are many pathological causes, including tumor, trauma, infarct, hemorrhage, sarcoidosis, Wernicke's encephalopathy, fat embolism, histiocytosis X, and multiple sclerosis (rare)

A rare syndrome of paroxysmal or periodic hypothermia has been described, and labeled as diencephalic epilepsy. Nonneurological causes of hypothermia are more common, including hypothyroidism, hypopituitarism, hypoglycemia, and drug overdose.

References

Thomas DJ, Green ID. Periodic hypothermia. *BMJ* 1973; **2**: 696-697

Cross References

Hyperthermia

Hypotonia, Hypotonus

Hypotonia (hypotonus) is a diminution or loss of normal muscular tone, causing floppiness of the limbs. This is particularly associated with peripheral nerve or muscle pathology, as well as lesions of the cerebellum and certain basal ganglia disorders, such as hemiballismus-hemichorea.

Weakness preventing voluntary activity rather than a reduction in stretch reflex activity appears to be the mechanism of hypotonia.

References

Van der Meche FG, van Gijn J. Hypotonia: an erroneous clinical concept. *Brain* 1986; **109**: 1169-1178

Cross References

Ataxia; Flaccidity; Hemiballismus; Hypertonia

Hypotropia

Hypotropia is a variety of heterotropia in which there is manifest downward vertical deviation of the visual axis of one eye. Using the cover test, this manifests as upward movement of the uncovered eye. Depending on the affected eye, this finding is often described as a "left-over-right" or "right-over-left."

Cross References

Cover tests; Heterotropia; Hypertropia

I

Ice Pack Test

The ice pack test is performed by holding an ice cube, wrapped in a towel or a surgical glove, over the levator palpebrae superioris muscle of a ptotic eye for 2-10 minutes. Improvement of ptosis is said to be specific for myasthenia gravis: cold improves transmission at the neuromuscular junction (myasthenic patients often improve in cold as opposed to hot weather). This phenomenon is not observed in other causes of ptosis. A pooled analysis of several studies gave a test sensitivity of 89% and specificity of 100% with correspondingly high positive and negative likelihood ratios. The test is easy to perform and without side effects (*cf*. Tensilon test).

Whether the ice pack test is also applicable to myasthenic diplopia has yet to be determined. False positives have been documented.

References

Larner AJ. The place of the ice pack test in the diagnosis of myasthenia gravis. *International Journal of Clinical Practice* 2004; **58**: 887-888

Larner AJ, Thomas DJ. Can myasthenia gravis be diagnosed with the "ice pack test"? A cautionary note. *Postgraduate Medical Journal* 2000; **76**: 162-163

Cross References

Diplopia; Fatigue; Ptosis

Ideational Apraxia

- see APRAXIA

Ideomotor Apraxia (IMA)

- see APRAXIA

Illusion

An illusion is a misinterpretation of a perception (*cf*. delusion, hallucination). Illusions occur in normal people when they are tired, inattentive, in conditions of poor illumination, or if there is sensory impairment. They also occur in disease states, such as delirium, and psychiatric disorders (affective disorders, schizophrenia).

Examples of phenomena which may be labeled illusory include:

Visual: metamorphopsia, palinopsia, polyopia, telopsia, Pulfrich phenomenon, visual alloesthesia

Auditory: palinacusis

Vestibular: vertigo

References

Tekin S, Cummings JL. Hallucinations and related conditions. In: Heilman KM, Valenstein E (eds.). *Clinical neuropsychology* (4th edition). Oxford: OUP, 2003: 479-494

Cross References
Delirium; Delusion; Hallucination

Imitation Behavior

Imitation behavior is the reproduction by the patient of gestures (echopraxia) and/or utterances (echolalia) made by the examiner in front of the patient; these "echophenomena" are made by the patient without preliminary instructions to do so. They are consistent and have a compulsive quality to them, perhaps triggered by the equivocal nature of the situation. There may be accompanying primitive reflexes, particularly the grasp reflex, and sometimes utilization behavior.

Imitation behavior occurs with frontal lobe damage; originally mediobasal disease was thought the anatomical correlate, but more recent studies suggest upper medial and lateral frontal cortex. Certainly imitation behavior never occurs with retrorolandic cortical lesions.

A distinction has been drawn between "naïve" imitation behavior, which ceases after a direct instruction from the examiner not to imitate his/her gestures, which may be seen in some normal individuals; and "obstinate" imitation behavior which continues despite an instruction to stop; the latter is said to be exclusive to frontotemporal dementia.

References

De Renzi E, Cavalleri F, Facchini S. Imitation and utilisation behavior. *Journal of Neurology, Neurosurgery and Psychiatry* 1996; **61**: 396-400

Lhermitte F, Pillon B, Serdaru M. Human autonomy and the frontal lobes. Part I: imitation and utilization behavior: a neuropsychological study of 75 patients. *Annals of Neurology* 1986; **19**: 326-334

Shimomura T, Mori E. Obstinate imitation behavior in differentiation of frontotemporal dementia from Alzheimer's disease. *Lancet* 1998; **352**: 623-624

Cross References
Echolalia; Echopraxia; Grasp reflex; Utilization behavior

Imitation Synkinesis

- see MIRROR MOVEMENTS

Impersistence

Impersistence is an inability to sustain simple motor acts, such as conjugate gaze, eye closure, protrusion of the tongue, or keeping the mouth open. It is most commonly seen with lesions affecting the right hemisphere, especially central and frontal mesial regions, and may occur in association with left hemiplegia, neglect, anosognosia, hemianopia, and sensory loss. These patients may also manifest perseveration, echolalia and echopraxia.

Impersistence is most often observed following vascular events but may also be seen in Alzheimer's disease and frontal lobe dementias, and metabolic encephalopathies. Impersistence of tongue protrusion

and hand grip may be seen in Huntington's disease. Neuropsychologically, impersistence may be related to mechanisms of directed attention which are needed to sustain motor activity.

References

Fisher M. Left hemiplegia and motor impersistence. *Journal of Nervous and Mental Disease* 1956; **123**: 201-218

Kertesz A, Nicholson I, Cancelliere A, Kassa K, Black SE. Motor impersistence: a right-hemisphere syndrome. *Neurology* 1985; **35**: 662-666

Cross References

Anosognosia, Echolalia; Echopraxia; Hemianopia; Milkmaid's grip; Neglect; Perseveration; Trombone tongue

Inattention

- see NEGLECT

Incontinence

Urinary incontinence may result from neurological disease. Neurological pathways subserving the appropriate control of micturition encompass the medial frontal lobes, a micturition centre in the dorsal tegmentum of the pons, spinal cord pathways, Onuf's nucleus in the spinal cord segments S2-S4, the cauda equina, and the pudendal nerves. Thus the anatomical differential diagnosis of incontinence is broad. Moreover incontinence may be due to inappropriate bladder emptying or a consequence of loss of awareness of bladder fullness with secondary overflow. Other features of the history and/or examination may give useful pointers as to localization. Incontinence of neurological origin is often accompanied by other neurological signs, especially if associated with spinal cord pathology (see Myelopathy). The pontine micturition centre lies close to the medial longitudinal fasciculus and local disease may cause an internuclear ophthalmoplegia. However, other signs may be absent in disease of the frontal lobe or cauda equina.

Causes of urinary incontinence include:

- Idiopathic generalized epilepsy with tonic-clonic seizures; however, the differential diagnosis of "loss of consciousness with incontinence" also encompasses syncopal attacks with or without secondary anoxic convulsions, nonepileptic attacks, and hyperekplexia
- Frontal lobe lesions: frontal lobe dementia; normal pressure hydrocephalus
- Spinal cord pathways: urge incontinence of multiple sclerosis; loss of awareness of bladder fullness with retention of urine and overflow in tabes dorsalis
- Sacral spinal cord injury; degeneration of the sacral anterior horn cells in Onuf's nucleus (multiple system atrophy)
- Cauda equina syndrome; tethered cord syndrome (associated with spinal dysraphism)
- Pelvic floor injury.

Neurogenic incontinence may be associated with urgency, which results from associated abrupt increases in detrusor pressure (detrusor hyperreflexia); this may be helped by anticholinergic medication (*e.g.*, oxybutynin). In addition there may be incomplete bladder emptying, which is usually asymptomatic, due to detrusor sphincter dyssynergia; for post-micturition residual volumes of greater than 100 ml (assessed by in-out catheterization or ultrasonography), this is best treated by clean intermittent self-catheterization.

References

Fowler CJ. Investigation of the neurogenic bladder. In: Hughes RAC (ed.). *Neurological Investigations*. London: BMJ Publishing, 1997: 397-414

Garg BP. Approach to the patient with bladder, bowel, or sexual dysfunction and other autonomic disorders. In: Biller J (ed.). *Practical neurology* (2nd edition). Philadelphia: Lippincott Williams & Wilkins, 2002: 366-376

Cross References

Cauda equina syndrome; Dementia; Frontal lobe syndromes; Hyperekplexia; Internuclear ophthalmoplegia; Myelopathy; Seizures; Urinary retention

Intention Myoclonus

- see MYOCLONUS

Intermanual Conflict

Intermanual conflict is a behavior exhibited by an alien hand (*le main étranger*) in which it reaches across involuntarily to interfere with the voluntary activities of the contralateral (normal) hand. Diagonistic dyspraxia probably refers to the same phenomenon. The hand acts at cross purposes to the other following voluntary activity. A "compulsive grasping hand" syndrome has been described which may be related to intermanual conflict, the difference being grasping of the contralateral hand in response to voluntary movement. Intermanual conflict is more characteristic of the callosal, rather than the frontal, subtype of anterior or motor alien hand. It is most often seen in patients with corticobasal degeneration, but may also occur in association with callosal infarcts or tumors or following callosotomy.

Cross References

Alien hand, alien limb; "Compulsive grasping hand"; Diagonistic dyspraxia

Intermetamorphosis

A form of delusional misidentification in which people known to the patient are believed to exchange identities with each other (*cf*. Fregoli syndrome, in which one person can assume different physical appearance).

References

Ellis HD, Whitley J, Luaute JP. Delusional misidentification. *History of Psychiatry* 1994; **5**: 117-146

Cross References
Delusion

Internal Ophthalmoplegia
- see OPHTHALMOPARESIS, OPHTHALMOPLEGIA

Internuclear Ophthalmoplegia (INO)
Internuclear ophthalmoplegia, or medial longitudinal fasciculus syndrome, consists of ipsilateral weakness of eye adduction with contralateral nystagmus of the abducting eye (ataxic or dissociated nystagmus), but with preserved convergence. This may be obvious with pursuit eye movements, but is better seen when testing reflexive saccades or optokinetic responses when the adducting eye is seen to "lag" behind the abducting eye. INO may be asymptomatic or, rarely, may cause diplopia, oscillopsia, or a skew deviation. INO may be unilateral or bilateral. The eyes are generally aligned in primary gaze, but if there is associated exotropia this may be labeled wall-eyed monocular/bilateral internuclear ophthalmoplegia (WEMINO, WEBINO syndromes).

The most common cause of INO by far is demyelination, particularly in young patients, but other causes include cerebrovascular disease (particularly older patients), Wernicke-Korsakoff syndrome, encephalitis, trauma, and paraneoplasia.

A similar clinical picture may be observed with pathology elsewhere, hence a "false-localizing " sign and referred to as a pseudo-internuclear ophthalmoplegia (*q.v.*), especially in myasthenia gravis.

References
Zee DS. Internuclear ophthalmoplegia: pathophysiology and diagnosis. In: Büttner U, Brandt Th. *Ocular motor disorders of the brain stem*. London: Baillière Tindall, 1992: 455-470

Cross References
Diplopia; "False-localizing signs"; One-and-a-half syndrome; Optokinetic nystagmus, Optokinetic response; Oscillopsia; Pseudo-internuclear ophthalmoplegia; Saccades; Skew deviation

Intrusion
An intrusion is an inappropriate recurrence of a response (verbal, motor) to a preceding test or procedure after intervening stimuli. Intrusions are thought to reflect inattention, and may be seen in dementing disorders or delirium. These phenomena overlap to some extent with the recurrent type of perseveration.

The term intrusion is also used to describe inappropriate saccadic eye movements which interfere with macular fixation during pursuit eye movements.

References
Fuld PA, Katzman R, Davies P, Terry RD. Intrusions as a sign of Alzheimer dementia: chemical and pathological verification. *Annals of Neurology* 1982; **11**: 155-159

Cross References
Delirium; Dementia; Perseveration; Saccadic intrusion, Saccadic pursuit

Inverse Marcus Gunn Phenomenon
- see JAW WINKING; PTOSIS

Inverse Uhthoff Sign
- see UHTHOFF'S PHENOMENON

Inverted Reflexes
A phasic tendon stretch reflex is said to be inverted when the movement elicited is opposite to that normally seen, *e.g.*, extension of the elbow rather than flexion when eliciting the supinator (brachioradialis) jerk; flexion of the forearm when tapping the triceps tendon (paradoxical triceps reflex); and flexion (hamstring contraction) rather than extension of the knee when tapping the patellar tendon.

The finding of inverted reflexes may reflect dual pathology, but more usually reflects a single lesion which simultaneously affects a root or roots, interrupting the local reflex arc, and the spinal cord, damaging corticospinal (pyramidal tract) pathways which supply segments below the reflex arc. Hence, an inverted supinator jerk is indicative of a lesion at C5/6, paradoxical triceps reflex occurs with C7 lesions; and an inverted knee jerk indicates interruption of the L2/3/4 reflex arcs, with concurrent damage to pathways descending to levels below these segments.

References
Boyle RS, Shakir RA, Weir AI, McInnes A. Inverted knee jerk: a neglected localizing sign in spinal cord disease. *Journal of Neurology, Neurosurgery and Psychiatry* 1979; **42**: 1005-1007

Cross References
Reflexes

Ipsipulsion
- see LATEROPULSION

Iridoplegia
Paralysis of the iris, due to loss of pupillary reflexes. This may be partial, as in Argyll Robertson pupil or Holmes-Adie pupil, or complete as in the internal ophthalmoplegia of an oculomotor (III) nerve palsy.

Cross References
Argyll robertson pupil; Holmes-adie pupil, Holmes-adie syndrome; Oculomotor (III) nerve palsy; Ophthalmoparesis, Ophthalmoplegia; Pupillary reflexes

J

Jacksonian March
Jacksonian march is the sequential spread of a simple partial seizure to involve other body parts, for example jerking may spread from one hand up the arm, to the ipsilateral side of the face. It may culminate in a secondary generalized seizure. The pathophysiological implication is of electrical disturbance spreading through the homunculus of the motor cortex. A sensory equivalent occurs but is rare.
Cross References
Seizures

Jactitation
Jactitation is literally "throwing about," but may also imply restlessness. The term has been used in various ways: to refer to jerking or convulsion of epileptic origin; or jerking of choreic origin; or of myoclonic origin, such as "hypnagogic jactitation" (physiological myoclonus associated with falling to sleep). It may also be used to refer to the restlessness seen in acute illness, high fever, and exhaustion, though differing from the restlessness implied by akathisia. Hence, it is essentially a nonspecific term.
Cross References
Akathisia; Myoclonus; Seizures

Jamais Entendu
A sensation of unfamiliarity akin to *jamais vu* but referring to auditory experiences.

Jamais vécu
- see *JAMAIS VU*

Jamais Vu
Jamais vu (literally "never seen") and *jamais vécu* ("never lived") are complex auras of focal onset epilepsy in which there is a sensation of strangeness or unfamiliarity about visual stimuli that have in fact been previously experienced (*cf*. *déjà vu*). This is suggestive of seizure onset in the limbic system, but is not lateralizing (*cf*. *déjà vu*).
Cross References
Aura*;* Déjà vu

Jargon Aphasia
Jargon aphasia is a fluent aphasia characterized by a jumbled, unintelligible and meaningless output, with multiple paraphasias and neologisms, and sometimes echolalia (as in transcortical sensory aphasia). There may be a pressure of speech (logorrhea).

There is debate as to whether jargon aphasia is simply a primary Wernicke/posterior/ sensory type of aphasia with failure to self-monitor speech output, or whether additional deficits (*e.g.*, pure word deafness, intellectual impairment) are also required. Others suggest that jargon aphasia represents aphasia and anosognosia, leading to confabulation and reduplicative paramnesia.

References

Hillis AE, Boatman DB, Hart J, Gordon B. Making sense out of jargon. A neurolinguistic and computational account of jargon aphasia. *Neurology* 1999; **53**: 1813-1824

Kinsbourne M, Warrington EK. Jargon aphasia. *Neuropsychologia* 1963; **1**: 27-37

Cross References

Anosognosia; Aphasia; Confabulation; Echolalia; Logorrhea; Pure word deafness; Reduplicative paramnesia; Transcortical aphasias; Wernicke's aphasia

Jaw Jerk

The jaw jerk, or masseter reflex, is contraction of the masseter and temporalis muscles in response to a tap on the jaw with the mouth held slightly open. Both the afferent and efferent limbs of the arc run in the mandibular division of the trigeminal (V) nerve, connecting centrally with the mesencephalic (motor) nucleus of the trigeminal nerve. The reflex is highly reproducible; there is a linear correlation between age and reflex latency, and a negative correlation between age and reflex amplitude.

Interruption of the reflex arc leads to a diminished or absent jaw jerk as in bulbar palsy (although an absent jaw jerk may be a normal finding, particularly in the elderly). Bilateral supranuclear lesions cause a brisk jaw jerk, as in pseudobulbar palsy (*e.g.*, in motor neurone disease).

References

Fitzek S, Fitzek C, Hopf HC. Normative values of the masseter reflex (myotatic masseter reflex). *Journal of Neurology* 2000; **247 (suppl3)**: 176-177 (abstract 724)

Cross References

Age-related signs; Bulbar palsy; Pseudobulbar palsy; Reflexes

Jaw Winking

Jaw winking, also known as the Marcus Gunn phenomenon, is widening of a congenital ptosis when a patient is chewing, swallowing, or opening the jaw (*i.e.*, a trigemino-oculomotor synkinesis). It is believed to result from aberrant innervation of the pterygoid muscles and levator palpebrae superioris.

Eyelid closure on jaw movement or opening of the mouth, inverse Marcus Gunn phenomenon, is also described, as the Marin-Amat syndrome, thought to be due to aberrant facial (VII) nerve regeneration.

References

Rana PVS, Wadia RS. The Marin-Amat syndrome: an unusual facial synkinesia. *Journal of Neurology, Neurosurgery and Psychiatry* 1985; **48**: 939-941

Cross References
Ptosis; Synkinesia, Synkinesis

Jendrassik's Maneuver

Jendrassik's maneuver is used to enhance or bring out absent or depressed tendon (phasic stretch) reflexes by isometric contraction of distant muscle groups, *e.g.*, clenching teeth, or making a fist, interlocking fingers and pulling the hands against one another. If previously absent reflexes are then elicited, this may be denoted +/–. Co-contraction increases the gain in the monosynaptic reflex arc, as distinct from facilitation or post-tetanic potentiation which is seen in Lambert-Eaton myasthenic syndrome following tetanic contraction of muscles involved in the reflex.

References

Jendrassik E. Ueber allgemeine Localisation der Reflexe. *Deutsche Archiv fur Klinische Medicin* 1894; **52**: 569-600

Delwaide P, Toulouse P. Facilitation of monosynaptic reflexes by voluntary contractions of muscle in remote parts of the body. Mechanisms involved in the Jendrassik maneuver. *Brain* 1981; **104**: 701-709

Cross References
Facilitation; Reflexes

Jitteriness

Jitteriness implies an exaggerated startle response, reflecting CNS overactivity. This may be confused in neonates with clonic seizures, but in the former there is stimulus sensitivity and an absence of associated ocular movements. However, both may occur in hypoxic-ischemic or metabolic encephalopathies or with drug withdrawal.

Cross References
Seizures

Joint Position Sense

- see PROPRIOCEPTION

Jugular Foramen Syndrome

The glossopharyngeal (IX), vagus (X), and accessory (XI) cranial nerves may be damaged by lesions at or around the jugular foramen, producing a jugular foramen (or Vernet's) syndrome. This produces:

- Dysphagia, dysphonia, palatal droop, impaired gag reflex; ipsilateral reduced taste sensation on the posterior one third of the tongue, and anesthesia of the posterior one third of the tongue, soft palate, pharynx, larynx and uvula, due to glossopharyngeal and vagus nerve involvement.
- Ipsilateral weakness and atrophy of sternocleidomastoid and trapezius due to accessory nerve involvement (atrophy may be the more evident, hence the importance of palpating the muscle bellies).

Recognized causes of the jugular foramen syndrome include:
Skull base trauma/fracture
Glomus jugulare tumor
Inflammatory/infective collection at the skull base
Ischemia.

The differential diagnosis includes retropharyngeal or retroparotid space occupying lesions, which may in addition involve the hypoglossal nerve (XII; Collet-Sicard syndrome) and the sympathetic chain with or without the facial nerve (VII; Villaret's syndrome).

Cross References
Dysphagia; Dysphonia; Gag reflex

Junctional Scotoma, Junctional Scotoma of Traquair

Despite the similarity of these terms, they are used to refer to different types of scotoma:

- *Junctional scotoma*:
  Unilateral central scotoma with contralateral superior temporal defect, seen with lesions at the anterior angle of the chiasm; this is said to damage the ipsilateral optic nerve plus the crossing loop of fibers (Wilbrand's knee) originating from the inferonasal portion of the contralateral eye (it may be noted that some authors have questioned whether such a loop in fact exists).
- *Junctional scotoma of Traquair*:
  A monocular temporal scotoma, sometimes even hemianopia, seen with optic nerve involvement sufficiently close to the chiasm to involve only ipsilateral crossing nasal axons, which subserve the temporal visual field, but sparing nasal axons crossing from the contralateral eye.

References
Larner AJ. A developing visual field defect. *Postgraduate Medical Journal* 2002; **78**: 106, 112-113

Cross References
Scotoma; Visual field defects

K

Kayser-Fleischer Rings

Kayser-Fleischer rings are deposits of copper, seen as a brownish discoloration, in Descemet's membrane. Although often visible to the naked eye (difficult in people with a brown iris), they are best seen with slit-lamp examination. Since they are a highly reliable sign of intracerebral copper deposition in Wilson's disease (hepatolenticular degeneration), any patient suspected of this diagnosis (*i.e.*, with parkinsonism or dystonia presenting before age 50 years) should have a slit-lamp examination (as well as blood copper and ceruloplasmin, and urinary copper, measurements). Very occasionally cases of neurological Wilson's disease without Kayser-Fleischer rings have been reported.

References

Finelli PF. Kayser-Fleischer ring: Hepatolenticular degeneration (Wilson's disease). *Neurology* 1995; **45**: 1261-1262

Cross References

Dystonia; Parkinsonism

Kernig's Sign

Kernig's sign is pain in the lower back (and also sometimes the neck) and resistance to movement with passive extension of the knee on the flexed thigh in a recumbent patient. It is indicative of meningeal mechanosensitivity due to inflammation, either infective (meningitis) or chemical (subarachnoid hemorrhage), in which case it may coexist with nuchal rigidity and Brudzinski's (neck) sign. If unilateral it may indicate irritation of the lumbosacral nerve roots from a ruptured intervertebral disc (in which case Lasègue's sign may also be present).

References

Pearce JMS. Kernig and Brudzinski. In: Pearce JMS. *Fragments of neurological history*. London: Imperial College Press, 2003: 365-366

Cross References

Brudzinski's (neck) sign; Lasègue's sign; Nuchal rigidity

Kernohan's Notch Syndrome

Raised intracranial pressure as a result of an expanding supratentorial lesion (*e.g.*, tumor, subdural hematoma) may cause herniation of brain tissue through the tentorium into the subtentorial space, putting pressure on the midbrain. If the midbrain is shifted against the contralateral margin (free edge) of the tentorium, the cerebral peduncle on that side may be compressed, resulting in a hemiparesis which is ipsilateral to the supratentorial lesion (and hence may be considered "false-localizing").

There may also be an oculomotor nerve palsy ipsilateral to the lesion, which may be partial (unilateral pupil dilatation).

References
Cohen AR, Wilson J. Magnetic resonance imaging of Kernohan's notch. *Neurosurgery* 1990; **27**: 205-207
Kernohan JW, Woltman HW. Incisura of the crus due to contralateral brain tumor. *Arch Neurol Psychiatry* 1929; **21**: 274-287
Kole MK, Hysell SE. MRI correlate of Kernohan's notch. *Neurology* 2000; **55**: 1751
Cross References
"False-localizing signs"; Hemiparesis; Hutchinson's pupil

Kinesis Paradoxica
Kinesis paradoxica is the brief but remarkably rapid and effective movement sometimes observed in patients with Parkinson's disease or post-encephalitic parkinsonism, despite the poverty and slowness of spontaneous movement (akinesia, hypokinesia; bradykinesia) seen in these conditions. It often occurs in response to alarm, excitement or emotion (*e.g.*, in response to a genuinely funny joke).
Cross References
Akinesia; Bradykinesia; Hypokinesia; Parkinsonism

Klazomania
Klazomania was the term applied to the motor and vocal tics seen as a sequel to encephalitis lethargica (von Economo's disease), along with parkinsonism and oculogyric crises. This observation helped to promote the idea that tics were due to neurological disease rather than being psychogenic, for example in Gilles de la Tourette syndrome.
References
Wohlfart G, Ingvar DH, Hellberg AM. Compulsory shouting (Benedek's "klazomania") associated with oculogyric spasm in chronic epidemic encephalitis. *Acta Psychiatrica Scandinavica* 1961; **36**: 369-377
Cross References
Coprolalia; Echolalia; Parakinesia, Parakinesis; Tic

Kleptomania
Kleptomania, a morbid impulse to steal, has been related to the obsessive-compulsive spectrum of behaviors in patients with frontal lobe dysfunction.
References
Kozian R, Otto FG. Treatment of a patient with kleptomania and frontal lobe dysfunction. *Journal of Neurology, Neurosurgery and Psychiatry* 2001; **70**: 279 (abstract)
Cross References
Frontal lobe syndromes

Klüver-Bucy Syndrome
The Klüver-Bucy syndrome consists of a variety of neurobehavioral changes, originally observed following bilateral temporal lobectomy (especially anterior tip) in monkeys, but subsequently described

in man. The characteristic features, some or all of which may be present, are:

Visual agnosia (*e.g.*, misrecognition of others)
Hyperorality
Hyperphagia, binge eating
Hypermetamorphosis
Hypersexuality
Emotional changes: apathy; loss of fear, rage reactions

Clinical causes of the Klüver-Bucy syndrome include:

Sequel of bilateral temporal lobectomy
Post-ictal phenomenon in a patient with a previous unilateral temporal lobectomy
Sequel to minor head trauma; subdural hematoma
Tumor
Meningoencephalitis
Pick's disease
Alzheimer's disease: especially hyperorality and hyperphagia, but it is rare to have all features

References

Anson JA, Kuhlman DT. Post-ictal Klüver-Bucy syndrome after temporal lobectomy. *Journal of Neurology, Neurosurgery and Psychiatry* 1993; **56**: 311-313

Klüver H, Bucy P. Preliminary analysis of functions of the temporal lobes in monkeys. *Archives of Neurology and Psychiatry* 1939; **42**: 979-1000.

Cross References

Apathy; Hypermetamorphosis; Hyperorality; Hyperphagia; Hypersexuality; Visual agnosia

Knee Tremor

A characteristic tremor of the patellae, sometimes known as knee bobbing, juddering, or quivering, may be seen in primary orthostatic tremor (POT; "shaky legs syndrome"). It is due to rapid rhythmic contractions of the leg muscles on standing, which dampen or subside on walking, leaning against a wall, or being lifted off the ground, with disappearance of the knee tremor; hence this is a task-specific tremor. Auscultation with the diaphragm of a stethoscope over the lower limb muscles reveals a regular thumping sound, likened to the sound of a distant helicopter. EMG studies show pathognomonic synchronous activity in the leg muscles with a frequency of 14-18Hz, thought to be generated by a central oscillator (peripheral loading does not alter tremor frequency).

A number of drugs may be helpful in POT, including phenobarbitone, primidone, clonazepam, and levodopa, but not propranolol (*cf.* essential tremor).

References

Heilman KM. Orthostatic tremor. *Archives of Neurology* 1984; **41**: 880-881

Brown P. New clinical sign for orthostatic tremor. *Lancet* 1995; **346**: 306-307

Cross References
Tremor

Körber-Salus-Elschnig Syndrome

- see NYSTAGMUS

Kyphoscoliosis

Kyphoscoliosis is twisting of the spinal column in both the anteroposterior (kyphosis) and lateral (scoliosis) planes. Although such deformity is often primary or idiopathic, thus falling within the orthopedic field of expertise, it may also be a consequence of neurological disease which causes weakness of paraspinal muscles.

Recognized neurological associations of kyphoscoliosis and scoliosis include:

Chiari I malformation, syringomyelia
Myelopathy (cause or effect? Skeletal disease, such as achondroplasia, is more likely to be associated with myelopathy than idiopathic scoliosis)
Cerebral palsy
Friedreich's ataxia
Neurofibromatosis
Hereditary motor and sensory neuropathies
Spinal muscular atrophies
Myopathies, *e.g.*, Duchenne muscular dystrophy

Stiff person syndrome may produce a characteristic hyperlordotic spine. Some degree of scoliosis occurs in virtually all patients suffering from paralytic poliomyelitis before the pubertal growth spurt.

Cross References
Camptocormia; Stiffness

L

Lagophthalmos

Lagophthalmos is an inability to close the eyelid in a peripheral facial (VII) nerve palsy, with partial opening of the palpebral fissure. A similar phenomenon may be observed with aberrant regeneration of the oculomotor nerve, thought to be due to co-contraction of the levator palpebrae superioris and superior rectus muscles during Bell's phenomenon.

Cross References

Bell's palsy; Bell's phenomenon; Facial paresis

Lambert's Sign

Lambert's sign is gradual increase in force over a few seconds when a patient with Lambert-Eaton myasthenic syndrome is asked to squeeze the examiner's hand as hard as possible, reflecting increased power with sustained exercise.

Cross References

Facilitation

Lasègue's Sign

Lasègue's sign is pain along the course of the sciatic nerve induced by stretching of the nerve, achieved by flexing the thigh at the hip while the leg is extended at the knee ("straight leg raising"). This is similar to the maneuver used in Kernig's sign (gradual extension of knee with thigh flexed at hip). Both indicate irritation of the lower lumbosacral nerve roots and/or meninges. The test may be positive with disc protrusion, intraspinal tumor, or inflammatory radiculopathy. Pain may be aggravated or elicited sooner using Bragard's test, dorsiflexing the foot while raising the leg thus increasing sciatic nerve stretch, or Neri's test, flexing the neck to bring the head on to the chest, indicating dural irritation.

A positive straight leg raising test is reported to be a sensitive indicator of nerve root irritation, proving positive in 95% of those with surgically proven disc herniation. The specificity may be somewhat lower.

Various modifications of Lasègue's sign have been described. crossed straight leg raising, when the complaint of pain on the affected side occurs with raising of the contralateral leg, is said to be less sensitive but highly specific. Femoral stretch test (*q.v.*) or "reverse straight leg raising" may detect L3 root or femoral nerve irritation.

References

Pearce JMS. JJ Forst and Lasègue's sign. In: Pearce JMS. *Fragments of neurological history*. London: Imperial College Press, 2003: 362-364

Cross References

Femoral stretch test; Kernig's sign

Lateral Medullary Syndrome

The lateral medullary syndrome (or Wallenberg's syndrome, after the neurologist who described it in 1895) results from damage (usually infarction) of the posterolateral medulla with or without involvement of the inferior cerebellum, producing the following clinical features:

- Nausea, vomiting, vertigo, oscillopsia (involvement of vestibular nuclei)
- Contralateral hypoalgesia, thermoanesthesia (spinothalamic tract)
- Ipsilateral facial hypoalgesia, thermoanesthesia, + facial pain (trigeminal spinal nucleus and tract)
- Horner's syndrome (descending sympathetic tract), +/– ipsilateral hypohidrosis of the body
- Ipsilateral ataxia of limbs (olivocerebellar/spinocerebellar fibers, inferior cerebellum)
- Dysphagia, dysphonia, impaired gag reflex
- +/– eye movement disorders, including nystagmus, abnormalities of ocular alignment (skew deviation, ocular tilt reaction, environmental tilt), smooth pursuit and gaze holding, and saccades (lateropulsion)
- +/– hiccups (singultus); loss of sneezing.

Infarction due to vertebral artery occlusion (occasionally posterior inferior cerebellar artery) or dissection is the most common cause of lateral medullary syndrome, although tumor, demyelination, and trauma are also recognized causes.

References

Fisher CM, Karnes W, Kubik C. Lateral medullary infarction: the pattern of vascular occlusion. *Journal of Neuropathology and Experimental Neurology* 1961; **20**: 103-113

Pearce JMS. Wallenberg's syndrome. In: Pearce JMS. *Fragments of neurological history*. London: Imperial College Press, 2003: 233-236

Sacco RL, Freddo L, Bello JA, Odel JG, Onesti ST, Mohr JP. Wallenberg's lateral medullary syndrome. Clinical-magnetic resonance imaging correlations. *Archives of Neurology* 1993; **50**: 609-614

Cross References

Anesthesia; Dysphagia; Dysphonia; Environmental tilt; Gag reflex; Hemiataxia; Hiccup; Horner's syndrome; Hypoalgesia; Hypohidrosis; Medial medullary syndrome; Nystagmus; Ocular tilt reaction; Oscillopsia; Saccades; Skew deviation; Sneezing; Vertigo

Lateral Rectus Palsy

- see ABDUCENS (VI) NERVE PALSY

Laterocollis

Laterocollis is a lateral head tilt; this may be seen in 10-15% of patients with torticollis.

Cross References

Torticollis

Lateropulsion

Lateropulsion or ipsipulsion is literally pulling to one side. The term may be used to describe ipsilateral axial lateropulsion after cerebellar infarcts preventing patients from standing upright causing them to lean to toward the opposite side. Lateral medullary syndrome may be associated with lateropulsion of the eye toward the involved medulla, and there may also be lateropulsion of saccadic eye movements.

Laughter

- see AUTOMATISM; PATHOLOGICAL CRYING, PATHOLOGICAL LAUGHTER

Lazarus Sign

Various spontaneous and reflex movements are described in brain death, the most dramatic of which has been labeled the Lazarus sign, after Lazarus, raised from the dead by Christ (John 11:1-44). This spinal reflex manifests as flexion of the arms at the elbow, adduction of the shoulders, lifting of the arms, dystonic posturing of the hands and crossing of the hands.

References

Saposnik G, Bueri JA, Mauriño J, Saizar R, Garretto NS. Spontaneous and reflex movements in brain death. *Neurology* 2000; **54**: 221-223

Bueri JA, Saposnik G, Mauriño J, Saizar R, Garretto NS. Lazarus' sign in brain death. *Movement Disorders* 2000; **15**: 583-586

Leadpipe Rigidity

- see RIGIDITY

Levator Inhibition

- see EYELID APRAXIA

Levitation

Spontaneous levitation may be displayed by an alien limb, more usually an arm than a leg, indicative of parietal lobe pathology. It is most often seen in corticobasal (ganglionic) degeneration, but a few cases with pathologically confirmed progressive supranuclear palsy have been reported.

References

Brunt ER, van Weerden TW, Pruim J, Lakke JW. Unique myoclonic pattern in corticobasal degeneration. *Movement Disorders* 1995; **10**: 132-142

Cross References

Alien hand, Alien limb

Lhermitte's Sign

Lhermitte's sign, or the "barber's chair syndrome," is a painless but unpleasant tingling or electric shock-like sensation in the back and spreading instantaneously down the arms and legs following neck flex-

ion (active or passive). It is associated with pathology within the cervical spinal cord. Although most commonly encountered (and originally described in) demyelination, it is not pathognomonic of this condition, and has been described with other local pathologies, such as:

subacute combined degeneration of the cord (vitamin B_{12} deficiency); nitrous oxide (N_2O) exposure
traumatic or compressive cervical myelopathy (*e.g.*, cervical spondylotic myelopathy)
epidural/subdural/intraparenchymal tumor
radiation myelitis
pyridoxine toxicity
inflammation, *e.g.*, systemic lupus erythematosus, Behçet's disease
cervical herpes zoster myelitis
cavernous angioma of the cervical cord

Pathophysiologically, this movement-induced symptom may reflect the exquisite mechanosensitivity of axons which are demyelinated, or damaged in some other way.

A "motor equivalent" of Lhermitte's sign, McArdle's sign, has been described, as has "reverse Lhermitte's sign," a label applied either to the aforementioned symptoms occurring on neck extension, or in which neck flexion induces electrical shock-like sensation traveling from the feet upward.

References

Lhermitte J, Bollack J, Nicolas M. Les douleurs à type de décharge electrique consécutives à la flexion céphalique dans la sclérose en plaques: un case de forme sensitive de la sclérose multiple. *Revue Neurologique* 1924; **39**: 56-62

Pearce JMS. Lhermitte's sign. In: Pearce JMS. *Fragments of neurological history*. London: Imperial College Press, 2003: 367-369

Smith KJ. Conduction properties of central demyelinated axons: the generation of symptoms in demyelinating disease. In: Bostock H, Kirkwood PA, Pullen AH (eds.). *The neurobiology of disease: contributions from neuroscience to clinical neurology*. Cambridge: CUP, 1996: 95-117

Cross References

Mcardle's sign; Myelopathy

Lid Lag

Lid lag is present if a band of sclera is visible between the upper eyelid and the corneal limbus on attempted downgaze (*cf.* lid retraction), seen for example in thyroid eye disease (von Graefe's sign), progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome), and Guillain-Barré syndrome.

Cross References

Lid retraction; von Graefe's sign

Lid Retraction

Lid retraction is present if a band of sclera is visible between the upper eyelid and the corneal limbus in the primary position (*cf.* lid lag). This

should be distinguished from contralateral ptosis. Recognized causes of lid retraction include:

- Overactivity of levator palpebrae superioris:
  - Dorsal mesencephalic lesion (Collier's sign)
  - Opposite to unilateral ptosis, *e.g.*, in myasthenia gravis; retracted lid may fall when ptotic lid raised; frontalis overactivity usually evident
  - Paradoxical lid retraction with jaw movement (jaw winking, Marcus Gunn phenomenon)
- Overactivity of Müller's muscle:
  - irritative oculosympathetic lesions (Claude-Bernard syndrome)
- Contracture of the levator muscle:
  - Hyperthyroidism, Graves' ophthalmopathy (Dalrymple's sign): may be associated lid lag
  - Myotonic syndromes
  - Aberrant oculomotor (III) nerve regeneration (pseudo-von Graefe's sign)
- Cicatricial retraction of the lid, *e.g.*, following trauma
- Hepatic disease (Summerskill's sign)
- Guillain-Barré syndrome.

Lower lid retraction may be congenital, or a sign of proptosis. Ectropion may also be seen with lower lid tumor or chalazion, trauma with scarring, and ageing.

Cross References

Collier's sign; Contracture; Dalrymple's sign; Jaw winking; Lid lag; Proptosis; Pseudo-von Graefe's sign; Ptosis; Stellwag's sign; Setting sun sign

Light-Near (Pupillary) Dissociation (LND)

Light-near pupillary dissociation refers to the loss of pupillary light reflexes, while the convergence-accommodation reaction is preserved (see Pupillary Reflexes). This dissociation may be seen in a variety of clinical circumstances:

- Argyll Robertson pupil: small irregular pupils with reduced reaction to light, typically seen in neurosyphilis; the absence of miosis and/or pupillary irregularity has been referred to as pseudo-Argyll Robertson pupil, which may occur with sarcoidosis, diabetes, and aberrant regeneration of the oculomotor (III) nerve
- Holmes-Adie pupil: dilated pupil showing strong but slow reaction to accommodation but minimal reaction to light (tonic > phasic)
- Parinaud's syndrome (dorsal rostral midbrain syndrome): due to a lesion at the level of the posterior commissure, and characterized by vertical gaze palsy, lid retraction (Collier's sign) or ptosis, and large regular pupils responding to accommodation but not light.

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; Collier's sign; Holmes-adie pupil, Holmes-adie syndrome; Lid retraction; Parinaud's syndrome; Pseudo-argyll Robertson pupil; Pupillary reflexes

Light Reflex

- see PUPILLARY REFLEXES

Locked-in Syndrome

The locked-in syndrome results from de-efferentation, such that a patient is awake, self-ventilating and alert, but unable to speak or move; vertical eye movements and blinking are usually preserved, affording a channel for simple (yes/no) communication.

The most common cause of the locked-in syndrome is basilar artery thrombosis causing ventral pontine infarction (both pathological laughter and pathological crying have on occasion been reported to herald this event). Other pathologies include pontine hemorrhage and central pontine myelinolysis. Bilateral ventral midbrain and internal capsule infarcts can produce a similar picture.

The locked-in syndrome may be mistaken for abulia, akinetic mutism, coma, and catatonia.

References

Bauby J-D. *The diving-bell and the butterfly*. London: Fourth Estate, 1997

Feldman MH. Physiological observations in a chronic case of locked in syndrome. *Neurology* 1971; **21**: 459-478

Cross References

Abulia; Akinetic mutism; Blinking; Catatonia; Coma; Pathological crying, Pathological laughter

Lockjaw

- see TRISMUS

Logoclonia

Logoclonia is the tendency for a patient to repeat the final syllable of a word when speaking; hence it is one of the reiterative speech disorders (*cf*. echolalia, palilalia). It may be described as the festinating repetition of individual phonemes. Logoclonia has also been used to describe continuous perseveration.

Logoclonia is an indicator of bilateral brain injury, usually involving subcortical structures, and may be seen in the late stages of dementia of Alzheimer type (but not in semantic dementia).

Cross References

Echolalia; Festination, Festinant gait; Palilalia; Perseveration

Logopenia

Logopenia is a reduced rate of language production, due especially to word finding pauses, but with relatively preserved phrase length and syntactically complete language, seen in aphasic syndromes, such as primary progressive aphasia.

Cross References

Aphasia

Logorrhea

Logorrhea is literally a flow of speech, or pressure of speech, denoting an excessive verbal output, an abnormal number of words produced during each utterance. Content is often irrelevant, disconnected and difficult to interpret. The term may be used of the output in the Wernicke/posterior type of aphasia, or of an output which superficially resembles Wernicke aphasia but in which syntax and morphology are intact, rhythm and articulation are usually normal, and paraphasias and neologisms are few. Moreover comprehension is better than anticipated in the Wernicke type of aphasia. Patients may be unaware of their impaired output (anosognosia) due to a failure of self-monitoring.

Logorrhea may be observed in subcortical (thalamic) aphasia, usually following recovery from lesions (usually hemorrhage) to the anterolateral nuclei. Similar speech output may be observed in psychiatric disorders, such as mania and schizophrenia.

References

Damasio AR. Aphasia. *New England Journal of Medicine* 1992; **326**: 531-539

Cross References

Aphasia; Delirium; Echolalia; Jargon aphasia; Wernicke's aphasia

Long Tract Signs

- see UPPER MOTOR NEURONE (UMN) SYNDROME

"Looking Glass Syndrome"

- see MIRROR AGNOSIA

Lower Motor Neurone (LMN) Syndrome

A lower motor neurone (LMN) syndrome constitutes a constellation of motor signs resulting from damage to lower motor neurone pathways, *i.e.*, from anterior horn cell distally, encompassing the motor roots, nerve plexuses, peripheral nerves, and neuromuscular junction. Following the standard order of neurological examination of the motor system, the signs include:

- *Appearance*:
  muscle wasting; fasciculations (or "fibrillations") may be observed or induced, particularly if the pathology is at the level of the anterior horn cell

- *Tone*:
  reduced tone (flaccidity, hypotonus), although this may simply reflect weakness
- *Power*:
  weakness, often marked; depending on the precise pathological process, weakness often affects both flexor and extensor muscles equally (although this is not always the case)
- *Coordination*:
  depending on the degree of weakness, it may not be possible to comment on the integrity or otherwise of coordination in LMN syndromes; in a pure LMN syndrome coordination will be normal
- *Reflexes*:
  depressed (hyporeflexia) or absent (areflexia); plantar responses are flexor.

It is often possible to draw a clinical distinction between motor symptoms resulting from lower or upper motor neurone pathology and hence to formulate a differential diagnosis and direct investigations accordingly. Sensory features may also be present in LMN syndromes if the pathology affects sensory as well as motor roots, or both motor and sensory fibers in peripheral nerves.

Cross References

Areflexia; Fasciculation; Fibrillation; Flaccidity; Hyporeflexia; Hypotonia, Hypotonus; Neuropathy; Reflexes; Upper motor neurone (UMN) syndrome; Weakness

M

Macrographia
Macrographia is abnormally large handwriting. It may be seen in cerebellar disease, possibly as a reflection of the kinetic tremor and/or the impaired checking response seen therein (*cf*. micrographia).
Cross References
Micrographia; Tremor

Macropsia
- see METAMORPHOPSIA

Macrosomatognosia
- see "ALICE IN WONDERLAND" SYNDROME

Macro-Square-Wave Jerks
- see SQUARE-WAVE JERKS

Macula Sparing, Macula Splitting
Macula sparing is a feature of an homonymous hemianopia in which central vision is intact, due to damage confined to the occipital cortex without involving the occipital pole. This may occur because anastomoses between the middle and posterior cerebral arteries maintain that part of area 17 necessary for central vision after occlusion of the posterior cerebral artery.

Cortical blindness due to bilateral (sequential or simultaneous) posterior cerebral artery occlusion may leave a small central field around the fixation point intact, also known as macula sparing.

Macula splitting, an homonymous hemianopia which cuts through the vertical meridian of the macula, occurs with lesions of the optic radiation.
Cross References
Cortical blindness; Hemianopia; Visual field defects

Maculopathy
Maculopathy is any process affecting the macula, with changes observable on ophthalmoscopy. These processes may produce a central or ring scotoma and visual failure. Common causes include:

- Diabetes mellitus: edema and hard exudates at the macula are a common cause of visual impairment, especially in noninsulin dependent diabetes mellitus.
- Hypertension: abnormal vascular permeability around the fovea may produce a macular star.
- Drug-induced: *e.g*., "bull's-eye" maculopathy of chloroquine.

- "Cherry red spot at the macula": this appearance may occur in sialidosis ("cherry red spot-myoclonus syndrome") and gangliosidoses (*e.g.*, Tay-Sachs disease).

Cross References
Cherry red spot at the macula; Retinopathy; Scotoma; Visual field defects

Magnetic Movements
Movements may be described as magnetic in varying contexts:

- the following or tracking movements of an alien hand in corticobasal degeneration, reaching out to touch or grasp the examiner's hand or clothing, as in forced groping;
- in a hesitant gait (ignition failure), with seeming inability to lift the feet ("stuck to the floor") in gait apraxia.

Cross References
Alien hand, Alien limb; Forced groping; Gait apraxia; Grasp reflex

Main d'Accoucheur
Main d'accoucheur, or carpopedal spasm, is a posture of the hand with wrist flexion in which the muscles are rigid and painful. *Main d'accoucheur* is so called because of its resemblance to the posture of the hand adopted for the manual delivery of a baby ("obstetrical hand").

This tetanic posture may develop in acute hypocalcemia (induced by hyperventilation, for instance) or hypomagnesemia, and reflects muscle hyperexcitability. Development of *main d'accoucheur* within 4 minutes of inflation of a sphygmomanometer cuff above arterial pressure (Trousseau's sign) indicates latent tetany. Mechanosensitivity of nerves may also be present elsewhere (Chvostek's sign).

Cross References
Chvostek's sign; Trousseau's sign

Main en Griffe
- see CLAW HAND

Main Étranger
- see ALIEN HAND, ALIEN LIMB

Main Succulente
Main succulente refers to a swollen hand with thickened subcutaneous tissues, hyperkeratosis and cyanosis, trophic changes which may be observed in an analgesic hand, *e.g.*, in syringomyelia.

Cross References
Charcot joint

"Man-in-a-Barrel"
"Man-in-a-barrel" is a clinical syndrome of brachial diplegia with preserved muscle strength in the legs.

This most usually occurs as a result of bilateral border zone infarcts in the territories between the anterior and middle cerebral arteries ("watershed infarction"). This may be as a consequence of cerebral hypoperfusion (*e.g.*, during cardiac arrest, cardiac surgery), in which case the prognosis is poor. The clinical picture has also been reported with cerebral metastases. Acute central cervical cord lesions may also produce a "man-in-a-barrel" syndrome, for example after severe hyperextension injury, or after unilateral vertebral artery dissection causing anterior cervical spinal cord infarction. This may follow a transient quadriplegia, and considerable recovery is possible. A neurogenic main-in-a-barrel syndrome has been reported ("flail arm syndrome"), which is a variant of motor neurone disease.

References

Mohr JP. Distal field infarction. *Neurology* 1969; **19**: 279 (abstract GS7)

Cross References

Flail arm; Quadriparesis, Quadriplegia

Marche à Petit Pas

Marche à petit pas is a disorder of gait characterized by impairments of balance, gait ignition, and locomotion. Particularly there is shortened stride (literally *marche à petit pas*) and a variably wide base. This gait disorder is often associated with dementia, frontal release signs, and urinary incontinence, and sometimes with apraxia, parkinsonism, and pyramidal signs. This constellation of clinical signs reflects underlying pathology in the frontal lobe and subjacent white matter, most usually of vascular origin. Modern clinical classifications of gait disorders have subsumed *marche à petit pas* into the category of frontal gait disorder.

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

Apraxia; Dementia; Frontal release signs; Parkinsonism

Marcus Gunn Phenomenon

- see JAW WINKING

Marcus Gunn Pupil, Marcus Gunn Sign

The Marcus Gunn pupil or sign, first described in 1902, is the adaptation of the pupillary light reflex to persistent light stimulation, that is, a dilatation of the pupil is observed with continuing stimulation with incident light ("dynamic anisocoria"). This is indicative of an afferent pathway defect, such as retrobulbar neuritis. The swinging flashlight sign or test (*q.v.*) may be used to demonstrate this by comparing direct and consensual pupillary light reflexes in one eye. Normally the responses are equal but in the presence of an afferent conduction defect an inequality is manifest as pupillary dilatation.

References

Pearce JMS. The Marcus Gunn pupil. In: Pearce JMS. *Fragments of neurological history*. London: Imperial College Press, 2003: 245-247.

Cross References

Pupillary reflexes; Relative afferent pupillary defect (RAPD); Swinging flashlight sign

Mask-like Facies

The poverty of spontaneous facial expression, hypomimia, seen in extrapyramidal disorders, such as idiopathic Parkinson's disease, is sometimes described as mask-like.

Cross References

Hypomimia; Parkinsonism

Masseter Hypertrophy

Masseter hypertrophy, either unilateral or bilateral, may occur in individuals prone to bruxism. A familial syndrome of hypertrophy of the masseter muscles has been described.

References

Matinelli P, Fabbri R, Gabellini AS. Familial hypertrophy of the masseter muscles. *Journal of Neurology* 1987; **234**: 251-253

Cross References

Bruxism

Masseter Reflex

- see JAW JERK

Masticatory Claudication

Pain in the muscles of mastication with chewing may be a sign, along with headache, of giant cell (temporal) arteritis.

McArdle's Sign

McArdle's sign is the combination of reduced lower limb strength, increased lower limb stiffness and impaired mobility following neck flexion. The difference may best be appreciated by comparing leg strength (*e.g.*, hip flexion) with the neck fully extended and fully flexed.

The sign was initially described in multiple sclerosis but may occur in other myelopathies affecting the cord at any point between the foramen magnum and the lower thoracic region. The mechanism is presumed to be stretch-induced conduction block, due to demyelinated plaques or other pathologies, in the corticospinal tracts. McArdle's sign may be envisaged as the motor equivalent of Lhermitte's sign.

References

McArdle MJ. McArdle's sign in multiple sclerosis. *Journal of Neurology, Neurosurgery and Psychiatry* 1988; **51**: 1110

O'Neill JH, Mills KR, Murray NMF. McArdle's sign in multiple sclerosis. *Journal of Neurology, Neurosurgery and Psychiatry* 1987; **50**: 1691-1693

Cross References
Lhermitte's sign; Myelopathy

Medial Medullary Syndrome

The medial medullary syndrome, or Dejerine's anterior bulbar syndrome, results from damage to the medial medulla, most usually infarction as a consequence of anterior spinal artery or vertebral artery occlusion. The clinical picture is of:

- Ipsilateral tongue paresis and atrophy, fasciculations (hypoglossal nerve involvement)
- Contralateral hemiplegia with sparing of the face (pyramid)
- Contralateral loss of position and vibration sense (medial lemniscus) with pain and temperature sensation spared
- +/– upbeat nystagmus (?nucleus intercalatus of Staderini).

References
Hirose G, Ogasawara T, Shirakawa T, *et al*. Primary position upbeat nystagmus due to unilateral medial medullary infarction. *Annals of Neurology* 1998; **43**: 403-406
Sawada H, Seriu N, Udaka F, Kameyama M. Magnetic resonance imaging of medial medullary infarction. *Stroke* 1990; **21**: 963-966
Cross References
Fasciculation; Hemiplegia; Lateral medullary syndrome; Nystagmus

Menace Reflex

- see BLINK REFLEX

Meningism

Meningism (meningismus, nuchal rigidity) is a stiffness or discomfort on passive movement (especially flexion) of the neck in the presence of meningeal irritation (*e.g*., infective meningitis, subarachnoid hemorrhage). A number of other, eponymous, signs of meningeal irritation have been described, of which the best known are those of Kernig and Brudzinski.

Meningism is not synonymous with meningitis, since it may occur in acute systemic pyrexial illnesses (pneumonia, bronchitis), especially in children. Moreover, meningism may be absent despite the presence of meningitis in the elderly and those receiving immunosuppression.

Cross References
Brudzinski's (neck) sign; Kernig's sign; Nuchal rigidity

Metamorphopsia

Metamorphopsia is an illusory visual phenomenon characterized by objects appearing distorted or misshapen in form. As with neglect, these phenomena may be classified as object- or person-centered:

- Object-centered: affecting size and spatial relationships
  Macropsia: objects appear larger than normal

Micropsia: objects appear smaller than normal
Pelopsia: objects appear closer to the observer than actual
Porropsia: objects appear farther away from the observer than actual

- Person centered:
  Micro- and macrosomatognosia: body image appears smaller or larger than normal ("Alice in Wonderland" syndrome).

Metamorphopsias are often transient and episodic, occurring for example during migraine attacks, epileptic seizures, with psychotropic drug abuse, and following petechial intraparenchymal hemorrhages. Rarely, they are long-lasting or permanent, for example following brain infarction (most commonly involving the occipito-parietal or temporo-parietal cortex: lesions on the right are more likely than those on the left to give metamorphopsia) or tumors. Retinal disease causing displacement of photoreceptors may produce metamorphopsia: micropsia due to receptor separation in retinal edema, macropsia due to receptor approximation in retinal scarring. Occasional cases of metamorphopsia have been reported with lesions of the optic chiasm, optic radiation, and retrosplenial region. Indeed, it seems that metamorphopsia may occur with pathology at any point along the visual pathway from retina to cortex. Differing patterns of metamorphopsia may assist with clinico-anatomical correlation:

- retinal lesions: ipsilateral monocular
- chiasmal lesions: bitemporal
- occipitoparietal lesions: contralateral homonymous

Metamorphopsia may be associated with visual hallucinations.

References

Shiga K, Makino M, Ueda Y, Nakajima K. Metamorphopsia and visual hallucinations restricted to the right visual hemifield after a left putaminal hemorrhage. *Journal of Neurology, Neurosurgery and Psychiatry* 1996; **61**: 420-421

Cross References

"Alice in Wonderland" syndrome; Hallucination; Illusion; Macropsia; Micropsia; Pelopsia; Porropsia; Telopsia

Micrographia

Micrographia is small handwriting. It is most often recognized in association with the extrapyramidal features of idiopathic Parkinson's disease (indeed it may be the presenting sign), but may occasionally occur with other parkinsonian syndromes (*e.g.*, progressive supranuclear palsy [PSP]) or in isolation with focal lesions of the midbrain or basal ganglia. In Parkinson's disease, handwriting may initially be of normal size but then become progressively smaller, slower, and more illegible as writing proceeds, an example of parkinsonian fatigue, a gradual

decline in the amplitude and speed of initiation of voluntary movements. Such "slow" micrographia may be distinguished from "fast" micrographia in which letters are small throughout although written at normal speed without fatigue, which may be seen in PSP or other pallidal pathologies.

There is a poor correlation between micrographia and the side, severity or duration of classical parkinsonian features, and its response to levodopa preparations is very variable. These observations, along with reports of isolated micrographia with cortical lesions demonstrated by neuroimaging, suggest that the anatomical basis of micrographia may be at the level of the cortex (dominant parietal lobe) rather than the basal ganglia.

Micrographia has also been described following large right anterior cerebral artery infarcts and lacunar infarcts involving the putamen and genu of the internal capsule.

References

McLennan JE, Nakano K, Tyler HR, Schwab RS. Micrographia in Parkinson's disease. *Journal of the Neurological Sciences* 1972; **15**: 141-152

Scolding NJ, Lees AJ. Micrographia associated with a parietal lobe lesion in multiple sclerosis. *Journal of Neurology, Neurosurgery and Psychiatry* 1994; **57**: 739-741

Cross References

Fast micrographia; Fatigue; Parkinsonism

Micropsia

Micropsia, or "Lilliput sight," is an illusory phenomenon in which the size of a normally recognized object is underestimated. It is the most common form of metamorphopsia, and is most often associated with lesions of the right temporo-parietal cortex, although macular edema and optic chiasm lesions may also cause micropsia. In migraine transient micropsia may occur. Hemimicropsia, micropsia confined to one visual hemifield, has been recorded.

The entirely subjective nature of the disorder may account for the relative rarity of reports.

References

Ceriani F, Gentileschi V, Muggia S, Spinnler H. Seeing objects smaller than they are: micropsia following right temporo-parietal infarction. *Cortex* 1998; **34**: 131-138

Cohen L, Gray F, Meyrignac C *et al*. Selective deficit of visual size perception: two cases of hemimicropsia. *Journal of Neurology, Neurosurgery and Psychiatry* 1994; **57**: 73-78

Cross References

Metamorphopsia

Microsomatognosia

- see "ALICE IN WONDERLAND" SYNDROME

Milkmaid's Grip

Milkmaid's grip is the descriptive term applied to the inability to maintain a firm grip (*e.g.*, of the examiner's fingers), detected as an alternating squeezing and releasing (as required for successful milking by hand). Seen in Huntington's disease, this may reflect a combination of chorea and motor impersistence.

Cross References

Chorea, Choreoathetosis; Impersistence; Trombone tongue

Miosis

Miosis is abnormal reduction in pupillary size, which may be unilateral or bilateral. Causes include:

Oculosympathetic paresis of whatever cause, *e.g.*, Horner's syndrome (unilateral), pontine hemorrhage (bilateral), early stages of central cephalic herniation (bilateral)
Drug-induced: *e.g.*, opiates (bilateral)
Pupils tend to be small and reactive in metabolic-toxic encephalopathies (bilateral)
"Senile miosis" (bilateral): age-related

If only one pupil appears small (anisocoria), it is important to distinguish miosis from contralateral mydriasis, when a different differential will apply.

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

Age-related signs; Anisocoria; Argyll Robertson pupil; Horner's syndrome; Mydriasis

Mirror Agnosia

Mirror agnosia, or the "looking glass syndrome," is a phenomenon observed in patients with left hemispatial neglect as a result of right parietal lobe lesions. There is inability to point to objects seen in a mirror, with repeated reaching "into" the mirror even when the actual location of the target is shown. In a milder form, known as "mirror ataxia," patients reach in the direction of the object but with increased errors of reach and grasp, suggesting that visual information is not adequately transformed into a body-centered frame of reference.

References

Binkofski F, Buccino G, Dohle C, Seitz RJ, Freund H-J. Mirror agnosia and mirror ataxia constitute different parietal lobe disorders. *Annals of Neurology* 1999; **46**: 51-61

Ramachandran VS, Altschuler EL, Hillyer S. Mirror agnosia. *Proceedings of the Royal Society of London, Series B* 1997; **264**: 645-647

Cross References

Agnosia; Neglect

Mirror Ataxia
- see MIRROR AGNOSIA

Mirror Hallucination
- see AUTOSCOPY

Mirror Movements
Mirror movements are involuntary movements of one side of the body that accompany and "mirror" (reflect) intentional movements on the opposite side of the body (also known as imitation synkinesis). They are usually symmetrical and most often seen when using distal muscles of the upper limb. Mirror movements are frequently present in young children but prevalence decreases with age. Persistence of mirror movements into adult life ("congenital mirror movements") is pathological, as is acquisition in adult life. These movements are uncommon after acquired brain lesions with no relationship to specific anatomical areas.

Congenital mirror movements are associated with skeletal developmental abnormalities, especially of the atlanto-occipital region, such as Klippel-Feil syndrome. They are also seen in 85% of patients with X-linked Kallmann syndrome (hypogonadotrophic hypogonadism and anosmia). Acquired mirror movements have been described following thalamic lesions, and in association with spastic paraparesis, extrapyramidal disorders, Friedreich's ataxia, phenylketonuria, and affecting hemiparetic limbs following stroke in young children.

There is some neurophysiological evidence from patients with X-linked Kallmann syndrome for the existence of an ipsilateral corticospinal pathway, consistent with other evidence that the congenital condition is primarily a disorder of axonal guidance during development. Concurrent activity within ipsilateral and contralateral corticospinal pathways may explain mirroring of movements. Alternatively, a failure of transcallosal inhibition, acquired at the time of myelination of these pathways, may contribute to the genesis of mirror movements. Loss of joint position sense following thalamic lesions may be of relevance. A deficit of sustained attention has also been postulated as the cause of mirror movements.

References

Farmer SF, Harrison LM, Mayston MJ, Parekh A, James LM, Stephens JA. Abnormal cortex-muscle interactions in subjects with X-linked Kallmann's syndrome and mirror movements. *Brain* 2004; **127**: 385-397

Mayston MJ, Harrison LM, Quinton R, Stephens JA, Krams M, Bouloux P-MG. Mirror movements in X-linked Kallmann's syndrome. I. A neurophysiological study. *Brain* 1997; **120**: 1199-1216

Cross References

Anosmia; Attention; Mirror writing; Proprioception; Synkinesia, Synkinesis

"Mirror Sign"
The term "mirror sign" has been applied to the phenomenon of misrecognition of self as another when seen in a mirror. It may be classi-

fied with the delusional misidentification syndromes. This may occur in Alzheimer's disease and frontotemporal dementia, and is associated with impaired cognition, confabulation, and prefrontal dysfunction. It may lead to a patient complaint of an intruder or a stranger living in the house ("phantom boarder" syndrome).

Some authors believe "the phenomenon of the mirror" to be an extreme example of prosopagnosia, but other studies have not found an association.

References

Caixeta LF, Caramelli P, Bahia V, Buchpiguel CA, Nitrini R. Clinical and neuroanatomical correlates of the mirror sign in frontotemporal dementia and Alzheimer's disease. *Neurobiology of Aging* 2000; **21(suppl1)**: S217 (abstract 988)

Cross References

Confabulation; Misidentification syndromes; "Picture sign"; Prosopagnosia

Mirror Writing

As the name implies, mirror writing is a mirror image of normal writing, hence running from right to left, with characters back to front. This may occur spontaneously, apparently more often in left-handers, or in right-handers attempting to write with the left hand following left-sided brain injury (*e.g.*, stroke).

Leonardo da Vinci is the most celebrated mirror writer: it is possible his left-handedness, and hence mirror writing, followed an injury to his right hand. The author Lewis Carroll occasionally wrote mirror letters but these differ from his normal script, unlike the situation with Leonardo whose two scripts are faithful mirror images. Carroll's letters may thus reflect not an inherent capacity but a contrivance, designed to amuse children who corresponded with him. The device was also used by the author Arthur Ransome in his 1939 novel *Secret Water*. Jane Austen wrote one letter (1817) to a young niece in which script runs from right to left but with word order reversed within words (*i.e.*, not mirror writing).

Various neural mechanisms are proposed to explain mirror writing, including bilateral cerebral representation of language, motor programs or visual memory traces or engrams. The mechanisms may differ between a true mirror writer like Leonardo and someone performing the task for amusement like Carroll.

Double mirror writing (*écriture en double miroir*) is inverted top to bottom (*i.e.*, script goes up the page, upside down) in addition to being mirror reversed.

References

Critchley M. *Mirror-writing*. London: Kegan Paul, Trench and Trubner, 1928

Larner AJ. The neurology of "Alice." *Advances in Clinical Neuroscience & Rehabilitation* 2005; **4(6)**: 35-36

Le Faye D (ed.). *Jane Austen's letters*. Oxford: Oxford University Press, 1995: 324

McManus C. *Right hand, left hand. The origins of asymmetry in brains, bodies, atoms and culture*. London: Phoenix, 2003: 348-350
Schott GD. Mirror writing: Allen's self observations, Lewis Carroll's "looking glass" letters, and Leonardo da Vinci's maps. *Lancet* 1999; **354**: 2158-2161

Misidentification Syndromes

These are defined as delusional conditions in which patients incorrectly identify and reduplicate people, places, objects, or events. Examples include:

Capgras syndrome; may be related to reduplicative paramnesia
Fregoli syndrome
Intermetamorphosis
Phantom boarder sign
Mirror sign

References

Feinberg TE, Roane DM. Misidentification syndromes. In: Feinberg TE, Farah MJ (eds.). *Behavioral neurology and neuropsychology*. New York: McGraw-Hill, 1997: 391-397
Forstl H, Almeida OP, Owen AM, Burns A, Howard R. Psychiatric, neurological and medical aspects of misidentification syndromes: a review of 260 patients. *Psychological Medicine* 1991; **21**: 905-910

Cross References

Delusion; Intermetamorphosis; "Mirror sign"; Reduplicative paramnesia

Misoplegia

Misoplegia is a disorder of body schema in which there is active hatred of a paralyzed limb, with or without personification of the limb, and attempts to injure the paralyzed limb. It occurs with right parietal region injury (hence left sided limbs most often involved) and may occur in conjunction with anosognosia, left hemispatial neglect, and (so called) constructional apraxia.

References

Critchley M. Misoplegia, a hatred of hemiplegia. *Mount Sinai Journal of Medicine* 1974; **41**: 82-87
Moss AD, Turnbull OH. Hatred of hemiparetic limbs (misoplegia) in a 10 year old child. *Journal of Neurology, Neurosurgery and Psychiatry* 1996; **61**: 210-211

Cross References

Anosognosia; Apraxia; Hemiparesis; Hemiplegia; Neglect

Mitbewegungen

- see SYNKINESIA, SYNKINESIS

Mitgehen

An abnormality of induced movement, in which limb movement occurs in response to application of the slightest pressure despite the

patient having been told to resist (German: to go too); a manifestation of negativism.
Cross References
Negativism

Mitmachen
A motor disorder in which the patient acquiesces to every passive movement of the body made by the examiner, but as soon as the examiner releases the body part, the patient returns it to the resting position.

Monoballismus
Monoballismus is ballism affecting a single limb.
Cross References
Ballism, Ballismus; Hemiballismus

"Monochromatopsia"
The author has seen a patient with a diagnosis of frontotemporal dementia who persistently and consistently complained that everything he saw was red, even though he was aware that they were not red, for example his wife's gray hair. His speech was fluent without paraphasia although impoverished in content, with recurrent themes repeated almost verbatim. He had mild orofacial dyspraxia. There was no alexia. Confronted with objects of different colors, he was unable to point to them by color since all appeared red to him. The features seem to be distinct from erythropsia (persistent) or phantom chromatopsia (normal visual acuity). The author proposes that this phenomenon might be termed "monochromatopsia."
Cross References
Erythropsia; Phantom chromatopsia

Monomelia
- see MONOPARESIS, MONOPLEGIA

Mononeuritis Multiplex, Mononeuropathy Multiplex
- see NEUROPATHY

Mononeuropathy
- see NEUROPATHY

Monoparesis, Monoplegia
Monoparesis is weakness, monoplegia complete weakness ("paralysis"), of a single limb. Monoparesis of the arm or leg of upper motor neurone type is usually cortical in origin, although may unusually arise from a cord lesion (leg more frequently than arm). Hoover's sign and Babinski's trunk-thigh test may be helpful in deciding whether monoparetic/monoplegic leg weakness is of nonorganic origin; the "arm drop" or "face-hand test" in arm weakness.

Peripheral disorders can sometimes present exclusively with single limb weakness, such as monomelic motor neurone disease (Hirayama disease), multifocal motor neuropathy with conduction block, and Guillain-Barré syndrome.

Cross References
"Arm drop"; Babinski's trunk-thigh test; Hemiparesis; Hoover's sign

Monotonia
Monotonia is a restricted range of speech inflection, occurring with hypophonia as part of the hypokinetic dysarthria observed in parkinsonism.
Cross References
Dysarthria; Hypophonia; Parkinsonism

Moria
Moria is literally folly, (as in Desiderius Erasmus' *Moriae Encomium* of 1509, literally "praise of folly"). In clinical usage, the meaning overlaps with that of emotional lability but has also be used in the context of pathological laughter.
Cross References
Emotionalism, Emotional lability; Pathological crying, Pathological laughter; Witzelsucht

Muscle Hypertrophy
Muscle hypertrophy is muscle enlargement due to an increase in the size of its myofibrils. Muscle hypertrophy may be generalized or focal; and occur in response to repetitive voluntary contraction (physiological) or repetitive abnormal electrical activity (pathological, *e.g.,* myotonia in Thomsen's disease; primary orthostatic tremor).

Muscle enlargement may also result from replacement of myofibrils by other tissues, such as fat or amyloid, a situation better described as pseudohypertrophy.
Cross References
Calf hypertrophy; Masseter hypertrophy; Myotonia

Mutism
Mutism is absence of speech output. This may be psychogenic, as in schizophrenia or affective disorders, with or without catatonia; or a consequence of neurological disease, for example:

Akinetic mutism
Dementia syndromes, especially frontal lobe dementia, late stages of primary progressive aphasia
Encephalopathy (toxic/drug-induced/metabolic)
Damage to Broca's area, supplementary motor area; severe pseudobulbar palsy, bilateral thalamic damage
Cerebellar mutism: rare, following midline cerebellar surgery in children
Bilateral vocal cord paralysis (although this may be better termed aphonia)

In neurological disorders there may be difficulty initiating movements, completing motor sequences, or inhibition of appropriate responses.

References

Altshuler LL, Cummings JL, Mills MJ. Mutism: review, differential diagnosis and report of 22 cases. *American Journal of Psychiatry* 1986; **143**: 1409-1414 (erratum: *American Journal of Psychiatry* 1987; **144**: 542)
Ersahin Y, Mutluer S, Cagli S, Duman Y. Cerebellar mutism: report of seven cases and review of the literature. *Neurosurgery* 1996; **38**: 60-66

Cross References

Akinetic mutism; Aphonia; Catatonia; Dementia; Encephalopathy; Pseudobulbar palsy

"Myasthenic Snarl"

Patients with weakness of facial musculature as a consequence of myasthenia gravis may have a "transverse smile," with lack of elevation of the corners of the mouth, or appear to snarl when asked to smile or laugh. This may give the impression that they seem peculiarly unamused by an examiner's attempted witticisms. These phenomena may be seen with other causes of facial weakness, such as facioscapulohumeral (FSH) dystrophy.

Mydriasis

Mydriasis is an abnormal dilatation of the pupil, either unilateral or bilateral. Causes include:

- Oculoparasympathetic paresis, from lesions at the Edinger-Westphal nucleus or anywhere along the course of the oculomotor (III) nerve (usually unilateral)
- Tonic enlargement of the pupil (Holmes-Adie pupil, usually unilateral)
- Sympathomimetic drugs, *e.g.,* adrenaline (usually bilateral)
- Later stages of central cephalic herniation
- If only one pupil appears large (anisocoria), it is important to distinguish mydriasis from contralateral miosis, when a different differential will apply (*e.g.,* Horner's syndrome).

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

Anisocoria; Holmes-adie pupil, Holmes-adie syndrome; Horner's syndrome; Hutchinson's pupil; Miosis; Oculomotor (III) nerve palsy

Myelopathy

A myelopathy is a disorder of the spinal cord. Such disorders may be further characterized according to whether the responsible lesion lies within or outside the spinal cord: intrinsic or intramedullary lesions are always intradural; extrinsic or extramedullary lesions may be

intradural or extradural. It may be possible to differentiate intramedullary from extramedullary lesions on clinical grounds, although this distinction is never absolute because of clinical overlap.

- Clinical features of extrinsic/extramedullary myelopathy:

  Motor: sequential spastic paraparesis below the level of the lesion; upper motor neurone (UMN) signs occur early; lower motor neurone (LMN) signs are unusual and have a segmental (radicular) distribution if present

  Sensory: symptoms of pain may be radicular (*e.g.,* secondary to a neurofibroma) or vertebral (*e.g.,* secondary to neoplastic or inflammatory processes); sensory signs are not usually marked until the later stages, and all modalities are often involved. A Brown-Séquard syndrome may be commoner in extrinsic than intrinsic myelopathies

  Sphincters: may have bladder urgency, impotence

  Pathologies commonly causing extrinsic myelopathy include:

  Prolapsed disc, osteophyte bar

  Tumor (primary, secondary)

  Arteriovenous malformation/hematoma

  Abscess

- Clinical features of intrinsic/intramedullary myelopathy:

  Motor: LMN signs may be prominent and diffuse; UMN signs tend to occur late (spastic paraparesis below level of lesion). A combination of UMN and LMN signs is much more likely to reflect intrinsic than extrinsic pathology

  Sensory: symptoms of central (funicular) pain may occur; dissociated sensory loss (spinothalamic > dorsal column involvement, or *vice versa*), suspended sensory loss, and sacral sparing are characteristic of intramedullary lesions; a Brown-Séquard syndrome may occur. Vibratory sensibility is more often affected than proprioception

  Sphincters: bladder involvement common, often early and slow to recover.

These features are dependent on the extent to which the cord is involved: some pathologies have a predilection for posterior columns, central cord, *etc*.

Pathologies commonly causing intrinsic myelopathy include:

Multiple sclerosis or other inflammatory process causing transverse myelitis (complete or partial), *e.g.,* viral infection, HTLV-1 infection, tabes dorsalis

Tumor (primary, secondary)

Syringomyelia

Infarction, *e.g.,* anterior spinal artery syndrome

Metabolic causes: vitamin B_{12} deficiency producing subacute combined degeneration of the cord.

Imaging of the cord, ideally with MRI, may be helpful in defining the cause of myelopathy.

References
Daly EC. Spinal cord disorders. In: Biller J (ed.). *Practical neurology* (2nd edition). Philadelphia: Lippincott Williams & Wilkins, 2002: 593-613
Johnston RA. Acute spinal cord compression. In: Hughes RAC (ed.). *Neurological emergencies* (2nd edition). London: BMJ Publishing, 1997: 272-294
Tartaglino LM, Flanders AE, Rapoport RJ. Intramedullary causes of myelopathy. *Seminars in Ultrasound, CT, and MRI* 1994; **15**: 158-188
Cross References
Brown-séquard syndrome; Lower motor neurone (LMN) syndrome; Paraparesis; Proprioception; Sacral sparing; Suspended sensory loss; Upper motor neurone (UMN) syndrome; Vibration

Myerson's Sign
- see GLABELLAR TAP REFLEX

Myoclonus
Myoclonus is involuntary, "shock-like," muscle jerking, arrhythmic more often than regular, of central nervous system (CNS) origin. This may be focal, multifocal, or generalized. Multiple irregular asynchronous myoclonic jerks may be termed polymyoclonus. Myoclonus may be characterized in several ways:

- Clinical classification (by observation, examination):
  - Spontaneous
  - Action or intention: following voluntary action; may be elicited by asking patient to reach out to touch the examiner's hand
  - Reflex, stimulus-sensitive: jerks produced by somatesthetic stimulation of a limb, in response to loud noises
- Anatomical/pathophysiological classification (by electrophysiological recordings):
  - Cortical
  - Subcortical/reticular
  - Propriospinal/segmental
- Etiological classification:
  - Physiological, *e.g.,* "sleep starts" (hypnic jerks)
  - Essential: in the absence of any other abnormality of the CNS
  - Epileptic: as a manifestation of idiopathic epilepsy
  - Symptomatic: of other neurological diseases, of which there are many, including:

Anoxic brain injury (Lance-Adams syndrome)
Vascular lesions
Neoplasia
Encephalopathies: especially of metabolic origin (hepatic, renal), but also toxic, viral, paraneoplastic, mitochondrial

Degenerations: basal ganglia, spinocerebellar
Malabsorption syndromes (coeliac disease, Whipple's disease)
Storage disorders, for example, Lafora body disease, Tay-Sachs disease, sialidosis
Dementias: Alzheimer's disease (usually late), prion disease (usually early in sporadic Creutzfeldt-Jakob disease)

The clinical differential diagnosis of myoclonus includes chorea, tic, tremor (especially with rhythmic myoclonus), and certain peripheral nerve disorders (fasciculation, myokymia).

Periodic limb movement disorder or periodic leg movements of sleep, frequently found in association with restless legs syndrome, is sometimes called "nocturnal myoclonus."

Brief lapses of muscle contraction with loss of posture are in some ways the converse of myoclonus and have in the past been labeled "negative myoclonus," although the term asterixis is now preferred.

Drugs useful in the treatment of myoclonus include clonazepam, sodium valproate, primidone, and piracetam. These may need to be given in combination to suppress severe action myoclonus.

References

Barker R. Myoclonus. *Advances in Clinical Neuroscience & Rehabilitation* 2003; **3(5)**: 20,22

Caviness JN. Myoclonus. *Mayo Clinic Proceedings* 1996; **71**: 679-688

Marsden CD, Hallett M, Fahn S. The nosology and pathophysiology of myoclonus. In: Marsden CD, Fahn S (eds.). *Movement Disorders.* London, Butterworth, 1982: 196-248

Obeso JA, Artieda J, Rothwell JC, Day B, Thompson P, Marsden CD. The treatment of severe action myoclonus. *Brain* 1989; **112:** 765-777

Cross References

Asterixis; Chorea, Choreoathetosis; Fasciculation; Hiccups; Jactitation; Myokymia; Palatal myoclonus; Tic; Tremor

Myoedema

Myoedema, or muscle mounding, provoked by mechanical stimuli or stretching of muscle, is a feature of rippling muscle disease, in which the muscle contractions are associated with electrical silence. It has also been reported as a neuromuscular feature of hypothyroidism.

References

Torbergsen T. Rippling muscle disease: a review. *Muscle and Nerve* 2002; **Suppl 11**: S103-S107

Myokymia

Myokymia is an involuntary, spontaneous, wave-like, undulating, flickering movement within a muscle (*cf*. fasciculation); it may be likened to a "bag of worms."

Electrophysiologically this corresponds to regular groups of motor unit discharges, of peripheral nerve origin. Myokymia is thus related to neuromyotonia and stiffness, since there may be concurrent impairment of muscle relaxation and a complaint of muscle cramps.

A syndrome of superior oblique myokymia is described, often following superior oblique palsy, which produces a microtremor of the eye and causes oscillopsia or transient diplopia. Facial myokymia is a rare facial dyskinesia, possibly related to disinhibition of the facial (VII) nerve nucleus by focal pontine lesions (tumor, demyelination).

References

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

Cross References

Fasciculation; Myotonia; Neuromyotonia; Stiffness

Myopathy

The term myopathy means a primary disorder of muscle causing wasting and/or weakness in the absence of sensory abnormalities. Clinically, myopathic processes need to be differentiated from neuropathies, particularly anterior horn cell diseases and motor neuropathies, and neuromuscular junction disorders. Generally in primary muscle disease there are no fasciculations, reflexes are lost late, and phenomena such as (peripheral) fatigue and facilitation do not occur.

Myopathies may be subdivided according to the clinical pattern of weakness, and/or their etiology:

- *Proximal*:
  Affecting shoulder abductors, hip flexors predominantly:
  Inflammatory: polymyositis, dermatomyositis
  Progressive muscular dystrophies: Duchenne, Becker, limb-girdle, facioscapulohumeral (FSH)
  Metabolic: acid-maltase deficiency; thyroid dysfunction, Cushing's syndrome
  Nonmetastatic feature of malignant disease.
- *Distal*:
  An unusual pattern for myopathy, which needs to be differentiated from distal polyneuropathy:
  Myotonic dystrophy
  Miyoshi dystrophy
  Desmin myopathy.
- *Bulbar palsy* (*q.v.*).
- *Facial paresis* (*q.v.*).
- *Diaphragm weakness*:
  Acid-maltase deficiency
  Acute polymyositis
  Neuralgic amyotrophy.
- *Axial myopathy*:
  Camptocormia ("bent spine syndrome")
  Dropped head syndrome.

References

Barnes PRJ, Hilton-Jones D, Dalakas MC, Palace JA, Rose MR. *Myopathy in clinical practice*. London: Martin Dunitz, 2003

Mandler RN. Myopathy. In: Biller J (ed.). *Practical neurology* (2nd edition). Philadelphia: Lippincott Williams & Wilkins, 2002: 623-641

Cross References

Atrophy; Bulbar palsy; Camptocormia; Dropped head syndrome; Facial paresis; Fatigue; Gowers' sign; Paradoxical breathing; Wasting; Weakness

Myorhythmia

Myorhythmia is an involuntary movement disorder characterized by rhythmic contraction (1-3 Hz) of muscles producing a coarse tremor, which may affect limbs, face, palate, head, jaw, neck, tongue, eyes or trunk. The movements are continuous and persist during sleep. They are associated with brainstem or thalamic vascular disease, trauma, alcohol-related nutritional deficiency, phenytoin intoxication, Hashimoto's encephalopathy, paraneoplasia, and Whipple's disease.

Although very rare, oculomasticatory myorhythmia is of diagnostic importance since it is pathognomonic for Whipple's disease of the nervous system. Characteristically there is also convergent-divergent pendular nystagmus with synchronous rhythmic movement of the mouth, tongue, jaw and sometimes proximal and distal skeletal muscles. The neurological manifestations of Whipple's disease are protean, and include dementia, ataxia, supranuclear ophthalmoplegia (with sparing of the pupils), seizures, myoclonus, nystagmus and psychosis. The condition is caused by the bacterium *Tropheryma whipplei*. Treatment is with antibiotics, usually a two-week intravenous course of trimethoprim-sulphamethoxazole or ceftriaxone followed by oral treatment for one year. Sodium valproate may be helpful for the involuntary movements which do not respond to antibiotics.

References

Anderson M. Neurology of Whipple's disease. *Journal of Neurology, Neurosurgery and Psychiatry* 2000; **68**: 2-5

Masucci EF, Kurtzke JF, Saini N. Myorhythmia: a widespread movement disorder. *Brain* 1984; **107**: 53-79

Simpson DA, Wishnow R, Gargulinski RB, Pawlak AM. Oculofacial-skeletal myorhythmia in central nervous system Whipple's disease: additional case and review of the literature. *Movement Disorders* 1995; **10**: 195-200

Cross References

Ataxia; Dementia; Myoclonus; Nystagmus

Myotonia

Myotonia is a stiffness of muscles with inability to relax after voluntary contraction (action myotonia), or induced by electrical or mechanical (*e.g.,* percussion myotonia) excitation. The phenomenon is often described by patients as "cramp" or stiffness. This is a reflection of primary muscle disease (*i.e.,* myogenic; *cf.* neuromyotonia, neurogenic muscle stiffness), which persists after peripheral nerve or neuromuscular junction blockade.

Electrophysiology reveals myotonic discharges, with prolonged twitch relaxation phase, which may be provoked by movement, percussion, and electrical stimulation of muscle; discharges typically wax and wane.

A similar clinical phenomenon of slow muscle relaxation may be observed in other circumstances, for example hypothyroidism, but without the characteristic EMG findings of myotonia, hence this is labeled as pseudomyotonia. Paramyotonia is myotonia exacerbated by cold and exertion (paradoxical myotonia).

Recognized causes of myotonia include:

- myotonic dystrophy (myotonia dystrophica; myotonic dystrophy type 1)
- hyperkalaemic periodic paralysis
- myotonia congenita (autosomal dominant Thomsen's disease, autosomal recessive Becker's myotonia)
- K^+-aggravated myotonia
- Schwartz-Jampel syndrome (chondrodystrophic myotonia)
- proximal myotonic myopathy (PROMM; myotonic dystrophy type 2)

Mutations in genes encoding voltage-gated ion channels have been identified in some of the inherited myotonias, hence these are channelopathies: skeletal muscle voltage-gated Na^+ channel mutations have been found in K^+-aggravated myotonia, and also paramyotonia congenita and hyperkalaemic periodic paralysis. Chloride (Cl^-) channel mutations have been identified in myotonia congenita. These latter conditions respond best to mexiletine.

References

Harper PS, van Engelen B, Eymard B, Wilcox DE (eds.). *Myotonic dystrophy: present management, future therapy*. Oxford: OUP, 2004

Mankodi A, Thornton CA. Myotonic syndromes. *Current Opinion in Neurology* 2002; **15**: 545-552

Cross References

Neuromyotonia; Paramyotonia; Percussion myotonia; Pseudomyotonia; Stiffness; Warm-up phenomenon; Woltman's sign

N

Narcolepsy, Narcoleptic Syndrome
- see HYPERSOMNOLENCE

Nasopalpebral Reflex
- see GLABELLAR TAP REFLEX

Negative Myoclonus
- see ASTERIXIS

Negative Tremor
- see ASTERIXIS

Negativism
Negativism is a motor sign of mental disorder, usually schizophrenia, consisting of the patient doing the opposite of what is asked and actively resisting efforts to persuade compliance. Movement of a limb in response to application of pressure despite the patient having been told to resist (*mitgehen*) is one element of negativism. It may also be a feature of catatonia. The similarity of some of these features to *gegenhalten* suggests the possibility of frontal lobe dysfunction as the underlying cause.
Cross References
Catatonia; Gegenhalten

Neglect
Neglect is a failure to orient toward, respond to, or report novel or meaningful stimuli. If failure to respond can be attributed to concurrent sensory or motor deficits (*e.g.,* hemiparesis, hemianopia, visuospatial deficits) neglect is not present.

Neglect can involve stimuli in the extrapersonal environment (*e.g.,* visual neglect) or personal space (*e.g.,* personal neglect or asomatognosia). Neglect of contralateral hemispace may also be called unilateral spatial neglect, hemi-inattention, or hemineglect. Lesser degrees of neglect may be manifest as extinction (double simultaneous stimulation). Motor neglect may be evident as hemiakinesia, hypokinesia, or motor impersistence. Alloesthesia and allokinesia may also be features of neglect.

Neglect may be obvious (*e.g.,* patient not dressing one side of the body), but is sometimes more subtle, in which case it may be tested for using various simple tests:

- Cancellation tests, for example, stars (unstructured array), letters (structured array)

- Figure copying, for example, Rey-Osterreith figure
- Line bisection, numbering a clock face
- Drawing from memory.

Neglect is commoner after right rather than left brain damage, usually of vascular origin. The angular gyrus and parahippocampal gyrus may be central to the development of visual neglect. Marked degrees of neglect may seriously hamper attempts at neurorehabilitation.

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Cross References

Alexia; Alloesthesia; Allokinesia; Asomatognosia; Extinction; Hemiakinesia; HypoKinesia; Impersistence

Negro's Sign

Negro has two eponymous signs:

- Cogwheel (jerky) type of rigidity in basal ganglia disorders.
- In both peripheral and central facial paralysis, the eyeball deviates outward and elevates more than normal when the patient attempts to look up due to overaction of the inferior oblique and superior rectus muscles, respectively.

Cross References

Bell's palsy; Facial paresis; Parkinsonism; Rigidity

Neologism

A neologism is a nonword approximating to a real word, produced in spontaneous speech; it is thought to result from an inability to organize phonemes appropriately in the process of speech production. Hence, this is a type of literal or phonemic paraphasia encountered in aphasic syndromes, most usually those resulting from left superior temporal lobe damage (Wernicke type). (The word "scientist" is said to be a neologism coined in the nineteenth century by William Whewell.)

Cross References

Aphasia; Paraphasia; Schizophasia; Wernicke's aphasia

Neri's test

- see LASÉGUE'S SIGN

Nerve Thickening

The characterization of a peripheral neuropathy should always include examination to see if any nerves are thickened. Good places to feel for nerve thickening include the elbow (ulnar nerve), anatomical snuff box (superficial radial nerves), and head of the fibula (common peroneal nerve).

Nerve thickening may be noted in a variety of conditions, in some by examination, in others using imaging techniques:

Leprosy
Hereditary motor and sensory neuropathies (HMSN), especially types I, III, and IV (Refsum's disease)
Hereditary neuropathy with liability to pressure palsies (HNLPP)/tomaculous neuropathy
Neurofibromatosis
Sarcoidosis
Chronic inflammatory demyelinating neuropathy/ophthalmoplegic migraine
Nerve tumors (localized)
Amyloidosis (familial amyloid polyneuropathy, primary systemic amyloidosis): rare.

References

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Cross References

Neuropathy

Neuromyotonia

Neuromyotonia is neurogenic muscle stiffness (*cf*. myotonia, myogenic muscle stiffness) which reflects peripheral nerve hyperexcitability. Clinically this is manifest as muscle cramps and stiffness, particularly during and after muscle contraction, and as muscular activity at rest (myokymia, fasciculations). Tendon areflexia and abnormal postures of hands and feet may also be observed.

A syndrome of ocular neuromyotonia has been described in which spasms of the extraocular muscles cause a transient heterophoria and diplopia.

Physiologically neuromyotonia is characterized by continuous motor unit and muscle fibre activity which is due to peripheral nerve hyperexcitability; it is abolished by curare (*cf*. myotonia). Neuromyotonia may be associated with autoantibodies directed against presynaptic voltage-gated K^+ channels. Around 20% of patients have an underlying small-cell lung cancer or thymoma, suggesting a paraneo-

plastic etiology in these patients. Neuromyotonia has also been associated with mutations within the voltage-gated K^+ ion channel gene.

Carbamazepine and phenytoin may help the stiffness and areflexia.

References

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Cross References

Fasciculation; Myokymia; Myotonia; Paramyotonia; Pseudomyotonia; Stiffness

Neuronopathy

- see NEUROPATHY

Neuropathy

Neuropathies are disorders of peripheral nerves. Various clinical patterns of peripheral nerve involvement may be seen:

- *Mononeuropathy*: sensory and/or motor involvement in the distribution of a single nerve
- *Mononeuropathy multiplex*: simultaneous involvement of two or more nerves, usually in different parts of the body; if due to inflammatory disease (as is often the case) this may be described as mononeuritis multiplex
- *Polyneuropathy*: a widespread process, predominantly affecting the distal parts of nerves; may be predominantly sensory ("glove and stocking" sensory loss) or motor, with or without concomitant autonomic involvement.

These clinical patterns may need to be differentiated in practice from disorders affecting the neuronal cell bodies in the ventral (anterior) horns of the spinal cord or dorsal root ganglia (motor and sensory neuronopathies, respectively); and disorders of the nerve roots (radiculopathy) and plexuses (plexopathy). Clinical signs resulting from neuropathies are of lower motor neurone type (wasting, weakness, reflex diminution or loss).

The causes of neuropathy are legion. Mononeuropathies often result from local compression (entrapment neuropathy), trauma, or diabetes. Mononeuropathy multiplex often reflects intrinsic inflammation (*e.g.,* polyarteritis nodosa, Churg-Strauss syndrome,

systemic lupus erythematosus, rheumatoid arthritis, Sjögren's syndrome, cryoglobulinemia, isolated PNS vasculitis). Polyneuropathies may have genetic, infective, inflammatory, toxic, nutritional, and endocrine etiologies. Many neuropathies, particularly polyneuropathies in the elderly, remain idiopathic or cryptogenic, despite intensive investigation.

Sensory neuronopathies (ganglionopathy, polyganglionopathy) have a rather more limited differential diagnosis, including:

- Paraneoplasia: anti-Hu antibody syndrome (although a similar syndrome, presumed paraneoplastic, may occur in the absence of these antibodies)
- Sjögren's syndrome
- Associated with anti-GD1b ganglioside antibodies
- CIDP
- HIV.

References

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Cross References

Amyotrophy; Lower motor neurone (LMN) syndrome; Plexopathy; Radiculopathy; Wasting; Weakness

Nominal Aphasia

- see ANOMIA

Nuchal Rigidity

Nuchal rigidity is neck stiffness, and is usually synonymous with meningism, in which case other signs of meningeal irritation are usually present (Kernig's sign, Brudzinski's neck sign). If these other signs are absent, then isolated nuchal rigidity may suggest a foraminal pressure cone. It may also occur in syndromes causing predominantly axial (as opposed to limb) rigidity (*e.g.,* progressive supranuclear palsy). In intubated patients, there may be resistance to passive neck movements.

Cross References

Brudzinski's (neck) sign; Kernig's sign; Meningism; Parkinsonism

Nuchocephalic Reflex

In a standing subject, rapid turning of the shoulders to either left or right (eyes closed to avoid fixation) is associated with bilateral con-

traction of the cervical musculature so that the head is held in the original position. This nuchocephalic reflex is present in infants and children up to the age of about 4 years. Beyond this age the reflex is inhibited, such that the head is actively turned in the direction of shoulder movement after a time lag of about half a second. If the reflex is present in adults (*i.e.,* disinhibited), it is claimed to be a "regressive" (primitive) sign, indicative of diffuse cerebral dysfunction.

References

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Cross References

Age-related signs; Primitive reflexes

Nyctalopia

Nyctalopia, or night blindness, is an impairment of visual acuity specific to scotopic vision, implying a loss or impairment of rod photoreceptor function. Patients may spontaneously complain of a disparity between daytime and nocturnal vision, in which case acuity should be measured in different ambient illumination.

Nyctalopia may be a feature of:
Retinitis pigmentosa
Vitamin A deficiency
Cancer-associated retinopathy: most commonly associated with small-cell lung cancer (antirecoverin antibodies may be detected), though gynecological malignancy and melanoma have also been associated (with antibipolar retinal cell antibodies in the latter).

Cross References

Hemeralopia; Retinitis pigmentosa

Nylen-Bárány Maneuver

- see HALLPIKE MANEUVER, HALLPIKE TEST

Nystagmoid Jerks

- see NYSTAGMUS

Nystagmus

Nystagmus, or talantropia, is an involuntary, usually bilateral, oscillation of the eyeballs (very occasionally it is unilateral, *e.g.,* in internuclear ophthalmoplegia), of which many varieties are described. This may be:

- *Physiological*:
  Optokinetic nystagmus (OKN; *e.g.,* looking out of a moving railway carriage)
  Induced by vestibular stimuli (*e.g.,* merry-go-round; caloric testing)

Nystagmoid jerks: in extremes of lateral or vertical gaze (end-point nystagmus, a form of gaze-evoked nystagmus)

- *Pathological*:

  Nystagmus may be classified according to direction, waveform, anatomy/etiology, or clinical frequency (common, rare). When describing nystagmus, it is necessary to make observations in the nine cardinal positions of gaze for the direction, amplitude and beat frequency of nystagmus. Nystagmus may be abortive (transient) or sustained in duration. The intensity of jerk nystagmus may be classified by a scale of three degrees:

  1st degree: present when looking in the direction of the fast phase;
  2nd degree: present in the neutral position;
  3rd degree: present when looking in the direction of the slow phase (*i.e.,* present in all directions of gaze).

It is important to distinguish nystagmus from other involuntary eye movements, such as square-wave jerks, ocular flutter, and opsoclonus (*q.v.*).

- *Directional classification of nystagmus*:

  Horizontal (common)
  Vertical (rare):
  Downbeat: seen with structural lesions of the cervicomedullary junction, midline cerebellum and floor of the 4th ventricle, but also with more diffuse cerebellar disease.
  Upbeat: of less localizing value than downbeat, upbeat nystagmus may occur with pontomesencephalic, pontomedullary, and even caudal medullary lesions (infarct, inflammation); bow-tie nystagmus is probably a variant of upbeat nystagmus.
  Torsional: usually accompanies horizontal nystagmus of peripheral vestibular (labyrinthine) origin.

- *Waveform classification of nystagmus*:

  Jerk nystagmus:
  characterized by a slow drift of the eyes in one direction (slow phase) followed by a rapid, corrective, saccadic movement in the opposite direction (fast phase); the direction of jerk nystagmus is named according to the direction of the fast phase, but it is the character of the slow phase, the pathological part of the process, which is more eloquent regarding anatomical correlation (*vide infra*).
  Pendular or undulatory nystagmus:
  in which the movements of the eyes are more or less equal in amplitude and velocity (sinusoidal oscillations) about a central (null) point. This is often congenital, may be conjugate or disconjugate (sometimes monocular), but is not related to concurrent internuclear ophthalmoplegia or asymmetry of visual acuity.

When studied using oculography, the slow phase of jerk nystagmus may show a uniform velocity ("saw-toothed"), indicative of imbalance in vestibulo-ocular reflex activity. A slow phase with exponentially decreasing velocity (negative exponential slow phase) is

ascribed to "leakiness" of a hypothetical neural integrator, a structure which converts eye or head velocity signals into approximations of eye or head position signals (thought to lie in the interstitial nucleus of Cajal in the midbrain for vertical eye movements, and in the nucleus propositus hypoglossi for horizontal eye movements). A slow phase with exponentially increasing velocity (high-gain instability, runaway movements) may be seen in congenital or acquired pendular nystagmus. The pathophysiology of acquired pendular nystagmus is thought to be deafferentation of the inferior olive by lesions of the red nucleus, central tegmental tract, or medial vestibular nucleus.

- *Anatomical/etiological classification of nystagmus*:

  Peripheral Vestibular:

  unidirectional (directed to side opposite lesion), and more pronounced when looking in direction of the fast phase (*i.e.,* 1st degree), usually with a rotatory component and associated with vertigo. Tends to fatigue, and usually transient (*e.g.,* in Hallpike maneuver). Nystagmus of peripheral vestibular origin is typically reduced by fixation (hence these patients hold their heads still) and enhanced by removal of visual fixation (in the dark, with Frenzel's lenses).

  Central Vestibular:

  unidirectional or multidirectional, 1st, 2nd, or 3rd degree; typically sustained and persistent. There may be other signs of central pathology (*e.g.,* cerebellar signs, upper motor neurone signs). Not affected by removal of visual fixation.

  Cerebellar/brainstem:

  commonly gaze-evoked due to a failure of gaze-holding mechanisms. It may be unidirectional with a unilateral cerebellar lesion (*e.g.,* vascular disease) in which case it typically occurs when the eyes are looking in the direction of the lesion (*cf*. peripheral vestibular nystagmus); multidirectional nystagmus of cerebellar origin may occur in multiple sclerosis, drug/toxin exposure, cerebellar degenerations.

  Congenital:

  usually horizontal, pendular type nystagmus; worse with fixation, attention, anxiety. It may appear with blindness of childhood onset, or be acquired with neurological disease (multiple sclerosis, mitochondrial disease, Whipple's disease, Pelizaeus-Merzbacher disease).

  Other forms of nystagmus include:

- *Ataxic/Dissociated*:

  in abducting >> adducting eye, as in internuclear ophthalmoplegia and pseudo-internuclear ophthalmoplegia.

- *Periodic Alternating*:

  primary position nystagmus, almost always in the horizontal plane, which stops and then reverses direction every minute or so; 4-5 minutes observation may be required to see the whole cycle; localizing value similar to downbeat nystagmus.

- *Convergence-retraction* (Körber-Salus-Elschnig syndrome):

adducting saccades (medial rectus contraction), occurring spontaneously or on attempted upgaze, often accompanied by retraction of the eyes into the orbits, associated with mesencephalic lesions of the pretectal region (*e.g.,* pinealoma).

- *See-saw*:
  a disconjugate cyclic movement of the eyes, comprising elevation and intorsion of one eye while the other eye falls and extorts, followed by reversal of these movements; may be congenital (*e.g.,* with albinism, retinitis pigmentosa) or acquired (mesodiencephalic or lateral medullary lesions, *e.g.,* brainstem stroke, head trauma, syringobulbia).

Many pathologies may cause nystagmus, the most common being demyelination, vascular disease, tumor, neurodegenerative disorders of cerebellum and/or brainstem, metabolic causes (*e.g.,* Wernicke-Korsakoff's syndrome), paraneoplasia, drugs (alcohol, phenytoin, barbiturates, sedative-hypnotic drugs), toxins, and epilepsy. Treatment of nystagmus is usually that of the underlying cause, where possible. Pendular nystagmus may respond to anticholinesterases, consistent with its being a result of cholinergic dysfunction. Periodic alternating nystagmus responds to baclofen, hence the importance of making this diagnosis. See-saw nystagmus may respond to baclofen, clonazepam, or alcohol.

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Cross References

Caloric testing; Hallpike maneuver, Hallpike test; Internuclear ophthalmoplegia; Myorhythmia; Optokinetic nystagmus (OKN), Optokinetic response; Opsoclonus; Oscillopsia; Palatal myoclonus; Pseudo-internuclear ophthalmoplegia; Spasmus nutans; Square-wave jerks; Vertigo

O

Obscurations

Visual obscurations are transient losses ("graying out") of vision lasting a few seconds, occurring in the context of raised intracranial pressure (ICP), and especially associated with activities known to elevate ICP (coughing, sneezing, bending down, straining at stool) and relieved by their cessation. These symptoms are thought to reflect critical compromise of optic nerve head perfusion and are invariably associated with the finding of papilledema. Obscurations mandate urgent investigation and treatment to prevent permanent visual loss. Transient visual obscurations may occasionally be due to optic disc drusen.

Cross References

Papilledema

Obtundation

Obtundation is a state of altered consciousness characterized by reduced alertness and a lessened interest in the environment, sometimes described as psychomotor retardation or torpor. An increased proportion of time is spent asleep and the patient is drowsy when awake. Obtundation is a less severe impairment of consciousness than stupor.

Cross References

Coma; Psychomotor retardation; Stupor

Ocular Apraxia

Ocular apraxia (ocular motor apraxia) is a disorder of voluntary saccade initiation; reflexive saccades and spontaneous eye movements are preserved. Ocular apraxia may be overcome by using dynamic head thrusting, with or without blinking (to suppress vestibulo-ocular reflexes): the desired fixation point is achieved through reflex contraversive tonic eye movements to the midposition following the overshoot of the eyes caused by the head thrust.

The anatomical substrate of ocular apraxia is not certain. Ocular apraxia may occur as a congenital syndrome (in the horizontal plane only: Cogan's syndrome), or may be acquired in ataxia telangiectasia (Louis-Bar syndrome), Niemann-Pick disease (mainly vertical plane affected), and Gaucher's disease (horizontal plane only).

Cross-References

Apraxia; Saccades

Ocular Bobbing

Ocular bobbing refers to intermittent abnormal vertical eye movements, usually conjugate, consisting of a fast downward movement followed by a slow return to the initial horizontal eye position. The sign has no precise localizing value, but is most commonly associated with intrinsic

pontine lesions, *e.g.,* infarct, hemorrhage, tumor, central pontine myelinolysis. It has also been described in encephalitis, Creutzfeldt-Jakob disease, and toxic encephalopathies. Its pathophysiology is uncertain but may involve mesencephalic and medullary burst neurone centres.

Variations on the theme include:

- Inverse ocular bobbing: slow downward movement, fast return (also known as fast upward ocular bobbing, or ocular dipping)
- Reverse ocular bobbing: fast upward movement, slow return to midposition
- Converse ocular bobbing: slow upward movement, fast down (also known as slow upward ocular bobbing, or reverse ocular dipping).

References

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Cross References

Ocular dipping

Ocular Dipping

Ocular dipping, or inverse ocular bobbing, consists of a slow spontaneous downward eye movement with a fast return to the midposition. This may be observed in anoxic coma or following prolonged status epilepticus and is thought to be a marker of diffuse, rather than focal, brain damage.

Reverse ocular dipping (slow upward ocular bobbing) consists of a slow upward movement followed by a fast return to the midposition.

References

Stark JR, Masucci EF, Kurtzke JF. Ocular dipping. *Neurology* 1984; **34**: 391-393

Cross References

Ocular bobbing

Ocular Flutter

Ocular flutter is an eye movement disorder characterized by involuntary bursts of back-to-back horizontal saccades without an intersaccadic interval (*cf*. square-wave jerks). Ocular flutter may be accurately diagnosed with oculography.

The postulated mechanism of ocular flutter is loss of "pause" neuronal inhibition of "burst" neurone function in the paramedian pontine reticular formation (PPRF). A case of ocular flutter with a circumscribed inflammatory pontine lesion involving the PPRF, in which clinical and neuroradiological improvement occurred together, has been reported, supporting the argument that, at least in some cases, PPRF lesions may be associated with ocular flutter.

References

Schon F, Hodgson TL, Mort D, Kennard C. Ocular flutter associated with a localized lesion in the paramedian pontine reticular formation. *Annals of Neurology* 2001; **50**: 413-416

Cross References
Opsoclonus; Saccades; Saccadic intrusion, Saccadic pursuit; Square-wave jerks

Ocular Myoclonus
- see MYOCLONUS; PALATAL MYOCLONUS

Ocular Tilt Reaction
The ocular tilt reaction is a postural synkinesis consisting of the triad of:

- Ocular torsion
- Lateral head tilt to the same side
- Skew deviation with hypotropia ipsilateral to the direction of head/eye torsion

The ocular tilt reaction (OTR) is due to disordered function of one utricle or its brainstem connections (vestibular nerve, vestibular nuclei, medial longitudinal fasciculus, interstitial nucleus of Cajal), hence a brainstem otolith-ocular reflex. It has occasionally been reported with cerebellar lesions, and may be under inhibitory cerebellar control. OTR may be tonic, as in the lateral medullary syndrome, or paroxysmal, as in multiple sclerosis.

References
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Cross References
Hypotropia; Lateral medullary syndrome; Skew deviation; Synkinesia, Synkinesis; Tullio phenomenon; Vestibulo-ocular reflexes

Oculocephalic Response
Oculocephalic responses are most commonly elicited in unconscious patients; the head is passively rotated in the horizontal or vertical plane (doll's head maneuver) and the eye movements are observed. Conjugate eye movement in a direction opposite to that in which the head is turned is indicative of an intact brainstem (intact vestibulo-ocular reflexes). With pontine lesions, the oculocephalic responses may be lost, after roving eye movements but before caloric responses disappear.

Cross References
Caloric testing; Coma; Doll's head maneuver, Doll's eye maneuver; Head impulse test; Roving eye movements; Supranuclear gaze palsy; Vestibulo-ocular reflexes

Oculogyric Crisis
Oculogyric crisis is an acute dystonia of the ocular muscles, usually causing upward and lateral displacement of the eye. It is often accompanied by a disorder of attention (obsessive, persistent thoughts), with or without dystonic or dyskinetic movements. It occurs particularly

with symptomatic (secondary), as opposed to idiopathic (primary), dystonias, for example post-encephalitic and neuroleptic-induced dystonia, the latter now being the most common cause. This is usually an acute effect but may on occasion be seen as a consequence of chronic therapy (tardive oculogyric crisis).

Treatment of acute neuroleptic-induced dystonia is either parenteral benzodiazepine or an anticholinergic agent, such as procyclidine, benztropine, or trihexyphenidyl.

References

Leigh RJ, Foley JM, Remler BF, Civil RH. Oculogyric crisis: a syndrome of thought disorder and ocular deviation. *Annals of Neurology* 1987; **22**: 13-17

Cross References

Dyskinesia; Dystonia

Oculomasticatory Myorhythmia

- see MYORHYTHMIA

Oculomotor (III) Nerve Palsy

Oculomotor (III) nerve palsy produces:

- Ptosis: weakness of levator palpebrae superioris (LPS), +/– Müller's muscle;
- Mydriasis: impaired parasympathetic outflow to the pupil ("internal ophthalmoplegia");
- Diplopia: weakness of medial rectus (MR), inferior rectus (IR), superior rectus (SR), and inferior oblique (IO) muscles causing the eye to point "down and out" (external ophthalmoplegia); the presence of intorsion confirms integrity of superior oblique muscle/trochlear (IV) nerve function.

These changes may be complete or partial.

Pathological correlates of third nerve palsy may occur anywhere from the brainstem to the orbit:

- Intramedullary (brainstem):

  Nuclear: very rare; SR subnucleus lesion causes bilateral denervation; other clinical signs may be expected, such as pupillary (Edinger-Westphal nucleus) and medial longitudinal fasciculus involvement

  Fascicular (within substance of midbrain): all muscles or specific muscles involved, + other clinical signs expected, such as contralateral ataxia (Claude's syndrome), hemiparesis (Weber's syndrome)
- Extramedullary:

  Subarachnoid space: peripherally located pupillomotor fibers often spared by ischemic lesions, but not by space-occupying lesions (*e.g.,* aneurysm), however the distinction is not absolute

Cavernous sinus: III runs over trochlear nerve; other oculomotor nerves +/– trigeminal nerve often affected
Superior orbital fissure: superior division/ramus to SR, LPS; inferior to MR, IR, IO; selective involvement (divisional palsy) may occur; proptosis with space occupying lesions
Orbit: paresis of isolated muscle almost always from orbital lesion or muscle disease

Oculomotor nerve palsies may be distinguished as "pupil involving" or "pupil sparing" (*q.v.*), the former implying a "surgical," the latter a "medical" cause, but this distinction only holds for complete palsies. Incomplete palsies are more likely to be of "surgical" origin (*e.g.,* posterior communicating artery aneurysm). Imaging is the appropriate management if in doubt. Transtentorial (uncal) herniation due to raised intracranial pressure may, particularly in its early stages, cause an oculomotor nerve palsy due to stretching of the nerve, a "false-localizing sign."

References

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Cross References

Diplopia; Divisional palsy; "False-localizing signs"; Hutchinson's pupil; Mydriasis; Ophthalmoparesis, Ophthalmoplegia; Ptosis; Pupil sparing

Oculovestibular Response

- see CALORIC TESTING; VESTIBULO-OCULAR REFLEXES

One-and-a-Half Syndrome

The one-and-a-half syndrome consists of an ipsilateral horizontal gaze palsy and an ipsilateral internuclear ophthalmoplegia, such that the only preserved horizontal eye movement is abduction in one eye; vertical movements and convergence are spared. This results from a brainstem lesion which involves both the abducens (VI) nerve nucleus or paramedian pontine reticular formation, causing ipsilateral horizontal gaze palsy, and the adjacent medial longitudinal fasciculus, causing internuclear ophthalmoplegia. In young patients this is most often due to demyelination, in the elderly to brainstem ischemia; brainstem arteriovenous malformation or tumor may also be responsible.

Myasthenia gravis may cause a pseudo-one-and-a-half syndrome.

A vertical one-and-a-half syndrome has also been described, characterized by vertical upgaze palsy, and monocular paresis of downgaze, either ipsilateral or contralateral to the lesion.

References

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Wall M, Wray SH. The one-and-a-half syndrome. A unilateral disorder of the pontine tegmentum: a study of 20 cases and a review of the literature. *Neurology* 1983; **33**: 971-980

Cross References

Eight-and-a-half syndrome; Gaze palsy; Internuclear ophthalmoplegia (INO)

Onion Peel, Onion Skin

These terms have been used to describe the pattern of facial sensory loss with perioral sparing (Dejerine pattern), seen with intramedullary or cervicomedullary lesions and with tabes dorsalis. It reflects the somatotopic sensory representation in the spinal nucleus of the trigeminal nerve: midline face (nose, mouth) represented rostrally, lateral facial sensation represented caudally. The pattern of sensory impairment may also be termed "balaclava helmet."

Cross References

Balaclava helmet

Ophthalmoparesis, Ophthalmoplegia

Ophthalmoparesis is a weakness or limitation, ophthalmoplegia a paralysis, of eye movements. Causes may be central (CNS pathways), or peripheral (cranial nerve nuclei, cranial nerves, neuromuscular junction, extraocular muscles).

A distinction is sometimes drawn between:

- *External ophthalmoplegia*:
  weakness of the extraocular muscles of central, neuromuscular, or myopathic origin (*e.g.,* chronic progressive external ophthalmoplegia [CPEO], a mitochondrial disorder); and
- *Internal ophthalmoplegia*:
  fixity of the pupil with loss of all pupillary reflexes (iridoplegia) and ciliary apparatus.

Hence in an oculomotor (III) nerve palsy there may be both internal and external ophthalmoplegia.

If structural disease and myasthenia gravis are excluded, then mitochondrial disorder (CPEO) may be responsible for ophthalmoplegia, even if this is not evident on quadriceps muscle biopsy.

References

Schaefer AM, Blakely EL, Barron MJ, Griffiths PG, Taylor RW, Turnbull DM. Ophthalmoplegia: when all the tests are negative. *Journal of Neurology, Neurosurgery and Psychiatry* 2004; **75**: 519 (abstract 020)

Cross References

Diplopia; Internuclear ophthalmoplegia (INO); Miosis; Mydriasis; Oculomotor (III) nerve palsy; Pupillary reflexes; Pupil sparing

Opisthotonos

Opisthotonos is an abnormal posture consisting of arching of the back and extension of the limbs such that the body may be supported just on the head and ankles (*arc de cercle*).

Opisthotonos may be seen in:
Coma; decerebrate rigidity
Basilar meningitis
Hydrocephalus
Structural lesions of the posterior fossa
Cerebellar fits due to intermittent tonsillar herniation
Acute drug- (neuroleptic-) induced dystonic reaction; or chronic feature of tardive dystonia
Tetanus
Syncope (especially in children)
Metabolic disorders: kernicterus, Gaucher's disease (type II)
Drug-induced: propofol
Pseudoseizures

As in decerebrate rigidity, opisthotonos may reflect unopposed extensor tone from the intact vestibular nuclei released from supratentorial control.

Cross References

Coma; Decerebrate rigidity; Emposthotonos

Oppenheim's Sign

Oppenheim's sign is a variant method for eliciting the plantar response, by application of heavy pressure to the anterior surface of the tibia, for example with the thumb, moving from patella to ankle. Extension of the hallux (upgoing plantar response, Babinski's sign) is pathological. Like Chaddock's sign, Oppenheim's sign always postdates the development of Babinski's sign as a reliable indicator of corticospinal pathway (upper motor neurone) pathology.

References

Pearce JMS. Oppenheim's sign. In: Pearce JMS. *Fragments of neurological history*. London: Imperial College Press, 2003: 359-361

Van Gijn J. *The Babinski sign: a centenary*. Utrecht: Universiteit Utrecht, 1996

Cross References

Babinski's sign (1); Chaddock's sign; Gordon's sign; Plantar response; Upper motor neurone (UMN) syndrome

Oppenheim's Useless Hand Sign

- see USELESS HAND OF OPPENHEIM

Opsoclonus

Opsoclonus, or saccadomania, is an eye movement disorder characterized by involuntary bursts of polydirectional saccades (sometimes with a horizontal preference) without an intersaccadic interval (*cf*. square-wave jerks). Like ocular flutter, opsoclonus may be accurately characterized with oculography.

Although some normal individuals can voluntarily induce opsoclonus, generally it reflects mesencephalic or cerebellar disease affecting the omnipause cells which exert tonic inhibition of the burst neurones which generate saccades.

Recognized causes of opsoclonus include:

- Paraneoplasia: in children with neuroblastoma (Kinsbourne's syndrome); in adults the opsoclonus-myoclonus syndrome is most commonly associated with small-cell lung cancer but it may also occur in association with breast cancer in which case onco neural antibodies (anti-Ri, or type 2 antineuronal nuclear antibodies [ANNA-2]) may be detected in serum and CSF;
- Postinfectious: a monophasic disorder following respiratory or gastrointestinal infection;
- Intraparenchymal (especially mesencephalic) lesions, *e.g.,* tumor, demyelination, sarcoidosis, metabolic/toxic encephalopathy.

Postinfectious opsoclonus generally remits spontaneously. Of the paraneoplastic disorders, opsoclonus associated with lung and breast tumors persists and the patients decline from their underlying illness; neuroblastoma associated opsoclonus may be steroid responsive.

Cross References

Ocular flutter; Saccadic intrusion, Saccadic pursuit; Square-wave jerks

Optic Aphasia

Optic aphasia is a visual modality-specific naming disorder. It has sometimes been grouped with associative visual agnosia, but these patients are not agnosic since they can demonstrate recognition of visually-presented stimuli by means other than naming, *e.g.*, gesture. Moreover, these patients are not handicapped by their deficit in everyday life, whereas agnosic patients are often functionally blind. Objects that are semantically related can be appropriately sorted, indicating intact semantics. This is not simply anomia, since the deficit is specific to visual stimuli; objects presented in tactile modality, or by sound, or by spoken definition can be named. Naming errors are often semantic, and perseverations ("*conduit d'approche*") are common. Perception is intact, evidenced by the ability to draw accurately objects which cannot be named. Reading is poorly performed.

Optic aphasia is associated with unilateral lesions of the left occipital cortex and subjacent white matter.

The neuropsychological explanation of optic aphasia is unclear. It may be a mild type of associative visual agnosia, despite the differences.

References

Beauvois MF. Optic aphasia: a process of interaction between vision and language. *Philosophical Transactions of the Royal Society, Series B* 1982; **298**: 35-47

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

Lhermitte F, Beauvois MF. A visual-speech disconnection syndrome: report of a case with optic aphasia, agnosic alexia and color agnosia. *Brain* 1973; **96**: 695-714

Cross References
Anomia; Conduit d'approche; Visual agnosia

Optic Ataxia

Optic ataxia is impaired voluntary reaching for a visually presented target, with misdirection and dysmetria. It may resemble cerebellar ataxia. Visual fixation is possible but reaching under visual guidance is impaired. Tactile search with the palm and fingers may be undertaken in searching for an object, using somatosensory cues to compensate for impaired access to visual information. Hence this may be characterized as a modality-specific apraxia, wherein visual information cannot be used to guide goal-directed movements. The disorder is both retinotopic and somatotopic.

Optic ataxia occurs with lesions of the intraparietal sulcus and regions medial and superior to it; the primary visual cortex is intact. It is one feature, along with psychic paralysis of gaze ("sticky fixation") and simultanagnosia (visual disorientation), of Balint's syndrome in which there is some evidence for parieto-occipital (and possibly frontal) lobe dysfunction (disconnection).

References
Perenin MT, Vighetto A. Optic ataxia: a specific disruption in visuomotor mechanisms. I. Different aspects of the deficit in reaching for objects. *Brain* 1988; **111**: 643-674

Cross References
Apraxia; Ataxia; Balint's syndrome; Dysmetria; Simultanagnosia; Visual disorientation; Visual form agnosia

Optic Atrophy

Optic atrophy is pallor of the optic nerve head as visualized by ophthalmoscopy. The temporal disc may appear pale in a normal fundus, so that optic atrophy can only be confidently diagnosed when there is also nasal pallor, although temporal pallor may follow damage to the macular fibre bundle with central visual defects.

Optic atrophy may be the consequence of any optic neuropathy which causes optic nerve damage leading to gliotic change of the optic nerve head. The appearance of optic atrophy is nonspecific with respect to etiology. Common causes include previous optic neuritis and chronic papilledema, but retinal lesions, optic chiasm and optic tract pathologies can all produce optic atrophy (*e.g.*, inherited optic neuropathies, tobacco-alcohol amblyopia; vitamin B_{12} deficiency). "Hemianopic" optic atrophy, indicates involvement of the optic tract or lateral geniculate body.

Cross References
Disc swelling; Papilledema; Temporal pallor

Optokinetic Nystagmus (OKN), Optokinetic Response

Optokinetic nystagmus (OKN) is familiar to anyone who has watched a railway passenger observing passing telegraph poles from the window

of a moving train: OKN is an involuntary rhythmic eye movement induced by observing moving stimuli. In clinical practice a striped drum serves to test both visual pursuit and saccades. Rotation of the stripe to the left produces leftward pursuit, followed by a compensatory saccade to the right, followed by pursuit to the left of the next stripe, with another compensatory saccade, and so on. Hence, OKN is a physiological nystagmus.

Parietal hemisphere lesions (vascular or neoplastic) typically impair OKN. Testing for OKN may be useful in patients with suspected hysterical visual loss, since OKN cannot occur unless visual function is present; the response is lost in blindness. An internuclear ophthalmoplegia may be made more evident by testing OKN.

Cross References

Cortical blindness; Internuclear ophthalmoplegia (INO); Nystagmus; Saccades; Vestibulo-ocular reflexes

Orofacial Dyspraxia

Orofacial dyspraxia, or buccofacial dyspraxia, is an inability to make voluntary, learned, movements with the orofacial musculature, such as blowing out a match, kissing, licking the lips.

Recognized causes of orofacial dyspraxia include:

- Transient accompaniment of Broca's aphasia, conduction aphasia, and transcortical motor aphasia of cerebrovascular origin
- Trauma to pre-Rolandic area just above the Sylvian fissure
- In some patients with primary progressive aphasia; a related but distinct condition of "progressive loss of speech output with orofacial dyspraxia" has also been described.

Clinical and imaging studies show a strong correlation between orofacial dyspraxia and lesions in the frontal operculum; it may also occur with subcortical lesions involving periventricular and/or peristriatal white matter as well as the basal ganglia.

References

Tyrrell PJ, Kartsounis LD, Frackowiak RSJ, Findley LJ, Rossor MN. Progressive loss of speech output and orofacial dyspraxia associated with frontal lobe hypometabolism. *Journal of Neurology, Neurosurgery and Psychiatry* 1991; **54**: 351-357

Cross References

Apraxia

Orthostatic Hypotension

Orthostatic hypotension or postural hypotension is the finding of a persistent drop in blood pressure on standing, defined as a greater than 20 mmHg fall in systolic pressure and/or a 5 mmHg fall in diastolic pressure one minute after a change from the supine to the upright position. Normally there is a drop in blood pressure of lesser magnitude on standing but this is usually quickly compensated for by the baroreceptor reflex. To demonstrate orthostatic hypotension, it may be necessary

to measure blood pressure not only on immediate standing but also after two to ten minutes, since the fall may be delayed. Measuring blood pressure automatically on a tilt table is also helpful in diagnosing orthostatic hypotension.

Symptoms which may be associated with orthostatic hypotension include exercise-induced or postprandial light-headedness, transient visual loss, blackouts (syncope), and pain in a "coathanger" distribution across the shoulders. There may be supine hypertension and reversal of the normal circadian blood pressure rhythm (normally lower at night), with increased frequency of micturition at night. Other features of autonomic dysfunction may be present, including dry eyes and dry mouth (xerophthalmia, xerostomia), a tendency to constipation, and lack of penile erections.

Orthostatic hypotension may be found in:

- Pure autonomic neuropathy
- Neurodegenerative disorders, such as multiple system atrophy, Parkinson's disease, dementia with Lewy bodies
- Phaeochromocytoma
- Other causes of autonomic neuropathy (*e.g.*, Guillain-Barré syndrome, amyloidosis).

However, the most common cause of orthostatic hypotension in hospital practice is probably dehydration or overzealous treatment with antihypertensive or diuretic agents.

Treatments for pure autonomic failure encompass both nonpharmacological approaches (*e.g.*, increased salt intake, head-up bed tilt, wearing a G-suit) and pharmacological therapies, including fludrocortisone, ephedrine, and midodrine.

References

Mathias CJ, Kimber JR. Treatment of postural hypotension. *Journal of Neurology, Neurosurgery and Psychiatry* 1998; **65**: 285-289

Cross References

Neuropathy; Parkinsonism; Xerophthalmia, Xerostomia

Oscillopsia

Oscillopsia is an illusory movement of the environment due to excessive slip of images on the retina ("retinal slip") during active or passive head movement, producing a complaint of blurring, jumping, or oscillation of the visual representation of the environment. Oscillopsia is most often due to acquired bilateral loss of vestibular function (loss of the vestibulo-ocular reflexes). Other recognized causes of oscillopsia include:

- Acquired nystagmus
- Superior oblique myokymia
- Other ocular oscillations.

Oscillopsia does not occur in congenital nystagmus, nor in opsoclonus, presumably due to the operation of the visual suppression mechanism which normally operates during saccadic eye movements.

Oscillopsia may be treated with clonazepam; if due to acquired pendular nystagmus anticholinesterase or alcohol may help.

References

Leigh RJ. Oscillopsia: impaired vision during motion in the absence of the vestibulo-ocular reflex. *Journal of Neurology, Neurosurgery and Psychiatry* 1998; **65**: 808

Cross References

Myokymia; Nystagmus; Opsoclonus; Vestibulo-ocular reflexes

Overflow

- see DYSTONIA; SYNKINESIA, SYNKINESIS

P

Pagophagia
- see PICA

Palatal Myoclonus
Palatal myoclonus, also known as palatal tremor, is a focal myoclonic syndrome characterized by rhythmic, unilateral or bilateral, palatal contractions which continue during sleep. This may be asymptomatic, or there may be a clicking sound in the inner ear (especially in essential palatal myoclonus). There may be associated contractions of external ocular muscles (oculopalatal myoclonus), larynx, neck, diaphragm (respiratory myoclonus, diaphragmatic flutter, or Leeuwenhoek's disease), trunk, and limbs, which may bring the palatal myoclonus to attention. Palatal myoclonus may be accompanied by pendular nystagmus and oscillopsia.

Palatal myoclonus is associated with lesions interrupting pathways between the red nucleus, inferior olivary nucleus and dentate nucleus (Guillain-Mollaret triangle). Hypertrophy of the inferior olivary nucleus may be evident neuroradiologically (structural or functional imaging) and pathologically. This is a consequence of a lesion in the dentato-olivary pathway which leads to transsynaptic degeneration and hypermetabolism of the olivary nucleus. Although many cases are essential/idiopathic, recognized symptomatic causes of palatal myoclonus include vascular lesions, trauma, neoplasia, demyelination, epilepsy and, rarely, adult-onset Alexander's disease.

Drug treatment of palatal myoclonus is often unsuccessful, although reports of benefit with 5-hydroxytryptophan, carbamazepine, sodium valproate, clonazepam, baclofen, and even sumatriptan have appeared. Botulinum toxin injections may also help.

Cross References
Eight-and-a-half syndrome; Myoclonus; Nystagmus; Oscillopsia; Tinnitus

Palatal Reflex
- see GAG REFLEX

Palilalia
Palilalia is a disorder of articulation characterized by the involuntary repetition of syllables within a word, whole words, or phrases, hence a reiterative speech disorder. The term stutter may be used for repetition of single syllables, and the term palilogia has sometimes been used for the repetition of phrases, to distinguish from palilalia. These phenomena may be encountered in:

Parkinson's disease (along with bradylalia, slowness of speech)
Progressive supranuclear palsy
Gilles de la Tourette syndrome (along with vocal and motor tics)
Pick's disease, as part of the so-called PES syndrome (palilalia, echolalia, stereotypy) or the PEMA syndrome (palilalia, echolalia, mutism, amimia)
Late stages of Alzheimer's disease
Postencephalitic parkinsonism (von Economo's disease)
Fahr's disease (bilateral basal ganglia calcification)
Normal finding in children below the age of about six years

In pathological states, palilalia may reflect difficulty in set shifting, as seen in frontal lobe (frontal convexity) syndromes.

Cross References

Bradylalia; Echolalia; Frontal lobe syndromes; Hypomimia; Mutism; Parkinsonism; Stereotypy; Stutter; Tic

Palilogia

- see PALILALIA

Palinacusis

Palinacusis, or palinacousis, is the persistence of prior auditory perception. Although sometimes classified as an illusory experience, musical hallucinations may occur concurrently. The symptom may be related to seizures of temporal lobe origin.

References

Takeshi T, Matsunaga K. Musical hallucinations and palinacousis. *Psychopathology* 1999; **32**: 57-59

Cross References

Hallucination; Illusion

Palinopsia

Palinopsia is an illusory visual phenomenon characterized by the persistence or recurrence of visual images immediately after the stimulus has been removed, hence visual perseveration. This is distinct from the physiological after-image. It may be associated with polyopia. The description of the symptom may lead to it being mistaken for diplopia ("pseudodiplopia").

Palinopsia occurs most frequently in the context of a left homonymous hemianopia, secondary to right occipitotemporal or occipitoparietal lesions: these may be vascular, neoplastic, metabolic, ictal, or drug- or toxin-induced (*e.g.*, carbon monoxide poisoning). It has also been described with retinal and optic nerve disease, and occasionally in normal individuals.

References

Michel EN, Troost BT. Palinopsia: cerebral localization with computed tomography. *Neurology* 1980; **30**: 887-889
Pomeranz HD, Lessell S. Palinopsia and polyopia in the absence of drugs or cerebral disease. *Neurology* 2000; **54**: 855-859

Smith PEM, Shah P, Sharpe J, Todd A, Goringe AP. Palinopsia. *Lancet* 2003; **361**: 1098
Stagno SJ, Gates TJ. Palinopsia: a review of the literature. *Behavioral Neurology* 1991; **4**: 67-74
Cross References
Hemianopia; Illusion; Perseveration; Polyopia

Pallesthesia

Pallesthesia is the appreciation of vibration sensation; its loss pallanesthesia.
Cross References
Vibration

Palmomental Reflex

The palmomental reflex consists of contraction of the mentalis muscle induced by stroking the ipsilateral palm with a blunt object. It may indicate damage to the contralateral paracentral cortex or its connections, but since it is observed in about one quarter of normal adults and is very common in the normal elderly, and may occur in other conditions, both its sensitivity and specificity are low. It may be considered a frontal release sign or primitive reflex, but is less specific than the grasp reflex. Induction of the reflex by stimulation of areas other than the palm is more likely to be associated with cerebral damage.
References
Owen G, Mulley GP. The palmomental reflex: a useful clinical sign? *Journal of Neurology, Neurosurgery and Psychiatry* 2002; **73**: 113-115
Cross References
Age-related signs; Frontal release signs

Papilledema

Papilledema is swelling (edema) of the optic nerve head due to raised intracranial pressure (*cf*. other causes of disc swelling, which may cause pseudopapilledema).

A number of stages of papilledema are described: in the acute stage, the only findings may be edema at the superior and inferior poles of the disc, absence of spontaneous venous pulsation, and enlargement of the blind spot. As papilledema progresses the whole disc is involved and splinter hemorrhages may be evident at the disc margin. These early stages may be asymptomatic, or may be associated with transient losses of vision (obscurations), often provoked by activities or movements which further raise intracranial pressure, thus compromising retinal perfusion pressure. Enlargement of the blind spot and constriction of the visual field may be evident, but visual acuity is often unimpaired (*cf*. disc swelling due to papillitis). Chronic papilledema produces gliosis of the optic nerve head and eventually optic atrophy ("sequential optic atrophy") with nerve fibre damage and permanent visual field defects.
Cross References
Blind spot; Disc swelling; Obscurations; Optic atrophy pseudopapilledema; Retinal venous pulsation; Scotoma

Paraballismus
- see BALLISM, BALLISMUS; HEMIBALLISMUS

Paradoxical Breathing
The normal movement of the diaphragm (*i.e.*, down in inspiration, causing outward abdominal wall movement) may be reversed (paradoxical) in conditions which cause diaphragm weakness (*i.e.*, inward abdominal wall movement on inspiration), *e.g.*, Guillain-Barré syndrome, acid-maltase deficiency, phrenic nerve injury, hence paradoxical abdominal movement, abdominal paradox, paradoxical breathing, or paradoxical diaphragm movement. This may be detectable clinically or by X-ray screening of the diaphragm. Vital capacity is lower when lying compared to standing. Paradoxical diaphragm movement is a potentially alarming sign since it may indicate incipient respiratory failure.

The term paradoxical breathing may also be used to describe thorax and abdomen moving in different directions when breathing, as with increased upper airway resistance.

Cross References
Myopathy

Paradoxical Flexor Reflex
- see GORDON'S SIGN

Paradoxical Head Tilt
- see BIELSCHOWSKY'S SIGN, BIELSCHOWSKY'S TEST

Paradoxical Triceps Reflex
- see INVERTED REFLEXES

Paragrammatism
- see WERNICKE'S APHASIA

Paragraphia
- see AGRAPHIA

Parakinesia, Parakinesis
These terms have been used in different ways by different authors, to describe:

- A volitional purposeful act designed to camouflage or draw attention away from an involuntary movement, such as chorea;
- Strange movements of presumed psychogenic origin. It should be remembered that many movements previously thought to conform to this definition have subsequently been recognized to have an organic basis (*e.g.*, klazomania).

The terms are now seldom used.

Cross References
Chorea, Choreoathetosis; Dyskinesia; Klazomania

Paralexia
- see ALEXIA

Paralogia
- see GANSER PHENOMENON

Paralysis
Paralysis is a total loss of power to move a body part; equivalent to the suffix -plegia. The use of the word has not been entirely consistent, for example *paralysis agitans* originally used by James Parkinson to describe the disease which now bears his name.

The periodic paralyses are a group of conditions characterized by episodic muscular weakness and stiffness (myotonia) associated with mutations in the skeletal muscle voltage-gated sodium and calcium ion channel genes (channelopathies).

Cross References
Myotonia; Plegia

Paramnesia
Paramnesia is recalling as memories things which have not in fact taken place, hence a distortion of episodic or autobiographical memory. This may be neurological or psychiatric in origin. Relation of paramnesias as the truth occurs in confabulation.

Cross References
Amnesia; Confabulation; Reduplicative paramnesia

Paramyotonia
Paramyotonia is similar to myotonia in that muscle does not relax normally following contraction (voluntary, percussion), which may prompt a complaint of muscle aching or stiffness, but differs in that repetitive muscle use (*e.g.*, exercise) accentuates the problem, leading to an increased delay in muscle relaxation (worsening stiffness). For example, repeated forced voluntary eyelid closure in a patient with paramyotonia may, after several attempts, lead to a failure of voluntary eyelid opening, the eyes remaining closed for a minute or so. Paramyotonia particularly affects the face and forearms. This type of muscle stiffness may also be sensitive to temperature, being made worse by cooling which may also provoke muscle weakness. Weakness may outlast exposure to cold by several hours.

Neurophysiological studies may assist in the diagnosis of paramyotonia. During the delayed muscle relaxation, electrical activity is not prominent, and after muscle cooling the resting muscle membrane potential may be reduced from around the normal –80 mV to –40 mV, at which point muscle fibers are inexcitable (contracture).

Paramyotonia congenita is a channelopathy with mutations affecting the α-subunit of the sodium channel. Mutations in the same gene have been documented in hyperkalaemic periodic paralysis and K^+-aggravated myotonia.

References
Davies NP, Eunson LH, Gregory RP, Mills KR, Morrison PJ, Hanna MG. Clinical, electrophysiological, and molecular genetic studies in a new family with paramyotonia. *Journal of Neurology, Neurosurgery and Psychiatry* 2000; **68**: 504-507 [erratum: *Journal of Neurology, Neurosurgery and Psychiatry* 2000; **69**: 139]
Ebers GC, George AL, Barchi RL, *et al*. Paramyotonia congenita and hyperkaliemic periodic paralysis are linked to the adult muscle sodium channel gene. *Annals of Neurology* 1991; **30**: 810-816
Cross References
Contracture; Myotonia; Paralysis; Warm-up phenomenon

Paraparesis

Paraparesis is a weakness of the lower limbs, short of complete weakness (paraplegia, *q.v.*). This may result from lesions anywhere from cerebral cortex (frontal, parasagittal lesions) to peripheral nerves, producing either an upper motor neurone (spastic) or lower motor neurone (flaccid) picture. A spinal cord lesion (myelopathy) is probably the most common cause. Paraparesis may be symmetrical or asymmetrical.
Cross References
Flaccidity; Myelopathy; Paraplegia; Spasticity

Paraphasia

Paraphasias are a feature of aphasias (disorders of language), particularly (but not exclusively) fluent aphasias resulting from posterior dominant temporal lobe lesions (*cf*. anterior lesions which tend to produce nonfluent aphasias with agrammatism). Paraphasias refer to a range of speech output errors including substitution, addition, duplication, omission and transposition of linguistic units, affecting letters within words, letters within syllables, or words within sentences. Paraphasic errors may be categorized as:

- *Phonemic* or *literal*:
  Errors involve individual phonemes; impaired phonology (*i.e*., sound based) causing approximations to real words; nonwords resulting from phonemic paraphasia may be referred to as neologisms. Phonemic paraphasias may be encountered in Broca's aphasia and conduction aphasia, when the patient may recognize them to be errors, and Wernicke's aphasia.
- *Formal*:
  Target word is replaced by another word that is phonemically similar.
- *Morphemic*:
  Errors involving word stems, suffixes, prefixes, inflections and other parts of words.
- *Verbal*:
  Errors involving whole words. These may be further classified as:

Semantic or *categoric*: substitution of a different exemplar from the same category (*e.g.*, "orange" for "apple"; *paradigmatic*) or of a thematically related word (*e.g.*, "sit" for "chair"; *syntagmatic*). Verbal paraphasias showing both semantic and phonemic resemblance to the target word are called *mixed* errors. These types may be observed in patients with Wernicke's aphasia, who often seem unaware of their paraphasias due to a failure of self-monitoring of output.

References

Buckingham HW, Kertesz A. *Neologistic jargon aphasia*. Amsterdam: Swets & Zeitlinger, 1976

Cross References

Aphasia; Broca's aphasia; Conduction aphasia; Jargon aphasia; Neologism; Schizophasia; Transcortical aphasias; Wernicke's aphasia

Paraplegia

Paraplegia is a total weakness (paralysis) of the lower limbs (*cf.* paraparesis). This may result from lower motor neurone lesions involving multiple nerve roots and/or peripheral nerves (*e.g.*, paraparetic Guillain-Barré syndrome) producing a flaccid, areflexic paraplegia; but more commonly it is due to upper motor neurone lesions interrupting corticospinal pathways (corticospinal tract, vestibulospinal tract, reticulospinal tracts, and other extrapyramidal pathways), most usually in the spinal cord. The latter may acutely produce a flaccid areflexic picture ("spinal shock"), but later this develops into an upper motor neurone syndrome (hypertonia, clonus, hyperreflexia, loss of superficial reflexes [*e.g.*, abdominal, cremasteric reflexes] and Babinski's sign) with possible lower motor neurone signs at the level of the lesion; bladder involvement is common (urinary retention). Because of the difficulty in distinguishing whether an acute paraplegia is of LMN or UMN origin, imaging to exclude potentially reversible cord compression is mandatory.

Recognized causes of paraplegia of upper motor neurone origin include:

- Traumatic section of the cord
- Cord compression
- Inflammatory lesions: acute transverse myelitis of viral origin, multiple sclerosis, neuromyelitis optica (Devic's syndrome)
- Ischemic lesions; anterior spinal artery syndrome, venous infarction of the cord.

In paraplegia of upper motor neurone origin, enhanced flexion defense reflexes ("flexor spasms") may occur, producing hip and knee flexion, ankle and toe dorsiflexion. Eventually such flexor responses may become a fixed flexion deformity with secondary contractures ("paraplegia in flexion"). Prevention of this situation may be possible by avoiding spasms, which are often provoked by skin irritation or ulceration, bowel constipation, bladder infection, and poor nutrition. Physiotherapy and pharmacotherapy with agents, such as baclofen,

dantrolene, and tizanidine may be used; botulinum toxin injections may be helpful for focal spasticity.

"Paraplegia in extension," with extension at the hip and knee, may be seen with incomplete or high spinal cord lesions.

References

Johnston RA. Acute spinal cord compression. In: Hughes RAC (ed.). *Neurological Emergencies* (2nd edition). London: BMJ Publishing, 1997: 272-294

Passmore AP, Taylor IC, McConnell JG. Acute Guillain-Barré syndrome presenting as acute spinal cord compression in an elderly woman. *Journal of the Royal Society of Medicine* 1990; **83**: 333-334

Cross References

Abdominal reflexes; Areflexia; Babinski's sign (1); Clonus; Contracture; Cremasteric reflex; Flaccidity; Hyperreflexia; Hypertonia, Hypertonus; Lower motor neurone (LMN) syndrome; Myelopathy; Paraparesis; Spasticity; Upper motor neurone (UMN) syndrome; Urinary retention

Paratonia

- see *GEGENHALTEN*

Paresis

Paresis denotes a weakness which is less than total paralysis (-plegia), which may be of upper or lower motor neurone origin. Various prefixes denote the location of such weakness, *e.g.*, hemiparesis, monoparesis, ophthalmoparesis, paraparesis, quadriparesis (*q.v.*).

Since localized pain may inhibit voluntary muscular exertion, apparent weakness in such circumstances may be labeled "algesic pseudoparesis."

Cross References

Lower motor neurone (LMN) syndrome; Plegia; Upper motor neurone (UMN) syndrome; Weakness

Paresthesia

Paresthesia is an abnormal sensation, often described as a tingling sensation, or likened to "pins and needles" or electricity, pricking, or even crawling (formication), *i.e.*, positive sensory symptoms. The sensation is not pleasant but nor is it painful (*cf.* dysesthesia). Some patients may describe this sensation as "numbness" or "deadness," in which case care needs to be taken to differentiate it from anesthesia (*i.e.*, a negative phenomenon). Some authorities reserve the term for spontaneous rather than evoked positive sensory phenomena, as a distinction from dysesthesia.

Paresthesia is a feature of neuropathy, and may occur in the distribution of a compressed or entrapped nerve, perhaps reflecting the mechanosensitivity of nerves in this situation (*e.g.*, Phalen's sign, Tinel's sign). Paresthesia is a more reliable indicator of the diagnosis of neuropathy than pain. Paresthesia may also be provoked by hyperventilation (especially perioral, hands and feet [acroparesthesia]). Central lesions may also produce paresthesia (*e.g.*, Lhermitte's sign).

References
Larner AJ. Missed diagnosis of vitamin B_{12} deficiency presenting with paresthetic symptoms. *International Journal of Clinical Practice* 2002; **56**: 377-378
Cross References
Anesthesia; Dysesthesia; Lhermitte's sign; Phalen's sign; Tinel's sign

Parinaud's Syndrome

Parinaud's syndrome, or the dorsal midbrain syndrome, or the pretectal syndrome, consists of:

- Paralysis of vertical gaze, especially upgaze, with or without mydriasis
- Loss of pupillary light reflexes (light-near dissociation)
- Loss of convergence
- Convergence or retraction nystagmus
- Upper eyelid retraction (Collier's "tucked lid" sign).

Bell's phenomenon is usually lost.

This constellation of signs results from dorsal midbrain lesions, such as pineal tumors, which affect the pretectum and posterior commissure and so interfere with conjugate eye movements in the vertical plane.

References
Keane JR. The pretectal syndrome: 206 patients. *Neurology* 1990; **40**: 684-690
Parinaud H. Paralysie des mouvements associés des yeux. *Archives de Neurologie Paris* 1883; **5**: 145-172
Pearce JMS. Parinaud's syndrome. *Journal of Neurology, Neurosurgery and Psychiatry* 2005; **76**: 99
Pierrot-Deseilligny C, Chain F, Gray M *et al*. Parinaud's syndrome. *Brain* 1982; **105**: 667-696
Cross References
Collier's sign; Light-near pupillary dissociation; Nystagmus; Supranuclear gaze palsy

Parkinsonism

Parkinsonism is a clinical syndrome characterized by the presence of some or all of the following features; there is overlap with so-called akinetic-rigid syndromes in which these features predominate:

- Akinesia, hypokinesia (*sine qua non*)
- Rigidity: consistent (lead-pipe) or jerky (cogwheeling; Negro's sign)
- Bradykinesia
- Tremor, usually at rest, of frequency 3.5-7.0 Hz, "pill rolling" type; there may sometimes be an additional action component to the tremor, and very occasionally there is exclusively an action tremor
- Stooped posture: forward flexion of trunk, flexion of knees, elbows; "simian posture"

Impaired postural reflexes, with or without a history of falls; propulsion, retropulsion
Mask-like facies, poverty of facial expression (hypomimia)
Reduced blink rate (this may be a particular feature of progressive supranuclear palsy)
Hypophonic, monotonic voice (hypokinetic dysarthria)
Widened palpebral fissure (Stellwag's sign)
Hypometria
Seborrhea
Sialorrhea
Festinant (shuffling) gait
Micrographia
Dystonic postures, for example, striatal toe
Apraxia
Akathisia
Cognitive impairment (usually of frontal-subcortical type)
Hallucinations: minor (*anwesenheit*; passage type), or formed, visual > auditory
Autonomic dysfunction, especially orthostatic hypotension

Conventionally parkinsonism is viewed as a disorder of the extrapyramidal system producing "extrapyramidal signs," although this term has limitations: despite the fact that some of the cardinal features of parkinsonism (bradykinesia, rigidity, postural instability, tremor) result from pathology in the basal ganglia, particularly affecting dopaminergic pathways, other features may reflect cortical involvement, at least in part (*e.g.*, apraxia, micrographia).

The incidence of parkinsonism increases dramatically with age; it is also associated with an increased risk of death, particularly in the presence of a gait disturbance.

The differential diagnosis of parkinsonism is broad, and includes:

Idiopathic Parkinson's disease
Multiple system atrophy
Progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome)
Corticobasal degeneration, cortical basal ganglionic degeneration
Drug-induced parkinsonism (*e.g.*, neuroleptics, MPTP)
Toxin-induced parkinsonism (*e.g.*, carbon monoxide, manganese)
Wilson's disease (hepatolenticular degeneration)
Dementia with Lewy bodies
Neuroleptic malignant syndrome
Normal pressure hydrocephalus
"Arteriosclerotic parkinsonism," resulting from multiple subcortical infarcts
Huntington's disease, especially juvenile onset (Westphal variant)
Post-encephalitic parkinsonism (encephalitis lethargica, von Economo's disease)
Dementia pugilistica, post-traumatic parkinsonism
Systemic lupus erythematosus

Sjögren's syndrome
Hypoparathyroidism
Parkinsonism-dementia complex of Guam

Obsessive slowness also enters the differential diagnosis but typical parkinsonian features (akinesia, rigidity) are not present in this condition.

It is crucial not to miss the diagnosis of Wilson's disease, although rare, since in the early stages this disorder is reversible with copper chelation therapy; hence copper and ceruloplasmin should be checked in all patients with young-onset (under age 50) parkinsonism (and dystonia).

Response to levodopa therapy is only reliably seen in idiopathic Parkinson's disease, although some patients with multiple system atrophy or progressive supranuclear palsy may benefit. The features particularly responsive in Parkinson's disease are bradykinesia and rigidity; tremor is less reliably helped.

References

Bennett DA, Beckett LA, Murray AM, *et al*. Prevalence of parkinsonian signs and associated mortality in a community population of older people. *New England Journal of Medicine* 1996; **334**: 71-76

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Gardner-Thorpe C. *James Parkinson 1755-1824*. Exeter: A Wheaton & Co. Ltd, 1987 [includes facsimile of Parkinson's book on the shaking palsy]

Litvan I, Agid Y, Calne D, *et al*. Clinical research criteria for the diagnosis of progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome): report of the NINDS-SPSP international workshop. *Neurology* 1996; **47**: 1-9

Oertel WH, Quinn NP. Parkinsonism. In: Brandt T, Caplan LR, Dichgans J, Diener HC, Kennard C (eds.). *Neurological disorders: course and treatment*. San Diego: Academic Press, 1996: 715-772

Cross References

Apraxia; Bradykinesia; Dysarthria; Dystonia; Hypokinesia; Hypomimia; Hypophonia; Mask-like facies; Micrographia; Orthostatic hypotension; Postural reflexes; Rigidity; Seborrhea; Sialorrhea; Striatal toe; Supranuclear gaze palsy; Tremor

Parosmia

Parosmia is a false smell, *i.e*., the subjective sensation of a smell which does not exist (*i.e*., an hallucination). Such smells are usually unpleasant (cacosmia), may be associated with a disagreeable taste (cacogeusia), and may be difficult for the patient to define. Causes include purulent nasal infections or sinusitis, and partial recovery following transection of olfactory nerve fibers after head injury. Transient parosmia may presage seizures of temporal lobe cortical origin (olfactory aura), particularly involving the medial (uncal) region.

Cross References
Aura; Seizures

Parry-Romberg Syndrome
- see HEMIFACIAL ATROPHY

Past-Pointing
- see DYSMETRIA

Patellar Reflex
- see REFLEXES

Pathological Crying, Pathological Laughter
Pathological laughter and pathological crying (PLC), or forced laughter and crying, have been defined as reflecting an incongruence of mood (subjective feeling) and expression or affect ("objective," observed), such that patients laugh involuntarily though not happy, or cry though not sad. There may be a sense that the patient is struggling against these displays of emotion, in contrast to the situation in other forms of emotional lability where there is said to be congruence of mood and affect, although sudden fluctuations and exaggerated emotional expression are common to both, suggesting a degree of overlap.

PLC are ascribed to a loss (release) of the normal inhibition of the motor component of facial expression (*i.e.*, cortical-subcortical disinhibition). PLC may occur in the context of a pseudobulbar palsy ("pseudobulbar affect") but not invariably so.
PLC has been reported in:

Multiple sclerosis: crying > laughing; related to intellectual impairment (more extensive brain involvement, but not brainstem)
Alzheimer's disease
Stroke: PLC may be the harbinger of brainstem stroke or a feature of anterior choroidal artery territory infarctions; rarely a feature of TIAs
Motor neurone disease
Head injury
Gelastic epilepsy.

Suggested treatments for PLC include:
Amitriptyline
Levodopa
Amantadine
Serotonin reuptake inhibitors: fluoxetine, citalopram.

References
Feinstein A. *The clinical neuropsychiatry of multiple sclerosis*. Cambridge: Cambridge University Press, 1999: 65-79
Larner AJ. Crying spells as symptoms of a transient ischemic attack. *Journal of Neurology, Neurosurgery and Psychiatry* 2000; **68**: 800-801

Robinson RG. *The clinical neuropsychiatry of stroke: cognitive, behavioral and emotional disorders following vascular brain injury*. Cambridge: Cambridge University Press, 1998: 455-471
Wild B, Rodden FA, Grodd W, Ruch W. Neural correlates of laughter and humor. *Brain* 2003; **126**: 2121-2138
Cross References
Automatism; Emotionalism, Emotional lability; Pseudobulbar palsy

"Peek Sign"
One of the eye signs of myasthenia gravis: on attempted forced eye closure, orbicularis oculi may fatigue, such that the patient "peeks" through the partially open palpebral fissure.

Pelopsia
A form of metamorphopsia characterized by the misperception of objects as closer to the observer than they really are (*cf*. porropsia).
Cross References
Metamorphopsia; Porropsia

Pelvic Thrusting
Pelvic thrusting may be a feature of seizures of frontal lobe origin; occasionally it may occur in temporal lobe seizures. Pelvic thrusting also occurs in pseudoseizures, particularly those of the "thrashing" variety.

Choreiform disorders may involve the pelvic region causing thrusting or rocking movements.
References
Geyer JD, Payne TA, Drury I. The value of pelvic thrusting in the diagnosis of seizures and pseudoseizures. *Neurology* 2000; **54**: 227-229
Cross References
Automatism; Chorea, Choreoathetosis; Seizure

Pendular Nystagmus
- see NYSTAGMUS

Percussion Myotonia
Percussion myotonia is the myotonic response of a muscle to a mechanical stimulus, *e.g*., when struck with a tendon hammer. For example, a blow to the thenar eminence may produce involuntary and sustained flexion of the thumb. This response, which may be seen in myotonic dystrophy, reflects the impaired muscle relaxation which characterizes myotonia.
Cross References
Myotonia

Periodic Alternating Nystagmus
- see NYSTAGMUS

Periodic Respiration
Periodic respiration is a cyclical waxing and waning of the depth and rate of breathing (Cheyne-Stokes breathing or respiration), over about

two minutes, the crescendo-decrescendo sequence being separated by central apneas. A so-called variant Cheyne-Stokes pattern has hypopneas rather than apneas.

Periodic respiration may be observed in unconscious patients with lesions of the deep cerebral hemispheres, diencephalon, or upper pons, or with central or tonsillar brain herniation; it has also been reported in multiple system atrophy. Prolonged circulatory time (congestive heart failure) and hypoxemia (*e.g.*, at altitude) may also cause periodic respiration, but with a shorter cycle.

References

Pearce JMS. Cheyne-Stokes respiration. In: Pearce JMS. *Fragments of neurological history*. London: Imperial College Press, 2003: 355-358

Cross References

Coma

Perseveration

Perseveration refers to any continuation or recurrence of activity without appropriate stimulus (*cf*. intrusions). Perseverations may be repeated motor behaviors (*e.g.*, drawing, writing) or speech. These are viewed as a failure to inhibit a previous response pattern. Sensory perseveration is also described, *e.g.*, palinopsia in the visual system.

A number of varieties of perseveration have been described, associated with lesions in different areas of the brain:

- "*Stuck-in-set*":
  Inappropriate maintenance of a current category or framework; thought to reflect a deficit in executive function; associated with frontal lobe (especially frontal convexity) damage, which is associated with an inert, apathetic pattern of behavior, rather than the disinhibited pattern associated with orbitofrontal damage.
- "*Recurrent*":
  Unintentional repetition of a previous response to a subsequent stimulus; thought to represent an abnormal post-facilitation of a memory trace; associated with posterior left (dominant) hemisphere damage; commonly seen in aphasics, Alzheimer's disease; this overlaps with "intrusions."
- "*Continuous*":
  Inappropriate prolongation or repetition of a current behavior without interruption; thought to represent a deficit of motor output; associated with basal ganglia damage.

References

Hudson AJ. Perseveration. *Brain* 1968; **91**: 571-582

Sandson J, Albert ML. Varieties of perseveration. *Neuropsychologia* 1984; **22**: 715-732

Cross References

Aphasia; Dysexecutive syndrome; Frontal lobe syndromes; Intrusion; Palinopsia

Pes Cavus

Pes cavus is a high-arched foot due to equinus (plantar flexion) deformity of the first ray, with secondary changes in the other rays (*i.e.*, deformity is more evident on the medial side of the foot in most cases). This may be due to imbalance of muscular forces during development (*e.g.*, strong peroneus longus, weak peroneus brevis and tibialis anterior, although the precise pattern may differ with cause), which may be a consequence of neurological disease. Hammer toes may also be present. Pes cavus may be associated with disease of genetic origin, *e.g.*, hereditary motor and sensory neuropathy (HMSN, Charcot-Marie-Tooth syndrome), hereditary spastic paraparesis, Friedreich's ataxia, Marfan's syndrome; or be due to an early neurological insult, *e.g.*, cerebral palsy, paralytic poliomyelitis. Familial pes cavus without other neurological signs also occurs.

Surgical treatment of pes cavus may be necessary, especially if there are secondary deformities causing pain, skin breakdown, or gait problems.

Cross References

Claw foot; Hammer toes

"Petite Madeleines Phenomenon"

- see PROUST PHENOMENON

Phalen's Sign

Phalen's sign is present when tingling (paresthesia) is experienced in the distribution of the median nerve when the wrist is held in forced flexion (90º for 30-60 seconds; Phalen's maneuver). Patients may volunteer that they experience such symptoms when carrying heavy items, such as shopping bags, that put the hand in a similar posture. Hyperextension of the wrist ("reverse Phalen's maneuver") may also reproduce symptoms. These are signs of compression of the median nerve at the wrist (carpal tunnel syndrome). Like other provocative tests (*e.g.*, Tinel's sign), the sensitivity of Phalen's sign for this diagnosis is not high compared to electrophysiological testing.

The pathophysiology of Phalen's sign is probably the lower threshold of injured nerves to mechanical stimuli, as for Tinel's sign and Lhermitte's sign.

References

Heller L, Ring H, Costeff H, Solzi P. Evaluation of Tinel's and Phalen's signs in the diagnosis of the carpal tunnel syndrome. *European Neurology* 1986; **25**: 40-42

Cross References

Flick sign; Lhermitte's sign; Paresthesia; Tinel's sign

"Phantom Alloesthesia"

- see ALLOESTHESIA

Phantom Chromatopsia

This term has been coined to refer to the complaint of patients who are blind or nearly so that a color, usually golden or purple, enlarges to

invade the entire visual field. This is presumably cortical in origin, and has been described as an hallucination. "Phantom vision" may describe a similar phenomenon.

References

Zeki S. *A vision of the brain*. Oxford: Blackwell Science, 1993: 278, 279

Cross References

Erythropsia; "Monochromatopsia"

Phantom Limb

Phantom limbs, or ghost limbs, are the subjective report of the awareness of a nonexisting or deafferented body part in a mentally otherwise competent individual. The term was coined by Weir Mitchell in the nineteenth century, but parts other than limbs (either congenitally absent or following amputation) may be affected by phantom phenomena, such as lips, tongue, nose, eye, penis, breast and nipple, teeth, and viscera. Phantom phenomena are perceived as real by the patient, may be subject to a wide range of sensations (pressure, temperature, tickle, pain), and are perceived as an integral part of the self. Such "limbless perception" is thought to reflect the mental representation of body parts generated within the brain (body schema), such that perception is carried out without somatic peripheral input. Reorganization of cortical connections following amputation may explain phantom phenomena, such as representation of a hand on the chest or face, for which there is also evidence from functional brain imaging.

References

Halligan PW. Phantom limbs: the body in mind. *Cognitive Neuropsychiatry* 2002; **7**: 251-268

Melzack R. Phantom limbs. *Scientific American* 1992; **266**: 120-126

Ramachandran VS, Hirstein W. The perception of phantom limbs. *Brain* 1998; **121**: 1603-1630

Phantom Vision

This name has been given to visual hallucinations following eye enucleation, by analogy with somesthetic sensation experienced in a phantom limb after amputation. Similar phenomena may occur after acute visual loss, and may overlap with phantom chromatopsia. Unformed or simple hallucinations are more common than formed or complex hallucinations.

References

Cohn R. Phantom vision. *Archives of Neurology* 1971; **25**: 468-471

Lepore FE. Spontaneous visual phenomena with visual loss. *Neurology* 1990; **40**: 444-447

Cross References

Hallucination; Phantom chromatopsia; Phantom limb

Pharyngeal Reflex

- see GAG REFLEX

Phonagnosia

Phonagnosia is an inability to recognize familiar voices in the absence of hearing impairment, hence a form of auditory agnosia. The patient can recognize and understand words and sentences (*cf.* pure word deafness). Phonagnosia is the equivalent in the auditory domain of prosopagnosia in the visual domain. The neuroanatomical substrate is thought to be right parietal lobe pathology.

Cross References

Agnosia; Auditory agnosia; Prosopagnosia; Pure word deafness

Phonemic Disintegration

Phonemic disintegration refers to an impaired ability to organize phonemes, the smallest units in which spoken language may be sequentially described, resulting in substitutions, deletions and misorderings of phonemes. Phonemic disintegration is relatively common in aphasic disorders, including Broca's aphasia, conduction aphasia, and transcortical motor aphasia. Isolated phonemic disintegration is rare. The neural substrate may be primary motor cortex of the left inferior precentral gyrus and subjacent white matter, with sparing of Broca's area.

References

Larner AJ, Robinson G, Kartsounis LD *et al*. Clinical-anatomical correlation in a selective phonemic speech production impairment. *Journal of the Neurological Sciences* 2004; **219**: 23-29

Taubner RW, Raymer AM, Heilman KM. Frontal-opercular aphasia. *Brain & Language* 1999; **70**: 240-261

Cross References

Aphasia; Aphemia; Broca's aphasia

Phonetic Disintegration

- see APHEMIA

Phonophobia

Phonophobia is a dislike, or fear, of sounds, especially loud sounds, often experienced during a migraine headache.

Cross References

Hyperacusis

Phosphene

Phosphenes are percepts in one modality induced by an inappropriate stimulus, *e.g.*, when pressure is applied to the eyeball, the mechanical stimulus may induce the perception of light. The perception of flashes of light when the eyes are moved has been reported in optic neuritis, presumably reflecting the increased mechanosensitivity of the demyelinated optic nerve fibers; this is suggested to be the visual equivalent of Lhermitte's sign. Eye gouging to produce phosphenes by mechanical stimulation of the retina is reported in Leber's congenital amaurosis.

Noise-induced visual phosphenes have also been reported, and may be equivalent to auditory-visual synesthesia.

References

Davis FA, Bergen D, Schauf C, McDonald I, Deutsch W. Movement phosphenes in optic neuritis: a new clinical sign. *Neurology* 1976; **26**: 1100-1104

Lessell JB, Cohen MM. Phosphenes induced by sound. *Neurology* 1979; **29**: 1524-1526

Cross References

Auditory-visual synesthesia; Gaze-evoked phenomena; Lhermitte's Sign; Photism; Synesthesia

Photism

Photisms are transient positive visual phenomenon, such as geometrical shapes or brightly colored spectral phenomena, occurring in the context of epilepsy, migraine, or in blind visual fields (hence overlapping with photopsia). Auditory-visual synesthesia may also be described as sound-induced photism.

Cross References

Auditory-visual synesthesia; Photopsia

Photophobia

Photophobia is a dislike, or fear, of light, often experienced with meningitis and other causes of meningeal irritation, and during a migraine headache.

Cross References

Dazzle; Meningism

Photopsia

Photopsias are simple visual hallucinations consisting of flashes of light which often occur with a visual field defect. They suggest dysfunction in the inferomedial occipital lobe, such as migraine or an epileptogenic lesion.

Cross References

Aura; Hallucination; Photism

Piano-Playing Fingers

- see PSEUDOATHETOSIS

Pica

Pica, or pagophagia, is a morbid craving for unusual or unsuitable food in association with iron deficiency. It has also been reported in tuberous sclerosis. Sufferers risk infection from contaminated foods.

"Picture Sign"

The "picture sign" is present when a patient believes that individuals seen on the television screen are actually present in the home; indeed they may be reported to emerge from the television set into the room.

This may occur as part of the cognitive disturbance of Alzheimer's disease or dementia with Lewy bodies, or as part of a psychotic disorder. Like the "mirror sign," the "picture sign" may be classified as a misidentification phenomenon.

Cross References

"Mirror sign"; Misidentification syndromes

"Picture Within a Picture" Sign

Following a right parieto-occipital infarction, a patient complained of seeing people moving about in the left lower quadrant of the visual field while vision was normal in the remainder of the visual field, a phenomenon labeled the "picture within a picture" sign. This has been categorized as a visual release hallucination.

References

Benegas MN, Liu GT, Volpe NJ, Galetta SL. "Picture within a picture" visual hallucinations. *Neurology* 1996; **47**: 1347-1348

Pied en Griffe

- see CLAW FOOT

"Pie-in-the-Sky" Defect

This name has sometimes been given to the superior homonymous quadrantanopia ending sharply at the vertical midline due to a temporal lobe lesion interrupting Meyer's loop, that part of the optic radiation coursing through the temporal lobe.

Cross References

Quadrantanopia

"Pill Rolling"

- see PARKINSONISM; TREMOR

"Pinch Sign"

The "pinch sign" is an inability to make a small circle ("form the letter O") by approximating the distal phalanges of the thumb and index finger, due to weakness of flexor digitorum profundus in the index finger and flexor pollicis longus in the thumb as a consequence of median nerve lesions in the forearm, *e.g.*, anterior interosseous syndrome, pronator teres syndrome. This results in a pinching posture of thumb and index finger. The "straight thumb sign" may also be present (*q.v.*).

Cross References

Froment's sign; "Straight thumb sign"

Pinhole Test

Impairments in visual acuity due to refractory defects (changes in shape of the globe or defects in the transparent media of the eye) may be improved or corrected by looking through a pinhole which restricts vision to the central beam of light.

Plantar Response

The plantar response is most commonly elicited by stroking the sole of the foot with a blunt object. The first response of the hallux is the critical observation, which may be facilitated by having ones line of vision directly above the axis of the toe. The normal response after maturation of the corticospinal tracts (*i.e.*, after about three years of age) is for the big toe to flex. An extensor response of the big toe in an adult (Babinski's sign), with or without fanning (abduction) of the other toes (fan sign, *signe de l'éventail*), is a reliable sign of upper motor neurone pathology. Use of the term "negative Babinski's sign" or "negative Babinski response" to mean "flexor plantar response" is incorrect and should not be used. This normal plantar response is a superficial cutaneous reflex, analogous to abdominal and cremasteric reflexes, whereas the pathological response is often accompanied by activity in other flexor muscles. In some individuals the toes do not move at all, in which case the response is labeled as "mute" or absent. Assessment of the response may be confounded by withdrawal of the foot in ticklish individuals. Differentiation from the striatal toe seen in parkinsonian syndromes is also important.

The plantar response may be elicited in a variety of other ways which are not in routine clinical use. Of these, perhaps the most frequently used are Chaddock's sign (application of a stimulus in a circular direction around the external malleolus, or the lateral aspect of the foot from heel to little toe) and Oppenheim's sign (application of heavy pressure to the anterior surface of the tibia from patella to ankle). These may be helpful in ticklish patients who object to having their feet stroked. If the plantar response thus elicited is upgoing, this suggests a spread of the "receptive field" of the reflex. Babinski's sign is the first to occur in the presence of upper motor neurone pathology.

It is often difficult to form a definite judgment on the plantar response and reproducibility is also questionable. A study of 24 experienced clinicians invited to examine plantar responses "blind" found that the interobserver percentage agreement beyond chance was on average only 16.7% (95% confidence interval [CI] 0.4-33%); intraobserver percentage agreement was a little better (average 59.6%; CI 39.6-79.6%). There remains a persistent belief, particularly amongst trainees, that an experienced neurologist can make the plantar response go which ever way s/he chooses.

References

Maher J, Reilly M, Daly L, Hutchinson M. Plantar power: reproducibility of the plantar response. *BMJ* 1992; **304**: 482

Van Gijn J. *The Babinski sign: a centenary*. Utrecht: Universiteit Utrecht, 1996

Cross References

Abdominal reflexes; Babinski's sign (1); Chaddock's sign; Gordon's sign; Oppenheim's sign; Reflexes; Striatal toe, Upper motor neurone (UMN) syndrome

Plegia

Plegia means stillness, implying a complete weakness (or paralysis in common parlance), as in monoplegia, diplegia, ophthalmoplegia, paraplegia, quadriplegia, cardioplegia. Hence plegia is a more severe weakness than paresis.

Cross References

Paresis; Weakness

Plexopathy

Lesions confined to the brachial, lumbar, or sacral plexi may produce a constellation of motor and sensory signs (weakness, reflex diminution or loss, sensory loss) which cannot be ascribed to single or multiple roots (radiculopathy) or peripheral nerves (neuropathy). Lesions may involve the whole plexus (panplexopathy):

Brachial: C5-T1
Lumbar: L2-L4
Sacral: L5-S3

or be partial, *e.g.*, upper trunk of brachial plexus (C5-C6), producing "waiter's tip" posture (as for C5/C6 root avulsion); lower trunk of brachial plexus (C8-T1; as for C8/T1 root avulsion).

Electrophysiological studies may be helpful in distinguishing plexopathy from radiculopathy: sensory nerve action potentials (SNAPs) are reduced or absent in plexopathies because the lesion is located distal to the dorsal root ganglion (DRG), whereas SNAPs are normal in radiculopathies because the lesion is proximal to the DRG. EMG shows sparing of paraspinal muscles in a plexopathy because the lesion is, by definition, distal to the origin of the dorsal primary rami (*cf*. radiculopathy). Coexistence of radiculopathy and plexopathy may invalidate these simple rules.

- Recognized causes of brachial plexopathy include:
  Trauma: Upper plexus: Dejerine-Klumpke paralysis ("waiter's tip" posture);
  Lower plexus: Erb-Duchenne paralysis (claw hand).
  Inflammation/Idiopathic: brachial neuritis, neuralgic amyotrophy.
  Malignant infiltration, *e.g.*, carcinoma of lung (Pancoast), breast, +/– Horner's syndrome; pain a significant symptom.
  Post-radiation (*e.g.*, after radiotherapy for malignant breast cancer with axillary spread; myokymic discharges may be seen on EMG).
  Tomaculous neuropathy.
  Hereditary neuropathy with liability to pressure palsies (HNLPP).
  Neurogenic thoracic outlet syndrome (rare): cervical rib or C7 transverse process or fibrous band compressing the lower trunk. May be surgically remediable.
- Recognized causes of lumbosacral plexopathy include:

Compression; *e.g.*, iliopsoas hematoma (anticoagulation, hemophilia), abscess (tuberculosis); abdominal aortic aneurysm; pregnancy (fetal head in the second stage of labor).
Neoplasia (direct spread > metastasis).
Trauma (rare; *cf*. brachial plexopathy).
Post-radiation.
Vasculitis (mononeuritis multiplex much commoner).
Idiopathic.

Imaging with MRI is superior to CT for defining structural causes of plexopathy.

References

Chad DF. Nerve root and plexus disorders. In: Bogousslavsky J, Fisher M (Eds.). *Textbook of neurology*. Boston: Butterworth-Heinemann, 1998: 491-506

Taylor BV, Kimmel DW, Krecke KN, Cascino TL. Magnetic resonance imaging in cancer-related lumbosacral plexopathy. *Mayo Clinic Proceedings* 1997; **72**: 823-829

Cross References

Amyotrophy; Claw hand; Horner's syndrome; Nerve thickening; Neuropathy; Radiculopathy; "Waiter's tip" posture

Polyganglionopathy

- see NEUROPATHY

Polymyoclonus

- see MYOCLONUS

Polyneuropathy

- see NEUROPATHY

Polyopia

Polyopia, or polyopsia, is a visual illusory phenomenon in which a single target is seen as multiple images, most usually double but sometimes higher multiples (*e.g.*, entomopia), persisting when looking away from the object. This may be likened to "echoes" of the image, and eye movement may produce a trailing effect. Polyopia may be related to palinopsia.

Polyopia is associated with occipital and occipito-parietal lesions, bilateral or confined to the nondominant hemisphere, and with drug abuse. It has also been described in disease of the retina and optic nerve, and occasionally in normal individuals.

The pathophysiology is unknown; suggestions include a defect of visual fixation or of visual integration; the latter may reflect pure occipital cortical dysfunction.

References

Pomeranz HD, Lessell S. Palinopsia and polyopia in the absence of drugs or cerebral disease. *Neurology* 2000; **54**: 855-859

Cross References

Entomopia; Illusion; Palinopsia

"Popeye Arms"
In facioscapulohumeral (FSH) muscular dystrophy, the deltoid muscle is normally well preserved, while biceps and triceps are weak and wasted, giving rise to an appearance of the upper limbs sometimes labeled as "Popeye arms" or "chicken wings."
Cross References
Winging of the scapula

Poriomania
A name sometimes given to prolonged wandering as an epileptic automatism, or a fugue state of nonconvulsive status epilepticus.
References
Mayeux R, Alexander MP, Benson DF *et al*. Poriomania. *Neurology* 1979; **29**: 1616-1619
Cross References
Automatism; Seizures

Porropsia
A form of metamorphopsia characterized by the misperception of objects as farther away from the observer than they really are (*cf*. pelopsia)
Cross References
Metamorphopsia; Pelopsia

Positional Maneuvers
- see HALLPIKE MANEUVER, HALLPIKE TEST; HEAD IMPULSE TEST; VESTIBULO-OCULAR RELFEXES

Post-Tetanic Potentiation
- see FACILITATION

Postural Hypotension
- see ORTHOSTATIC HYPOTENSION

Postural Reflexes
Postures, such as standing, are largely reflex in origin, dependent upon involuntary muscle contraction in antigravity muscles. Interference with such reflex activity impairs normal standing. Postural and righting reflexes depend on the integration of labyrinthine, proprioceptive, exteroceptive, and visual stimuli, mostly in the brainstem but also involving the cerebral cortex. However, abnormalities in these reflexes are of relatively little diagnostic value except in infants.

One exception is extrapyramidal disease (parkinsonism, Huntington's disease, but not idiopathic dystonia) in which impairment or loss of postural reflexes may be observed. In the "pull test" the examiner stands behind the patient, who is standing comfortably, and pulls briskly on the shoulders; this may provoke repetitive steps backward (retropulsion, festination) or even *en bloc* falling, due to the failure of reflex muscle contraction necessary to maintain equilibrium. Pushing the patient may likewise provoke propulsion or festination, but

this maneuver is less safe since the examiner will not be placed to catch the patient should they begin to topple over.

Cross References

Dystonia; Festinant gait, Festination; Parkinsonism; Proprioception; "Rocket sign"

Pout Reflex

The pout reflex consists of a pouting movement of the lips elicited by lightly tapping orbicularis oris with a finger or tendon hammer, or by tapping a spatula placed over the lips. This myotactic stretch reflex is indicative of a bilateral upper motor neurone lesion, which may be due to cerebrovascular small vessel disease, motor neurone disease or multiple sclerosis. It differs from the snout reflex, which refers to the reflex elicited by constant pressure on the philtrum.

References

Rossor M. Snouting, pouting and rooting. *Practical Neurology* 2001; **1**: 119-121

Cross References

Frontal release signs; Primitive reflexes

"Prayer Sign"

An inability to fully oppose the palmar surfaces of the digits with the hands held in the praying position, recognized causes of which include ulnar neuropathy (*main en griffe*), Dupuytren's contracture, diabetic cheiroarthropathy, and camptodactyly.

Cross References

Camptodactyly; "Table top" sign

Prehensile Thumb Sign

- see FROMENT'S SIGN

Presbycusis

Presbycusis is a progressive sensorineural hearing loss, especially for high frequencies, developing with increasing age, which may reduce speech discrimination. It is thought to be due to age-related attrition of hair cells in the organ of Corti and/or spiral ganglion neurones.

Cross References

Age-related signs

Presbyopia

Presbyopia is progressive far-sightedness which is increasingly common with increasing age, thought to be due to an age-related impairment of accommodation.

Cross References

Age-related signs

Prevost's Sign

Also known as Vulpian's sign, this refers to the acute and transient gaze palsy in a frontal lesion (*e.g.*, infarct) which is toward the side of

the lesion and away from the concurrent hemiparesis. The eyes can be brought to the other side with the oculocephalic maneuver or caloric testing. In contrast, thalamic and basal ganglia hemorrhages produce forced deviation of the eyes to the side contralateral to the lesion (wrong-way eyes).

Priapism

Priapism is an unintended, sustained, and usually painful erection of the penis unrelated to sexual activity. It may occur with intramedullary spinal cord lesions (*e.g.*, multiple sclerosis) which damage the lumbosacral erection centres, and has also been reported with lumbar canal spinal stenosis. There are also nonneurological causes, such as hematological conditions (sickle cell anemia, polycythemica rubra vera) which may cause intrapenile thromboses.

Primitive Reflexes

Reflexes which are normally found in infancy but which disappear with brain maturation during childhood may be labeled as "primitive reflexes" if they reemerge in adulthood as a consequence of pathological states. Many of these reflexes are seen with frontal lobe pathology (*e.g.*, grasp, pout/snout, palmomental, rooting, corneomandibular) and hence may also be known as "frontal release signs." However, the term "primitive reflex" could equally apply to Babinski's sign which is not necessarily frontal in origin.

References

Schott JM, Rossor MN. The grasp and other primitive reflexes. *Journal of Neurology, Neurosurgery and Psychiatry* 2003; **74**: 558-560

Cross References

Babinski's sign (1); Corneomandibular reflex; Frontal release signs; Grasp reflex; Palmomental reflex; Pout reflex; Rooting reflex

Procerus Sign

A focal dystonia of the procerus muscle, denoted the procerus sign, has been suggested to contribute to the "astonished," "worried" or "reptile-like" facial expression typical of progressive supranuclear palsy, which may also be characterized by reduced blinking, lid retraction, and gaze palsy. All contrast with the hypomimia of Parkinson's disease.

References

Romano S, Colosimo C. Procerus sign in progressive supranuclear palsy. *Neurology* 2001; **57**: 1928

Cross References

Blinking; Dystonia; Hypomimia; Parkinsonism

Proprioception

Proprioception sensation, or joint position sense, is knowledge about ones position in space, originating from sensory receptors in skin, muscle, and viscera. Proprioceptive information is carried within the dorsal

columns of the spinal cord (more reliably so than vibration sensation, though not necessarily exclusively). Lesions affecting this part of the cord, particularly in the cervical region (*e.g.*, subacute combined degeneration of the cord due to vitamin B_{12} deficiency, tabes dorsalis), lead to impairments of proprioception with sparing of spinothalamic sensations (pin-prick, temperature) producing a dissociated sensory loss. Impairment of proprioception leads to sensory ataxia which may manifest clinically with pseudoathetosis or pseudochoreoathetosis (also seen in useless hand of Oppenheim) and with a positive Romberg's sign.

References

Gilman S. Joint position sense and vibration sense. *Journal of Neurology, Neurosurgery and Psychiatry* 2002; **73**: 473-477

Cross References

Ataxia; Dissociated sensory loss; Pseudoathetosis; Pseudochoreoathetosis; Rombergism, Romberg's sign; Useless hand of oppenheim; Vibration

Proptosis

Proptosis is forward displacement of the eyeball, an exaggerated degree of exophthalmos. There may be lower lid retraction. Proptosis may be assessed clinically by standing directly behind the patient and gradually tipping the head back, observing when the globe of the eyeball first comes into view; this is most useful for asymmetric proptosis. An exophthalmometer may be used to measure proptosis. Once established, it is crucial to determine whether the proptosis is axial or nonaxial. Axial proptosis reflects increased pressure within or transmitted through the cone of extraocular muscles (*e.g.*, thyroid ophthalmopathy, cavernous sinus thrombosis), whereas nonaxial proptosis suggests pressure from an orbital mass outside the cone of muscles (*e.g.*, orbital lymphoma, pseudotumor, mucocele). Pulsatile axial proptosis may occur in carotico-cavernous fistula, in which case there may be a bruit audible by auscultation over the eye. Venous angioma of the orbit may cause an intermittent proptosis associated with straining, bending, coughing or blowing the nose.

Dedicated orbital CT or MRI, the latter with fat-suppression sequences and intravenous gadolinium contrast, may be required to detect intraorbital masses.

Middle fossa tumors may cause pressure on the veins of the cavernous sinus with secondary intraorbital venous congestion causing a "false-localizing " proptosis.

Cross References

Exophthalmos; "False-localizing signs"; Lid retraction

Propulsion

- see FESTINANT GAIT, FESTINATION; POSTURAL REFLEXES

Prosopagnosia

Prosopagnosia is a form of visual agnosia characterized by an inability to recognize previously known human faces or equivalent stimuli

(hence, a retrograde defect) and to learn new ones (anterograde defect). As with more pervasive visual agnosia (*q.v.*), this may be:

- *Apperceptive*: due to faulty perceptual analysis of faces; or
- *Associative*: a semantic defect in recognition.

Familiar individuals may be recognized by their voices or clothing or hair; hence, the defect may be one of visually triggered episodic memory. It is important to note that the defect is not limited solely to faces; it may encompass animals ("zooagnosia"), or cars.

Prosopagnosia is often found in association with a visual field defect, most often a left superior quadrantanopia or even hemianopia, although for the diagnosis of prosopagnosia to be made this should not be sufficient to produce a perceptual deficit. Alexia and achromatopsia may also be present, depending on the exact extent of the underlying lesion.

Anatomically, prosopagnosia occurs most often in association with bilateral occipito-temporal lesions involving the inferior and mesial visual association cortices in the lingual and fusiform gyri, sometimes with subjacent white matter. Unilateral nondominant (right) hemisphere lesions have occasionally been associated with prosopagnosia, and a syndrome of progressive prosopagnosia associated with selective focal atrophy of the right temporal lobe has been reported. Involvement of the periventricular region on the left side may explain accompanying alexia, and disconnection of the inferior visual association cortex (area V4) may explain achromatopsia.

Pathological causes of prosopagnosia include:

- Cerebrovascular disease: this is by far the most common cause
- Tumor, for example, glioma, extending from one hemisphere to the other via the splenium of the corpus callosum
- Epilepsy (paroxysmal prosopagnosia), due to bilateral foci or spread from one occipital focus to the contralateral hemisphere
- Right temporal lobe atrophy
- Herpes simplex encephalitis, usually as part of an extensive amnesic syndrome (although memory impairment may put this outwith the operational criteria for an agnosia)
- Rare cases of developmental (or "congenital") prosopagnosia have been described.

References

Evans JJ, Heggs AJ, Antoun N, Hodges JR. Progressive prosopagnosia associated with selective right temporal lobe atrophy. A new syndrome? *Brain* 1995; **118**: 1-13

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

Nunn JA, Postma P, Pearson R. Developmental prosopagnosia: should it be taken at face value? *Neurocase* 2001; **7**: 15-27

Cross References

Achromatopsia; Agnosia; Alexia; Hemianopia; Phonagnosia; Quadrantanopia; Visual agnosia; Zooagnosia

Prosopoplegia
- see BELL'S PALSY; FACIAL PARESIS

Proust Phenomenon
The Proust phenomenon, named after the author Marcel Proust (1871-1922), is the observation that particular odors may trigger reminders of autobiographical memories. There is some experimental evidence that olfactory stimuli can cue autobiographical memories more effectively than cues from other sensory modalities. The "petite Madeleines phenomenon" has been used to describe sudden triggering of memories in individuals with amnesia due to thalamic infarction.
References
Chu S, Downes JJ. Odor-evoked autobiographical memories: psychological investigations of Proustian phenomena. *Chemical Senses* 2000; **25**: 111-116
Lucchelli F, Muggia S, Spinnler H. The "Petites Madeleines" phenomenon in two amnesic patients: sudden recovery of forgotten memories. *Brain* 1995; **118**: 167-183
Cross References
Amnesia

Pseudoachromatopsia
Pseudoachromatopsia is failure on tests of color vision (*e.g.*, pseudoisochromatic plates) not due to central or peripheral achromatopsia, for example due to visual neglect.
Cross References
Achromatopsia; Neglect

Pseudo-Abducens Palsy
- see ABDUCENS (VI) NERVE PALSY

Pseudoagnosia
- see AGNOSIA

Pseudo-Argyll Robertson Pupil
A pseudo-Argyll Robertson pupil shows light-near dissociation of pupillary reactions, but, unlike the "true" Argyll Robertson pupil, there is no miosis or pupil irregularity. Indeed the pupil may be dilated (mydriasis) and resemble a Holmes-Adie pupil. The latter may be differentiated on the basis of its response to dilute (0.2%) pilocarpine: Holmes-Adie pupil results from a peripheral lesion and shows denervation supersensitivity constricting with dilute pilocarpine, whereas the pseudo-Argyll Robertson pupil results from a central lesion and does not respond.
Pseudo-Argyll Robertson pupil has been reported in:

Diabetes mellitus
Multiple sclerosis
Wernicke's encephalopathy

Sarcoidosis
Tumor
Hemorrhage
Aberrant oculomotor (III) nerve regeneration
Spinocerebellar ataxia type 1 (SCA1)

Cross References
Argyll Robertson pupil; Holmes-adie pupil, Holmes-adie syndrome; Miosis; Mydriasis

Pseudoathetosis

Pseudoathetosis is the name given to athetoid-like movements, most usually of the outstretched fingers ("piano-playing fingers") and hands, resulting from sensory ataxia (impaired proprioception); it is worse with the eyes closed. There may also be chorea-like movements (see Pseudochoreoathetosis). Causes include any interruption to the anatomical pathway mediating proprioception, most often lesions in the dorsal cervical cord (*e.g.*, multiple sclerosis, subacute combined degeneration of the cord due to vitamin B_{12} deficiency or nitrous oxide overuse), but also lesions of the large (myelinated) peripheral nerve fibers, and of the parietal lobe.

Cross References
Athetosis; Chorea, Choreoathetosis; Proprioception; Pseudochoreoathetosis

Pseudobitemporal Hemianopia

- see HEMIANOPIA

Pseudobulbar Affect

- see EMOTIONALISM, EMOTIONAL LABILITY; PATHOLOGICAL CRYING, PATHOLOGICAL LAUGHTER; PSEUDOBULBAR PALSY

Pseudobulbar Palsy

Pseudobulbar palsy, or spastic bulbar palsy, describes bilateral upper motor neurone lesions affecting fibers passing to the cranial nerve nuclei (*cf.* bulbar palsy). This leads to a variety of clinical problems, including:

Difficulty with speech: spastic dysarthria, dysphonia
Difficulty with swallowing: dysphagia
Brisk jaw jerk and pout reflex; there may be trismus
Slow, spastic, tongue movements
Gag reflex may be depressed or exaggerated.

There may be associated emotional lability, or pathological laughter and crying ("pseudobulbar affect"), and a gait disorder with *marche à petit pas*. There are otherwise few signs in the limbs, aside from brisk reflexes and upgoing plantar responses (Babinski's sign).

Recognized causes of pseudobulbar palsy include:

Motor neurone disease (in which there may be coincident bulbar palsy)
Multiple sclerosis
Bilateral internal capsule lacunar infarctions, widespread small vessel disease (Binswanger's disease)
Congenital childhood suprabulbar palsy (Worster-Drought syndrome; perisylvian syndrome).

Cross References
Babinski's sign (1); Bulbar palsy; Dysarthria; Dysphagia; Dysphonia; Emotionalism, Emotional lability; Gag reflex; Jaw jerk; Marche à petit pas; Pathological crying, Pathological laughter; Trismus; Upper motor neurone (UMN) syndrome

Pseudochoreoathetosis

Pseudochoreoathetosis is the name given to choreoathetoid type involuntary movements, including dystonic movements, which result from a loss or impairment of proprioception. These may be observed with lesions anywhere along the proprioceptive pathways, including parietal cortex, thalamus (may be associated with ataxic hemiparesis and hemihypoesthesia), spinal cord, dorsal root ganglia (neuronopathy), and mononeuropathy.

References
Sharp FR, Rando TA, Greenberg SA, Brown L, Sagar SM. Pseudochoreoathetosis. Movements associated with loss of proprioception. *Archives of Neurology* 1994; **51**: 1103-1109

Cross References
Ataxic hemiparesis; Chorea, Choreoathetosis; Dystonia; Proprioception; Pseudoathetosis; Useless hand of oppenheim

Pseudodementia

Pseudodementia is a label given to cognitive impairments resulting from affective disorders, most commonly anxiety and depression; the terms "dementia syndrome of depression" and "depression-related cognitive dysfunction" have also been used. The pattern of cognitive deficits in individuals with depression most closely resembles that seen in so-called subcortical dementia, with bradyphrenia, attentional and executive deficits. In addition there may be evident lack of effort and application, "don't know" answers, approximate answers (Ganser phenomenon, *vorbereiden*), and evidence of mood disturbance (tearfulness). Memory loss for recent and distant events may be equally severe (*cf*. temporal gradient of memory loss in dementia, *e.g*., due to Alzheimer's disease). A 22-item checklist to help differentiate pseudodementia from Alzheimer's disease has been described, based on clinical history, behavior and mental status.

The recognition of pseudodementia is important since the deficits are often reversible with appropriate treatment with antidepressants. However, it should be borne in mind that depression is sometimes the presenting symptom of an underlying neurodegenerative dementing

disorder, such as Alzheimer's disease. Longitudinal assessment may be required to differentiate between these diagnostic possibilities.

References

Andrews C. Pseudodementia responding to antidepressant therapy. *Progress in Neurology and Psychiatry* 2002; **6(1)**: 26

Kiloh L. Pseudodementia. *Acta Psychiatrica Scandinavica* 1961; **37**: 336-351

Roose SP, Devanand DP. *The interface between dementia and depression*. London: Martin Dunitz, 1999

Wells CE. Pseudodementia. *American Journal of Psychiatry* 1979; **136**: 895-900

Cross References

Attention; Bradyphrenia; Dementia; Ganser phenomenon

Pseudodiplopia

- see PALINOPSIA

Pseudohypertrophy

- see CALF HYPERTROPHY; MUSCLE HYPERTROPHY

Pseudo-Internuclear Ophthalmoplegia

Pseudo-internuclear ophthalmoplegia is a disorder of eye movements with impaired adduction in one eye and horizontal nystagmus in the abducting eye (*i.e.*, signs as in an internuclear ophthalmoplegia) but without an intrinsic brainstem lesion. This sign may be seen in:

- Myasthenia gravis (a diagnosis which is always worthy of consideration in a patient with an "isolated INO") due to extraocular muscle weakness
- Brainstem compression due to subdural hematoma with transtentorial herniation
- Cerebellar mass lesion
- Guillain-Barré syndrome, Miller Fisher syndrome
- Thyroid ophthalmopathy
- Orbital pseudotumor.

Cross References

Internuclear ophthalmoplegia (INO); One-and-a-half syndrome

Pseudomyotonia

The term pseudomyotonia has been used to describe the clinical appearance of myotonia (slow muscular relaxation after contraction) in the absence of myotonic discharges on electromyography. Pseudomyotonia is most commonly observed as the slow-relaxing or "hung-up" tendon reflexes (Woltman's sign) of hypothyroidism, although other causes are described.

Pseudomyotonia has also been used to describe difficulty opening the hand in cervical osteoarthritis, although muscle relaxation is normal; finger flexion on attempted extension has been explained as aberrant regeneration of the C7 root.

The term pseudomyotonia has also been used to describe neuromyotonia and myokymia (as, for example, in Isaacs syndrome), to distinguish it from myotonia.

References

Satoyoshi E, Doi Y, Kinoshita M. Pseudomyotonia in cervical root lesions with myelopathy. A sign of the misdirection of regenerating nerve. *Archives of Neurology* 1972; **27**: 307-313

Cross References

Myotonia; Neuromyotonia; Woltman's sign

Pseudo-One-and-a-Half Syndrome

Pseudo-one-and-a-half syndrome is the eye movement disorder of one-and-a-half syndrome (*q.v.*) without a brainstem lesion. Myasthenia gravis and Guillain-Barré syndrome are recognized causes.

Cross References

One-and-a-half syndrome

Pseudopapilledema

Pseudopapilledema is the name given to elevation of the optic disc that is not due to edema (*i.e.*, intracranial pressure is not raised). There may or may not be visible drusen (hyaline bodies). In distinction to edematous disc swelling, the nerve fiber layer is not hazy and the underlying vessels are not obscured; however, spontaneous retinal venous pulsation is usually absent, and hemorrhages may be seen, so these are not reliable distinguishing features. Visual acuity is usually normal, but visual field defects (most commonly in the inferior nasal field) may be found.

Cross References

Disc swelling; Papilledema; Retinal venous pulsation

Pseudoparesis

- see PARESIS; WEAKNESS

Pseudoptosis

Ptosis, drooping of the eyelid, may need to be differentiated from pseudoptosis or functional ptosis. This may result simply from a redundant tarsal skin fold, especially in older patients, or be a functional condition. Frontalis underactivity may be a clinical indicator of the latter diagnosis (*cf.* compensatory overactivity of frontalis with other causes of ptosis).

The term has also been used in hypotropia; when the nonhypotropic eye fixates, the upper lid follows the hypotropic eye and appears ptotic, disappearing when fixation is with the hypotropic eye.

References

Hop JW, Frijns CJ, van Gijn J. Psychogenic pseudoptosis. *Journal of Neurology* 1997; **244**: 623-624

Stone J. Pseudo-ptosis. *Practical Neurology* 2002; **2**: 364-365

Cross References

Ptosis

Pseudoradicular Syndrome
Thalamic lesions may sometimes cause contralateral sensory symptoms in an apparent radicular (*e.g.*, C8) distribution. If associated with perioral sensory symptoms this may be known as the cheiro-oral syndrome.
References
Kim JS. Restricted acral sensory syndrome following minor stroke: further observations with special reference to differential severity of symptoms among individual digits. *Stroke* 1994; **25**: 2497-2502

Pseudo-Von Graefe's Sign
Pseudo-von Graefe's sign is seen on attempted downgaze or adduction of the eye, when there is involuntary retraction or elevation of the upper eyelid (*cf.* von Graefe's sign), medial rotation of the eye, and pupillary constriction. This constellation of findings is said to be a lid-gaze synkinesis following aberrant regeneration after an oculomotor (III) nerve palsy, usually of traumatic or chronic compressive rather than ischemic origin.
Cross References
Lid retraction; Synkinesia, Synkinesis; von Graefe's sign

Psychic Blindness
- see VISUAL AGNOSIA

Psychic Paralysis of Gaze
- see BALINT'S SYNDROME; OCULAR APRAXIA

Psychomotor Retardation
Psychomotor retardation is a slowness of thought (bradyphrenia) and movement (bradykinesia) seen in psychiatric disorders, particularly depression. It may be confused with the akinesia of parkinsonism, and abulic or catatonic states. Psychomotor retardation may also be a feature of the "subcortical" type of dementia, or of impairments of arousal (obtundation).
Cross References
Abulia; Akinesia; Catatonia; Dementia; Obtundation; Parkinsonism

Psychomotor Signs
- see FRONTAL RELEASE SIGNS

Ptarmus
- see SNEEZING

Ptosis
Ptosis, or blepharoptosis, is the name given to drooping of the eyelid. This may be due to mechanical causes, such as aponeurosis dehiscence, or neurological disease, in which case it may be congenital or acquired, partial or complete, unilateral or bilateral, fixed or variable, isolated or accompanied by other signs, *e.g.*, miosis in a Horner's

syndrome; diplopia in myasthenia gravis; mydriasis and downward and outward deviation of the eye in an oculomotor (III) nerve palsy.

Ptosis may result from pathology in a variety of locations: brainstem disease involving the oculomotor (III) nerve; anywhere along the oculosympathetic autonomic pathway causing a Horner's syndrome; or cortical disease (*e.g.*, infarction) reflecting hemispheric control of the eyelid (probably bilaterally represented).

When considering the cause of ptosis, the differential diagnosis is broad. Recognized causes include:

- Congenital:
  - Cranial nerve dysinnervation disorder
  - Congenital Horner's syndrome
  - Oculomotor-trigeminal (or trigeminal-levator) synkinesis: Marcus Gunn jaw-winking phenomenon, or inverse Marcus Gunn phenomenon (ptosis on jaw opening).
- Neurogenic:
  - Supranuclear lesion:
    - Hemiparesis: due to cortical infarct; ptosis usually ipsilateral, incomplete
    - Duane syndrome: ptosis on eye adduction, due to supranuclear levator inhibition; usually with family history
  - Oculomotor (III) nerve:
    - Hypertension, diabetes: ptosis often complete; in a superior divisional third nerve palsy partial ptosis is associated with superior rectus weakness only
    - Compressive lesion (*e.g.*, posterior communicating artery aneurysm): ptosis usually incomplete; ptosis may be present with subarachnoid hemorrhage
    - Guillain-Barré syndrome
    - Facial paresis
- Neuromuscular junction:
  - Myasthenia gravis: ptosis variable, bilateral or unilateral
  - Excessive botulinum toxin, *e.g.*, given for treatment of blepharospasm
- Myogenic: ptosis usually bilateral
  - Mitochondrial disease (CPEO)
  - Myotonic dystrophy
  - Oculopharyngeal muscular dystrophy (OPMD)
- Local, ophthalmological causes:
  - Age-related aponeurosis dehiscence, trauma, thyroid eye disease, lid inflammation (chalazion), lymphoma.

Pseudoptosis (*q.v.*) enters the differential diagnosis.

Enhanced ptosis, worsening of ptosis on one side when the other eyelid is held elevated in a fixed position, may be demonstrated in myasthenia gravis and Lambert Eaton myasthenic syndrome.

References

Caplan LR. Ptosis. *Journal of Neurology, Neurosurgery and Psychiatry* 1974; **37**: 1-7

Cross References
Blepharospasm; Diplopia; Divisional palsy; Ewart phenomenon; Horner's syndrome; Jaw winking; Miosis; Mydriasis; Pseudoptosis; Pupil sparing; Synkinesia, Synkinesis

Ptyalism
- see SIALORRHEA

Pulfrich Phenomenon
The Pulfrich phenomenon is the observation that a pendulum swung from side to side appears to traverse a curved trajectory. This is a stereo-illusion resulting from latency disparities in the visual pathways, most commonly seen as a consequence of conduction slowing in a demyelinated optic nerve following unilateral optic neuritis.

References
Diaper CJ. Pulfrich revisited. *Survey of Ophthalmology* 1997; **41**: 493-499
Rushton D. Use of the Pulfrich pendulum for detecting abnormal delay in the visual pathways in multiple sclerosis. *Brain* 1975; **98**: 283-296

Cross References
Phosphene; Relative afferent pupillary defect (RAPD)

"Pull Test"
- see POSTURAL REFLEXES

Pupillary Reflexes
Two pupillary reflexes are routinely examined in clinical practice:

- *Light reflex*:
  The eye is illuminated directly and the reaction (constriction) observed; the *consensual light reflex* is observed by illuminating the contralateral eye. In an eye with poor acuity, a *relative afferent pupillary defect* may be observed using the "swinging flashlight test." The afferent pathway subserving the light reflex is optic nerve to thalamus, brainstem, and Edinger-Westphal nucleus, with the efferent limb (pupillomotor parasympathetic fibers) in the oculomotor (III) nerve. The contralateral (consensual) response results from fibers crossing the midline in the optic chiasm and in the posterior commissure at the level of the rostral brainstem.
  Paradoxical constriction of the pupil in darkness (Flynn phenomenon) has been described.
- *Accommodation reflex*:
  This is most conveniently examined by asking the patient to look into the distance, then focus on a near object (sufficiently close to necessitate convergence of the visual axes) when pupil constriction should occur (accommodation-convergence synkinesis). The afferent pathways subserving this response are less certain than for the light reflex, and may involve the occipital cortex, although the final (efferent)

pathway via Edinger-Westphal nucleus and oculomotor nerve is common to both accommodation and light reflexes.

In comatose patients, fixed dilated pupils may be observed with central diencephalic herniation, whereas midbrain lesions produce fixed midposition pupils.

A dissociation between the light and accommodation reactions (light-near pupillary dissociation, *q.v.*) may be observed.

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; Ciliospinal response; Cortical blindness; Flynn phenomenon; Light-near pupillary dissociation; Miosis; Mydriasis; Relative afferent pupillary defect (RAPD); Swinging flashlight sign

Pupil Sparing

Oculomotor (III) nerve lesions may be pupil sparing (normal response to light) or pupil-involving (mydriasis, loss of light reflex). The latter situation usually implies a "surgical" cause of oculomotor palsy (*e.g.*, posterior communicating artery aneurysm), especially if extraocular muscle function is relatively preserved. Pupil sparing suggests a "medical" cause (*e.g.*, diabetes mellitus, hypertension) especially if the palsy is otherwise complete (complete ptosis, eye deviated downwards and outwards). This disparity arises because pupillomotor fibers run on the outside of the oculomotor nerve, and are relatively spared by ischemia but are vulnerable to external compression. However, the distinction is not absolute; imaging for an aneurysm (by means of spiral CT, MRA, or catheter angiography) may be necessary if the clinical scenario leaves room for doubt.

Cross References

Oculomotor (III) nerve palsy; Ophthalmoparesis, Ophthalmoplegia; Ptosis; Pupillary reflexes

Pure Word Blindness

- see ALEXIA

Pure Word Deafness

Pure word deafness is a rare condition characterized by an inability to comprehend and discriminate spoken language, despite adequate hearing as measured by audiometry, and with preserved spontaneous speech, reading, reading comprehension, and writing (*i.e.*, no aphasia, alexia, agraphia). Lip reading may assist in the understanding of others who sometimes seem to the patient as though they are speaking in a foreign language. Patients can copy and write spontaneously, follow written commands, but cannot write to dictation. Word repetition tasks are impaired. There may be associated amusia, depending on the precise location of cerebral damage.

Pure word deafness has been variously conceptualized as a form of auditory agnosia or a subcortical sensory aphasia.

Pure word deafness is most commonly associated with bilateral lesions of the temporal cortex or subcortical lesions whose anatomical effect is to damage the primary auditory cortex or isolate it (*e.g.*, from Wernicke's area) through lesions of the auditory radiation; unilateral lesions producing this syndrome have been reported. Very rarely pure word deafness has been associated with bilateral brainstem lesions at the level of the inferior colliculi.

References

Meyer B, Kral T, Zentner J. Pure word deafness after resection of a tectal plate glioma with preservation of wave V of brain stem auditory evoked potentials. *Journal of Neurology, Neurosurgery and Psychiatry* 1996; **61**: 423-424

Roberts M, Sandercock P, Ghadiali E. Pure word deafness and unilateral right temporo-parietal lesions: a case report. *Journal of Neurology, Neurosurgery and Psychiatry* 1987; **50**: 1708-1709

Tanaka Y, Yamadori A, Mori E. Pure word deafness following bilateral lesions. A psychophysical analysis. *Brain* 1987; **110**: 381-403

Cross References

Agnosia; Amusia; Aphasia

Pyramidal Signs, Pyramidal Weakness

- see HEMIPARESIS; UPPER MOTOR NEURONE (UMN) SYNDROME; WEAKNESS

Q

Quadrantanopia

Quadrantanopia (quadrantanopsia), a defect in one quarter of the visual field, suggests an optic radiation lesion. Occipital lobe pathology is the most common cause of both inferior and superior quadrantanopias, although temporal lobe pathology damaging Meyer's loop typically must be considered with a superior homonymous quadrantanopia ("pie-in-the-sky" defect). Parietal lobe lesions may produce inferior quadrantic defects, usually accompanied by other localizing signs. Damage to extrastriate visual cortex (areas V2 and V3) has also been suggested to cause quadrantanopia; concurrent central achromatopsia favors this localization.

References

Horton JC, Hoyt WF. Quadrantic visual field defects. A hallmark of lesions in extrastriate (V2/V3) cortex. *Brain* 1991; **114**: 1703-1718

Jacobson DM. The localizing value of quadrantanopia. *Archives of Neurology* 1997; **54**: 401-404

Cross References

Achromatopsia; Hemianopia; "Pie-in-the-sky" defect

Quadriparesis, Quadriplegia

Quadriparesis and quadriplegia (tetraparesis, tetraplegia) refer to weakness (partial or total, respectively) of all four limbs which may be of upper motor neurone or, less commonly, lower motor neurone type (*e.g.*, in Guillain-Barré syndrome). As with hemiplegia, upper motor neurone quadriplegia may result from lesions of the corticospinal pathways anywhere from motor cortex to cervical cord via the brainstem, but is most commonly seen with brainstem and upper cervical cord lesions. In such circumstances, respiration may be affected.

Cross References

Hemiparesis; Paraplegia

R

Rabbit Syndrome

Rabbit syndrome is a rest tremor of the perioral and nasal muscles, which may occur with antipsychotic drug therapy and in idiopathic Parkinson's disease. It is therefore presumably related to dopamine deficiency. Drug-induced rabbit syndrome may remit with drug withdrawal.

References

Decina P, Caracci G, Scapicchio PL. The rabbit syndrome. *Movement Disorders* 1990; **5**: 263

Cross References

Parkinsonism

"Raccoon Eyes"

"Raccoon eyes" refers to an appearance of bilateral periorbital ecchymosis, appearing 48-72 hours after an anterior basal skull fracture.

Radiculopathy

A radiculopathy is a disorder of nerve roots, causing pain in a radicular distribution, paresthesia, sensory diminution or loss in the corresponding dermatome, and lower motor neurone type weakness with reflex diminution or loss in the corresponding myotome. Radiculopathies may be single or multiple (polyradiculopathy, *e.g.*, cauda equina syndrome). There may be concurrent myelopathy, typically of extrinsic or extramedullary type.

Most radiculopathies are in the lumbosacral region (60-90%), followed by the cervical region (5-30%).

Electrophysiological studies may be helpful in distinguishing radiculopathy from a neuropathy or plexopathy: sensory nerve action potentials (SNAPs) are normal for intrathecal root lesions, and EMG shows involvement of paraspinal muscles.

- Recognized causes of radiculopathy include:

  Structural lesions:

  Compression: disc protrusion: cervical (especially C6, C7), lumbar (L5, S1) >>> thoracic; bony metastases; spondylolisthesis; fracture; infection.

  Root avulsion, *e.g.*, C5/C6, "waiter's tip" posture; C8/T1, claw hand +/– Horner's syndrome.

  Diabetic polyradiculopathy: thoraco-abdominal, lumbosacral (= diabetic amyotrophy, also known as diabetic lumbar sacral plexopathy, proximal diabetic neuropathy; especially involves L2-L4).

  Neoplasia: with meningeal symptoms, due to spread from carcinoma of breast or lung, melanoma, nonHodgkin's lymphoma, leukemia.

Infection: HIV (CMV late in the course), *Borrelia* (Lyme disease), syphilis (tabes dorsalis), herpes zoster (thoracic > cervical > lumbosacral; sensory >> motor), leprosy.
Demyelination: Guillain-Barré syndrome, chronic inflammatory demyelinating polyradiculopathy (CIDP).

References

Chad DF. Nerve root and plexus disorders. In: Bogousslavsky J, Fisher M (Eds.). *Textbook of neurology*. Boston: Butterworth-Heinemann, 1998: 491-506
Feldman EL, Grisold W, Russell JW, Zifko U. *Atlas of neuromuscular disease. A practical guideline*. Vienna: Springer, 2005: 117-139

Cross References

Cauda equina syndrome; Lasègue's sign; Myelopathy; Neuropathy; Paresthesia; Plexopathy; Reflexes; "Waiter's tip" posture; Weakness

Rebound Phenomenon

This is one feature of the impaired checking response seen in cerebellar disease, along with dysdiadochokinesia and macrographia. It may be demonstrated by observing an overshoot of the outstretched arms when they are released suddenly after being pressed down by the examiner, or suddenly releasing the forearm flexed against resistance so that it hits the chest (Stewart-Holmes sign). Although previously attributed to hypotonia, it is more likely a reflection of asynergia between agonist and antagonist muscles.

Cross References

Asynergia; Ataxia; Dysdiadochokinesia; Dysmetria; Hypotonia, Hypotonus; Macrographia

Recruitment

Recruitment, or loudness recruitment, is the phenomenon of abnormally rapid growth of loudness with increase in sound intensity, which is encountered in patients with sensorineural (especially cochlear sensory) hearing loss. Thus patients have difficulty with sounds of low to moderate intensity ("Speak up, doctor") but intense sounds are uncomfortably loud ("There's no need to shout, doctor!"). Speech discrimination is relatively unimpaired in conductive hearing loss.

"Recruitment" may also be used to refer to pathological "spread" of tendon reflexes, implying broadening of their receptive field.

Cross References

Reflexes

Reduplicative Paramnesia

Reduplicative paramnesia is a delusion in which patients believe familiar places, objects, individuals or events to be duplicated. The syndrome is probably heterogeneous and bears some resemblance to the Capgras delusion as described by psychiatrists.

Reduplicative paramnesia is commoner with right (nondominant) hemisphere damage; frontal, temporal and limbic system damage has

been implicated. This may occur transiently as a consequence of cerebrovascular disease, following head trauma, or even after migraine attacks, or more persistently in the context of neurodegenerative disorders, such as Alzheimer's disease.

References

Benson DF, Gardner H, Meadows JC. Reduplicative paramnesia. *Neurology* 1976; **26**: 147-151

Cross References

Delusion; Paramnesia

Reflexes

Reflex action – a sensory stimulus provoking an involuntary motor response – is a useful way of assessing the integrity of neurological function, since disease in the afferent (sensory) limb, synapse, or efferent (motor) limb of the reflex arc may lead to dysfunction, as may changes in inputs from higher centres.

Different types of reflex may be distinguished. Muscle tendon reflexes (myotactic reflexes) may be either tonic (in response to a static applied force: "stretch reflex") or phasic (in response to a brief applied force, for example a blow from a tendon hammer to the muscle tendon). The latter are of particular use in clinical work because of their localizing value (see Table). However, there are none between T2 and T12, and thus for localization one is dependent on sensory findings, or occasionally cutaneous (skin or superficial) reflexes, such as the abdominal reflexes.

Reflex	Root Value
Jaw jerk	Trigeminal (V) nerve
Supinator (Brachioradialis, Radial)	C5, C6
Biceps	C5, C6
Triceps	C7
Finger flexion (Digital)	C8, T1
Abdominal	T7 to T12
Cremasteric	L1, L2
Knee (Patellar)	L3, L4
Hamstring	L5, S1
Ankle (Achilles)	(L5) S1 (S2)
Bulbocavernosus	S2, S3, S4
Anal	S4, S5

Tendon reflex responses are usually graded on a five point scale:

– absent (areflexia; as in lower motor neurone syndromes, such as peripheral nerve or anterior horn cell disorders; or acute upper motor neurone syndromes, *e.g.*, "spinal shock")

+/– present only with reinforcement (Jendrassik's maneuver); hyporeflexia

\+ normal

++ brisk normal

+++ pathologically brisk (hyperreflexia, as in upper motor neurone syndromes)

Reflex "spread," or "recruitment," for example a finger jerk when eliciting the supinator or biceps jerk, is suggestive of corticospinal pathway (upper motor neurone) pathology, producing an enlarged receptive field for the reflex response; concurrent disruption of the local reflex arc may result in inverted reflexes (*q.v.*).

Reflex responses may vary according to the degree of patient relaxation or anxiety (pre-contraction). Moreover, there is interobserver variation in the assessment of tendon reflexes (as with all clinical signs): a biasing effect of prior knowledge upon reflex assessment has been recorded.

There is also a class or "primitive," "developmental," or "psychomotor" signs, present in neonates but disappearing with maturity but which may re-emerge with ageing or cerebral (especially frontal lobe) disease, hence sometimes known as "frontal release signs."

References

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

Donaghy M, Compston A, Rossor M, Warlow C. Clinical diagnosis. In: Donaghy M (ed.). *Brain's diseases of the nervous system* (11th edition). Oxford: OUP, 2001: 1-59 [at 26-32]

Stam J, van Crevel H. Reliability of the clinical and electromyographic examination of tendon reflexes. *Journal of Neurology* 1990; **237**: 427-431

Cross References

Age-related signs; Areflexia; Crossed adductor reflex; Facilitation; Frontal release signs; Hyperreflexia; Hyporeflexia; Inverted reflexes; Jendrassik's maneuver; Lower motor neurone (LMN) syndrome; Primitive reflexes; Pupillary reflexes; Upper motor neurone (UMN) syndrome; Woltman's sign. See also specific (named) reflexes

Relative Afferent Pupillary Defect (RAPD)

An afferent pupillary defect (APD), or relative afferent pupillary defect (RAPD), is an abnormal pupillary response in which the normally equal direct and consensual pupillary reflexes are asymmetric, the direct response being less than the consensual. This may be particularly evident using the "swinging flashlight" test, in which the two pupils are alternately illuminated every 2-3 seconds in a darkened room. Quickly moving the light to the diseased side may produce pupillary dilatation (Marcus Gunn pupil). Subjectively patients may note that the light stimulus seems less bright in the affected eye.

RAPD suggests an asymmetric optic nerve pathology, such as optic neuritis or tumor, causing a conduction defect; indeed this is the most sensitive sign of optic nerve pathology. Although visual acuity may also be impaired in the affected eye, and the disc appear abnormal on funduscopy, this is not necessarily the case. Since RAPD depends on asymmetry of optic nerve conduction, no defect may be observed if both optic nerves are affected.

RAPD has also been described with lesions of the retina, optic chiasm, optic tract (contralateral), brachium of the superior colliculus and pretectal nucleus (in the latter two situations without visual impairment).

References

Chen CJ, Scheufele M, Sheth M, Torabi A, Hogan N, Frohman EM. Isolated relative afferent pupillary defect secondary to contralateral midbrain compression. *Archives of Neurology* 2004; **61**: 1451-1453

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

Amblyopia; Marcus gunn pupil, Marcus gunn sign; Pupillary reflexes; Swinging flashlight sign

Religiosity

- see HYPERRELIGIOSITY

Remote Atrophy

- see ATROPHY; "FALSE-LOCALIZING SIGNS"

Retinal Venous Pulsation

Venous pulsation is evident in the normal retina when observed with an ophthalmoscope, particularly at the margin of the disc. It is sometimes difficult to see, and may be more obvious in the recumbent position (because of higher pressure within the retinal veins). Venous pulsation is lost when intracranial pressure rises above venous pressure. This is a sensitive marker of raised intracranial pressure, and may be an early sign of impending papilledema; however, venous pulsation may also be absent in pseudopapilledema, and sometimes in normal individuals.

References

Jacks AS, Miller NR. Spontaneous retinal venous pulsation: etiology and significance. *Journal of Neurology, Neurosurgery and Psychiatry* 2003; **74**: 7-9

Cross References

Papilledema; Pseudopapilledema

Retinitis Pigmentosa

Retinitis pigmentosa is a generic name for an inherited retinal degeneration, characterized clinically by typical appearances on ophthalmoscopy, with peripheral pigmentation of "bone-spicule" type, arteriolar attenuation, and eventually unmasking of choroidal vessels and optic atrophy. This process may be asymptomatic in its early stages, but may later be a cause of nyctalopia (night blindness), and produce a mid-peripheral ring scotoma on visual field testing.

A variety of genetic causes of isolated retinitis pigmentosa have been partially characterized:

- autosomal recessive: linked to chromosome 1q
- X-linked: Xp11, Xp21

- autosomal dominant: 3q, 6p, 8

In most cases, patients with retinitis pigmentosa have no associated systemic or extraocular abnormalities, but there are a number of multisystem disorders in which it occurs:

- Abetalipoproteinemia (Bassen-Kornzweig syndrome)
- Kearns-Sayre syndrome, mitochondrial disorders in general
- Lawrence-Moon-Bardet-Biedl syndrome
- Refsum's disease
- Usher's syndrome.

Cross References
Nyctalopia; Optic atrophy; Scotoma

Retinopathy

Retinopathy is a pathological process affecting the retina, with changes observable on ophthalmoscopy; dilatation of the pupil aids observation of the peripheral retina. Common causes include:

- Diabetes mellitus: various abnormalities may occur, in both insulin-dependent (IDDM) and noninsulin dependent (NIDDM) patients. "Background" diabetic retinopathy is manifest as microaneuryms, dot and blot hemorrhages, hard exudates, and diffuse retinal edema, all of which may be asymptomatic. Edema and hard exudates at the macula are a common cause of visual impairment. Proliferative retinopathy is characterized by neovascularization of the disc due to retinal hypoxia, typically in IDDM, with the risk of vitreous hemorrhage, traction retinal detachment and irreversible visual loss. Laser treatment of new vessels is the treatment of choice.
- Hypertension: hypertensive retinopathy may cause arteriolar constriction, with the development of cotton-wool spots; and abnormal vascular permeability causing flame-shaped hemorrhages, retinal edema and hard exudates; around the fovea, the latter may produce a macular star. Optic disc swelling may be seen in malignant hypertension. Arteriosclerosis, thickening of vessel walls with prolonged hypertension, may cause changes at arteriovenous crossings ("AV nipping"). Systemic hypertension is associated with an increased risk of branch retinal vein and central retinal artery occlusion.
- Drug-induced, for example, antimalarials (chloroquine); chlorpromazine.
- Retinitis pigmentosa (*q.v.*).
- Serous retinopathy or chorioretinopathy: leakage of fluid into the subretinal space, causing unilateral sudden nonprogressive visual loss.
- Cancer-associated retinopathy: arteriolar narrowing, optic atrophy.
- "Salt and pepper" retinopathy of Kearns-Sayre syndrome (mitochondrial disorder)

An electroretinogram (ERG) may be helpful in confirming the presence of a retinopathic disorder.

Cross References

Maculopathy; Retinitis pigmentosa; Scotoma

Retrocollis

Retrocollis is an extended posture of the neck. Progressive supranuclear palsy (PSP; Steele-Richardson-Olszewski syndrome) is commonly associated with retrocollis (*cf*. antecollis in multiple system atrophy). Retrocollis may also be a feature of cervical dystonia (torticollis) and of kernicterus.

Cross References

Antecollis; Dystonia; Parkinsonism; Torticollis

Retropulsion, Retropulsion Test

- see FESTINANT GAIT, FESTINATION; POSTURAL REFLEXES; "PULL TEST"

Reverse Lhermitte's Sign

- see LHERMITTE'S SIGN

Reverse Phalen's Maneuver

- see PHALEN'S SIGN

"Reverse Ptosis"

- see HORNER'S SYNDROME; PTOSIS

Reverse Straight Leg Raising

- see FEMORAL STRETCH TEST

Revilliod's Sign

Revilliod's sign is an acquired inability to wink. This is a sign, possibly early, of corticobulbar disease.

Riddoch's Phenomenon

Riddoch's phenomenon is the dissociation of the perception of static and kinetic visual stimuli (statokinetic dissociation). This phenomenon does not have particular localization value, since it may occur with both occipital and anterior visual pathway lesions.

References

Zeki S, Ffytche DH. The Riddoch syndrome: insights into the neurobiology of conscious vision. *Brain* 1998; **121**: 25-45

Cross References

Akinetopsia; Visual agnosia

Right-Left Disorientation

Right-left disorientation is an inability to say whether a part of the body is on the right or left side, or to use a named body part to command.

This may occur in association with acalculia, agraphia, and finger agnosia, collectively known as the Gerstmann syndrome. Although all these features are dissociable, their concurrence indicates a posterior parietal dominant hemisphere lesion involving the angular and supramarginal gyri.

Cross References

Acalculia; Agraphia; Autotopagnosia; Finger agnosia; Gerstmann syndrome

Rigidity

Rigidity is an increased resistance to the passive movement of a joint which is constant throughout the range of joint displacement and not related to the speed of joint movement; resistance is present in both agonist and antagonist muscles. In these particulars, rigidity differs from spasticity. Rigidity also needs to be differentiated from stiffness (*q.v.*).

Rigidity may be described as:

- *Consistent*: "lead-pipe rigidity"; or
- *Jerky*: "cogwheel rigidity" or Negro's sign, when a rhythmic fluctuation (*i.e.*, tremor), like a ratchet or cogwheel, is superimposed on the background of sustained rigidity (NB cogwheeling, reflecting underlying tremor, may occur in the absence of rigidity, *e.g.*, in essential tremor).

The presence of rigidity may be made more obvious by reinforcing maneuvers (*e.g.*, clenching and relaxing the contralateral fist, performing mental arithmetic), a finding variously known as activated rigidity, or Froment's sign, or synkinesis (but note that both Froment's sign and synkinesis have other meanings too). However, this may occur in some normal subjects; it is most helpful in the diagnosis of Parkinson's disease if unilateral. Rigidity may also be demonstrated using Wartenberg's swing test (*q.v.*).

Rigidity is a feature of parkinsonism and may coexist with any of the other clinical features of extrapyramidal system disease, but particularly akinesia (akinetic-rigid syndrome); both are associated with loss of dopamine projections from the substantia nigra to the putamen. Rigidity is a feature of pathology within the basal ganglia.

The pathophysiology of rigidity is thought to relate to overactivity of tonic stretch reflexes in the spinal cord due to excessive supraspinal drive to spinal cord α-motor neurones following loss of descending inhibition as a result of basal ganglia dysfunction. In other words, there is a change in the sensitivity of the spinal interneurones which control α-motor neurones due to defective supraspinal control. Hence rigidity is a positive or release symptom, reflecting the operation of intact suprasegmental centres. The physiological correlate of this is the increased EMG activity found in rigid muscles with increased 1A afferent fiber activity, suggesting maintained α-γ linkage. In support of this, pyramidotomy has in the past been shown to produce some relief of rigidity.

Rigidity in Parkinson's disease may be lessened by treatment with levodopa preparations. The techniques of modern stereotactic neurosurgery may also be helpful, particularly stimulation of the subthalamic nucleus, although both thalamotomy and pallidotomy may also have an effect.

The term rigidity may also be used to describe:

- Posturing associated with coma: decorticate or decerebrate, flexor and extensor posturing respectively;
- A lack of mental flexibility, particularly evident in patients with frontal lobe dysfunction.

References

Cantello R, Gianelli M, Civardi C, Mutani R. Pathophysiology of Parkinson's disease rigidity: role of corticospinal motor projections. *Advances in Neurology* 1996; **69**: 129-133

Meara RJ, Cody FWJ. Relationship between electromyographic activity and clinically assessed rigidity studied at the wrist joint in Parkinson's disease. *Brain* 1992; **115**: 1167-1180

Cross References

Decerebrate rigidity; Decorticate rigidity; Froment's sign; Frontal lobe syndromes; Parkinsonism; Stiffness; Synkinesia, Synkinesis; Tremor; Wartenberg's swing test

Ring Scotoma

- see ANNULAR SCOTOMA; SCOTOMA

Rinne's Test

Rinne's test is one of the tuning fork tests (512 Hz fork preferred), which is used to define whether there is a conductive element to hearing loss. The patient is asked to compare the loudness of a vibrating tuning fork held at the external auditory meatus (air conduction; AC) with the loudness of the fork held against the mastoid process (bone conduction; BC); masking of the other ear, for example by rubbing the tragus, is advised. Normally air conduction is louder (AC > BC). If bone conduction sounds louder (BC > AC), then this is indicative of a conductive hearing loss. In sensorineural hearing loss, AC and BC are diminished to a similar extent, and air conduction remains louder (AC > BC).

References

Miyamoto RT, Wynne MK. Approach to the patient with hearing loss. In: Biller J (ed.). *Practical neurology* (2nd edition). Philadelphia: Lippincott Williams & Wilkins, 2002: 206-226

Cross References

Weber's test

Risus Sardonicus

Risus sardonicus ("sardonic smile") due to spasm of the facial musculature is a classic feature of the neuromuscular syndrome of tetanus, now

exceptionally rarely seen in the developed nations. Risus sardonicus may also occur in the context of dystonia, more usually symptomatic (secondary) than idiopathic (primary) dystonia.

Cross References

Dystonia; Spasm

"Rocket Sign"

The so-called "rocket sign" is a toppling backward, after jumping to the feet from the sitting position, due to postural instability, seen in progressive supranuclear palsy (PSP) and ascribed to frontal lobe dysfunction. A history of falls due to postural instability in the first year after disease onset is one of the mandatory inclusion criteria for the diagnosis of PSP.

Cross References

Parkinsonism; "Wheelchair sign"

Roger's Sign

Roger's sign, or the numb chin syndrome, is an isolated neuropathy affecting the mental branch of the mandibular division of the trigeminal (V) nerve, causing pain, swelling, and numbness of the lower lip, chin and mucous membrane inside the lip. This is usually a sign of metastatic spread of cancer to the jaw.

Hypoesthesia involving the cheek, upper lip, upper incisors and gingiva, due to involvement of the infraorbital portion of the maxillary division of the trigeminal nerve ("numb cheek syndrome") is also often an ominous sign, resulting from recurrence of squamous cell carcinoma of the face infiltrating the nerve.

References

Acarin N. Roger's sign. Chin neuropathy. *Medicina Clâinica (Barcelona)* 1985; **84**: 546

Campbell WW Jr. The numb cheek syndrome: a sign of infraorbital neuropathy. *Neurology* 1986; **36**: 421-423

Roger H, Paillas J. Le signe du mentonnier (parasthésie et anesthésie unilatérale) révélateur d'un processus néoplasique métastatique. *Revue Neurologique (Paris)* 1937; **2**: 751-752

"Rolex" Sign

Apparent malfunction of self-winding (Rolex) watches, which depend on movement of the arm, may occur when they are worn on a hypokinetic, rigid arm; this may be the first sign of a parkinsonian syndrome.

Cross References

Parkinsonism; Wartenberg's swing test

Rombergism, Romberg's Sign

Romberg's sign is adjudged present (or positive) when there is a dramatic increase in unsteadiness, sometimes with falls, after eye closure in a patient standing comfortably (static Romberg's test). Before asking the patient to close his or her eyes, it is advisable to position ones arms in such a way as to be able to catch the patient should they begin to fall.

Patients may fall forward immediately on eye closure ("sink sign"). These phenomena result from sensory ataxia (*i.e.*, loss of proprioception from the feet), which occurs most commonly with posterior column spinal cord disease: Romberg's sign may be seen in tabes dorsalis.

A modest increase in sway on closing the eyes may be seen in normal subjects, and patients with cerebellar ataxia, frontal lobe ataxia, and vestibular disorders (toward the side of the involved ear); on occasion these too may produce an increase in sway sufficient to cause falls. Hence, Romberg's test is not specific. Posturography is an attempt to quantify the Romberg test.

Large amplitude sway without falling, due to the patient clutching hold of the physician, has been labeled "psychogenic Romberg's sign," an indicator of functional stance impairment.

Heel-toe (tandem) walking along a straight line is sometimes known as the dynamic Romberg's test.

References

Pearce JMS. Romberg's sign. In: Pearce JMS. *Fragments of neurological history*. London: Imperial College Press, 2003: 373-374

Cross References

Ataxia; Functional weakness and sensory disturbance; Proprioception; Tandem walking

Rooting Reflex

The rooting reflex is a turning of the head toward a tactile stimulus on the face or an object approaching the mouth, a normal response in infants which is lost during childhood. Its presence in adults is indicative of diffuse premotor frontal disease, this being a primitive reflex or frontal release sign.

References

Rossor M. Snouting, pouting and rooting. *Practical Neurology* 2001; **1**: 119-121

Cross References

Age-related signs; Frontal release signs; Primitive reflexes

Rosenbach's Sign

- see ABDOMINAL REFLEXES

Ross's Syndrome

- see HOLMES-ADIE PUPIL, HOLMES-ADIE SYNDROME

"Round the Clock" Weakness

- see SEQUENTIAL PARESIS

"Round the Houses" Sign

This name has been proposed for the inability of patients with progressive supranuclear palsy to produce pure vertical saccades along a straight line in the visual midline. Full vertical excursions are still present, but only accomplished by saccades moving the eyes in a lateral arc ("round the houses").

References
Quinn N. The "round the houses" sign in progressive supranuclear palsy. *Annals of Neurology* 1996; **40**: 951

Roving Eye Movements

Roving eye movements consist of slow drifting movements of the eyes from side to side; the eyelids are closed and there may be slight divergence of the ocular axes. Roving eye movements may be seen in normal sleep, but also in comatose patients in whom they are indicative of an intact brainstem (*e.g.*, the early diencephalic stage of central herniation) but are otherwise nonlocalizing . As coma deepens, roving eye movements are lost before the movements provoked by the oculocephalic (doll's head) maneuver (oculocephalic reflexes, vestibulo-ocular reflexes), or the caloric tests. Roving eye movements cannot be mimicked, hence their presence excludes psychiatric coma or pseudocoma.

Cross References
Caloric testing; Coma; Vestibulo-ocular reflexes

Rubral Tremor

- see HOLMES'S TREMOR; TREMOR

S

Saccades

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

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

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

References

Carpenter R. The saccadic system: a neurological microcosm. *Advances in Clinical Neuroscience & Rehabilitation* 2004; **4(1)**: 6-8

Leigh RJ, Riley DE. Eye movements in parkinsonism: it's saccadic speed that counts. *Neurology* 2000; **54**: 1018-1019

Cross References

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

Saccadic Intrusion, Saccadic Pursuit

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

Cross References

Ocular flutter; Opsoclonus; Saccades; Square-wave jerks

Saccadomania

- see OPSOCLONUS

Sacral Sparing

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

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

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

Cross References

Dissociated sensory loss; Myelopathy; Paraparesis

Saddle Anesthesia

- see ANESTHESIA; CAUDA EQUINA SYNDROME

Saturday Night Palsy

- see WRIST DROP

Scanning Speech

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

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

Cross References

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

Scapula Alata
- see WINGING OF THE SCAPULA

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

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

Scoliosis
- see KYPHOSCOLIOSIS

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

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

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

Central scotoma
Cecocentral or centrocecal scotoma
Arcuate scotoma
Annular or ring scotoma
Junctional scotoma
Junctional scotoma of Traquair
Peripapillary scotoma (enlarged blind spot)

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

"Scratch Test"

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

References

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

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

Cross References
Proprioception; Vibration

Seborrhea

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

Cross References
Parkinsonism

Seelenblindheit

- see VISUAL AGNOSIA

Seizures

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

- *Epileptic*:
  Idiopathic generalized: tonic-clonic ("grand mal"); absence attack ("petit mal"); myoclonic epilepsy
  Partial: simple (no impairment of consciousness), for example jerking of one arm, which may spread sequentially to other body parts (jacksonian march); or complex, in which

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

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

- *Nonepileptic*:

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

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

References

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

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

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

Cross References

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

Self-Mutilation

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

Sensory Ataxia

- see ATAXIA; ROMBERGISM, ROMBERG'S SIGN

Sequential Paresis

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

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

Cross References

Hemiparesis; Paresis; Quadriparesis, Quadriplegia

Setting Sun Sign

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

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

Cross References

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

Shadowing

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

Cross References

Dementia

Shin-Tapping

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

References

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

Cross References

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

Sialorrhea

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

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

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

References

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

Cross References
Bulbar palsy; Parkinsonism

Sighing
Occasional deep involuntary sighs may occur in multiple system atrophy. Sighing is also a feature, along with yawning, of the early (diencephalic) stage of central herniation of the brainstem with an otherwise normal respiratory pattern. Sudden inspiratory or expiratory sighs are said to be a feature of the hyperkinetic choreiform dysarthria characteristically seen in choreiform disorders, such as Huntington's disease.
References
Quinn N. Multiple system atrophy. In: Marsden CD, Fahn S (eds.). *Movement disorders 3*. Boston: Butterworth, 1994: 262-281
Cross References
Blinking; Dysarthria; Yawning

Signe de l'Eventail **(Fan Sign)**
- see BABINSKI'S SIGN (1)

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

Signe du Journal
- see FROMENT'S SIGN

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

Simian Posture
- see PARKINSONISM

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

Two types of simultanagnosia are described:

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

References

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

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

Cross References

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

Singultus

- see HICCUPS

"Sink Sign"

- see ROMBERG'S SIGN, ROMBERGISM

Skew Deviation

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

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

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

References

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

Cross References

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

Smile-Wink Phenomenon

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

References

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

Cross References

Dysarthria; Facial paresis; Hemiparesis; Hypoesthesia

Snarling Facies

- see "MYASTHENIC SNARL"

Sneezing

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

References

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

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

Cross References

Lateral medullary syndrome

Snoring

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

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

References

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

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

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

Cross References

Hypersomnolence

Snout Reflex

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

Cross References

Frontal release signs; Pout reflex; Primitive reflexes

Somatoparaphrenia

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

Cross References

Anosognosia; Autotopagnosia; Confabulation

Somatotopagnosia

- see AUTOTOPAGNOSIA

Spasm

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

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

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

References

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

Cross References

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

Spasmus Nutans

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

Cross References

Nystagmus

Spastic Catch

- see SPASTICITY

Spasticity

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

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

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

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

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

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

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

Treatment of severe spasticity, for example in multiple sclerosis, often requires a multidisciplinary approach. Urinary infection, constipation, skin ulceration and pain can all exacerbate spasticity, as may inappropriate posture; appropriate management of these features may ameliorate spasticity. Drugs which may be useful include baclofen, dantrolene (a blocker of muscle excitation-contraction coupling), and tizanidine (α_2-adrenoreceptor agonist). Intrathecal baclofen given via a pump may also be of benefit in selected cases, and for focal spasticity injections of botulinum toxin may be appropriate. For painful immobile spastic legs with reflex spasms and double incontinence, irreversible nerve injury with intrathecal phenol or alcohol may be advocated to relieve symptoms. The place of cannabinoids has yet to be fully determined.

References

Shakespeare DT, Boggild M, Young C. Anti-spasticity agents for multiple sclerosis. *Cochrane Database of Systematic Reviews* 2003; **4**: CD001332

Sheean G. Neurophysiology of spasticity. In: Barnes MP, Johnson GR (eds.). *Upper motor neurone syndrome and spasticity. Clinical management and neurophysiology*. Cambridge: CUP, 2001: 12-78

Cross References

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

Speech Apraxia

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

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

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

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

References

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

Cross References

Aphasia; Aphemia; Apraxia

Spinal Mass Reflex

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

Cross References

Coma

Spurling's Sign

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

Cross References

Radiculopathy

Square-Wave Jerks

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

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

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

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

Square-Wave Oscillations
- see SQUARE-WAVE JERKS

Squint
- see HETEROTROPIA

Stapedius Reflex
- see HYPERACUSIS

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

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

Cross References
Blinking; Lid lag; Lid retraction; Setting sun sign

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

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

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

Stereoanesthesia
- see ASTEREOGNOSIS

Stereohypesthesia
- see ASTEREOGNOSIS

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

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

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

References

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

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

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

Cross References

Aphasia; Broca's aphasia; Tic

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

References

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

Cross References

Functional weakness and sensory disturbance; Hemiparesis

Stethoscope Loudness Imbalance Test
- see HYPERACUSIS

Stewart-Holmes Sign
- see REBOUND PHENOMENON

Stiffness

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

Recognized causes of stiffness include:
Stiff man/stiff person syndrome
Stiff limb syndrome
Progressive encephalomyelitis with rigidity +/–myoclonus
Neuromyotonia (Isaac's syndrome; armadillo syndrome)
Schwartz-Jampel syndrome (chondrodystrophic myotonia)
Tetanus
Strychnine poisoning

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

References

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

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

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

Cross References

Myokymia; Myotonia; Neuromyotonia; Paramyotonia; Rigidity; Spasticity

"Stork Legs"

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

Cross References

Foot drop; Steppage, Stepping gait; Wasting

Strabismus
- see HETEROPHORIA; HETEROTROPIA

Straight Leg Raising
- see LASÈGUE'S SIGN

"Straight Thumb Sign"
Median nerve lesions in the forearm cause weakness of flexor pollicis longus, which normally flexes the distal phalanx of the thumb. Hence the thumb remains straight when the patient attempts to grasp something or make a fist. The "pinch sign" may also be present.
References
Cherington M. Anterior interosseous nerve syndrome straight thumb sign. *Neurology* 1977; **27**: 800-801
Cross References
"Pinch sign"

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

Striatal toe may be confused with Babinski's sign (extensor plantar response), the principal difference being that the latter is elicited by stimulation whereas the former is a tonic response.
References
Winkler AS, Reuter I, Harwood G, Chaudhuri KR. The frequency and significance of "striatal toe" in parkinsonism. *Parkinsonism and Related Disorders* 2002; **9**: 97-101
Cross References
Babinski's sign (1); Parkinsonism

Stupor
Stupor is a state of altered consciousness characterized by deep sleep or unresponsiveness, susceptible to arousal only by vigorous and/or repeated stimuli, with lapse back into unresponsiveness when the stimulus stops. Stupor is a less severe impairment of conscious level than coma, but worse than obtundation (torpor). It is suggestive of diffuse cerebral dysfunction, *e.g.*, drug-induced.
References
Plum F, Posner JB. *The diagnosis of stupor and coma* (3rd edition). Philadelphia: FA Davis, 1980
Cross References
Coma; Delirium; Encephalopathy; Obtundation

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

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

References

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

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

Cross References

Aphasia; Echolalia; Palilalia

Sucking Reflex

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

Cross References

Akinetic mutism; Dementia; Frontal release signs

Summerskill's Sign

- see LID RETRACTION

"Sundowning"

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

References

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

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

Cross References

Delirium; Dementia

Sunset Sign

- see SETTING SUN SIGN

Suppression
- see EXTINCTION

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

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

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

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

References
Lees AJ. The Steele-Richardson-Olszewski syndrome (progressive supranuclear palsy). In: Marsden CD, Fahn S (eds.). *Movement Disorders 2*. London: Butterworth, 1987: 272-287
Cross References
Gaze palsy; Parinaud's syndrome; Parkinsonism; Prevost's sign; Skew deviation; Vestibulo-ocular reflexes

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

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

Cross References

Dissociated sensory loss; Myelopathy

"Swan Neck"

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

Swinging Flashlight Sign

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

References

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

Cross References

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

Synesthesia

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

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

References

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

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

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

Cross References

Auditory-visual synesthesia; Phosphene

Synkinesia, Synkinesis

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

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

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

Cross References

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

T

"Table Top" Sign

Inability to place the hand flat on a level surface, recognized causes of which include ulnar neuropathy (*main en griffe*), Dupuytren's contracture, diabetic cheiroarthropathy, and camptodactyly.

Cross References

"Prayer sign"

Tachylalia

Tachylalia is increased speech velocity. This has been reported in patients with cerebrotendinous xanthomatosis, particularly in the 20 to 40 year age group.

References

Verrips A, Van Engelen B, de Swart B *et al*. Increased speech rate (tachylalia) in cerebrotendinous xanthomatosis: a new sign. *Journal of Medical Speech-Language Pathology* 1998; **6**: 161-164.

Tachyphemia

Repetition of a word or phrase with increasing rapidity and decreasing volume; may be encountered as a feature of parkinsonian syndromes.

Cross References

Parkinsonism

Tactile Agnosia

A selective impairment of object recognition by touch despite (relatively) preserved somatesthetic perception. This is a unilateral disorder resulting from lesions of the contralateral inferior parietal cortex.

References

Reed CL, Caselli RJ, Farah MJ. Tactile agnosia: underlying impairment and implications for normal tactile object recognition. *Brain* 1996; **119**: 875-888

Cross References

Agnosia

Tadpole Pupils

Pupillary dilatation restricted to one segment may cause peaked elongation of the pupil, a shape likened to a tadpole's pupil. This has been recorded in Horner's syndrome, migraine, and Holmes-Adie pupil.

References

Thompson HS, Zackon DH, Czarnecki JSC. Tadpole-shaped pupils caused by segmental spasm of the iris dilator muscle. *American Journal of Ophthalmology* 1983; **96**: 467-477

Talantropia
- see NYSTAGMUS

Tandem Walking
Tandem walking, or heel-toe walking, also known as the dynamic Romberg's test, is the ability to walk along a straight line placing one foot directly in front of the other, heel to toe, which may be likened to walking a tightrope. In ataxic disorders, cerebellar (midline cerebellum, in which axial coordination is most affected) or sensory (loss of proprioception), the ability to tandem walk is impaired, as reflected by the tendency of such patients to compensate for their incoordination by developing a broad based gait.
Cross References
Ataxia; Cerebellar syndromes; Proprioception; Rombergism, Romberg's sign

Tay's Sign
- see CHERRY RED SPOT AT THE MACULA

Teichopsia
- see FORTIFICATION SPECTRA

Telegraphic Speech
- see AGRAMMATISM

Telopsia
A visual illusion in which the image is altered in position.
Cross References
Illusion; Pelopsia; Porropsia

Temporal Desaturation
Temporal desaturation refers to an impairment in perception of red targets confined to the temporal visual hemifield. This may be the earliest indication of a developing temporal field defect, as in a bitemporal hemianopia due to a chiasmal lesion, or a monocular temporal field defect (junctional scotoma of Traquair) due to a distal ipsilateral optic nerve lesion.
Cross References
Hemianopia; Scotoma

Temporal Pallor
Pallor of the temporal portion of the optic nerve head may follow atrophy of the macular fibre bundle in the retina, since the macular fibers for central vision enter the temporal nerve head. This may be associated with impairment of central vision.
Cross References
Optic atrophy

Terson Syndrome
Vitreous hemorrhage in association with any form of intracranial or subarachnoid hemorrhage.

Tetanus, Tetany
- see *MAIN D'ACCOUCHEUR*; RISUS SARDONICUS; SPASM

Tetraparesis, Tetraplegia
- see QUADRIPARESIS, QUADRIPLEGIA

Threat Reflex
- see BLINK REFLEX

Tic
A tic is an abrupt, jerky repetitive movement involving discrete muscle groups, hence a less complex movement than a stereotypy. Vocal (phonic) tics are also described. Tics vary in intensity, lack rhythmicity, and are relatively easy to imitate. They may temporarily be voluntarily suppressed by will power (perhaps accounting for their previous designation as "habit spasms") but this is usually accompanied by a growing inner tension or restlessness, only relieved by the performance of the movement. The pathophysiology of tics is uncertain. The belief that Gilles de la Tourette syndrome was a disorder of the basal ganglia has now been superseded by evidence of dysfunction within the cingulate and orbitofrontal cortex, perhaps related to excessive endorphin release.

The etiological differential diagnosis of tic includes:

- Idiopathic
- Gilles de la Tourette syndrome
- Tics related to structural brain damage
- Drug-induced tics.

Treatment of tics is most usually with dopamine antagonists (haloperidol, sulpiride) and opioid antagonists (naltrexone); clonidine (central α_2 adrenergic receptor antagonist) and tetrabenazine (dopamine-depleting agent) have also been reported to be beneficial on occasion.

The word tic has also been used to describe the paroxysmal, lancinating pains of trigeminal neuralgia (tic douloureux).

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

Weeks RA, Turjanski N, Brooks DJ. Tourette's syndrome: a disorder of cingulate and orbitofrontal function? *Quarterly Journal of Medicine* 1996; **89**: 401-408

Cross References
Klazomania; Stereotypy

"Tie Sign"
- see VISUAL DISORIENTATION

Tinel's Sign (Hoffmann-Tinel Sign)

Tinel's sign (Hoffmann-Tinel sign) is present when tingling (paresthesia) is experienced when tapping lightly with a finger or a tendon hammer over a compressed or regenerating peripheral nerve. The tingling (Tinel's "sign of formication") is present in the cutaneous distribution of the damaged nerve ("peripheral reference"). Although originally described in the context of peripheral nerve regeneration after injury, Tinel's sign may also be helpful in diagnosing focal entrapment neuropathy, such as carpal tunnel syndrome. However, it is a "soft" sign; like other provocative tests for carpal tunnel syndrome (*e.g.*, Phalen's sign) it is not as reliable for diagnostic purposes as electromyography (EMG). One study found a specificity of 59-77%, and sensitivity of 60-67%.

A "motor Tinel sign" has been described, consisting of motor EMG activity and jerking of muscles evoked by manipulation of an entrapped nerve trunk.

The neurophysiological basis of Tinel's sign is presumed to be the lower threshold of regenerating or injured (demyelinated) nerves to mechanical stimuli, which permits ectopic generation of orthodromic action potentials, as in Lhermitte's sign.

References

Heller L, Ring H, Costeff H, Solzi P. Evaluation of Tinel's and Phalen's signs in the diagnosis of the carpal tunnel syndrome. *European Neurology* 1986; **25**: 40-42

Montagna P. Motor Tinel sign: a new localizing sign in entrapment neuropathy. *Muscle Nerve* 1994; **17**: 1493-1494

Pearce JMS. Hoffmann and Tinel's sign of formication. In: Pearce JMS. *Fragments of neurological history*. London: Imperial College Press, 2003: 375-377

Cross References

Flick sign; Lhermitte's sign; Phalen's sign

Tinnitus

Tinnitus is the perception of elementary nonenvironmental sound or noise in the ear. This is most usually a subjective phenomenon (*i.e.*, heard only by the sufferer), occurring in the absence of acoustic stimulation. It may occur in conjunction with either conductive or sensorineural hearing loss. However, in about one-fifth of sufferers, tinnitus is objective (*i.e.*, heard also by an observer). This may result from:

- Vascular causes: *e.g.*, arteriovenous malformation, fistula; carotid or vertebrobasilar bruit
- Mechanical causes: *e.g.*, palatal myoclonus (ear click).

The common causes of subjective tinnitus are:

- Middle/inner ear disease: cochlear hydrops (Ménière's disease), presbycusis, acoustic tumor
- Pulsatile: normal heartbeat, glomus jugulare tumor, raised intracranial pressure, cervical/intracranial aneurysm, arteriovenous malformation.

Cross References
Hallucination; Palatal myoclonus

"Tip-of-the-Tongue" Phenomenon
- see CIRCUMLOCUTION

Titubation
- see HEAD TREMOR

Todd's Paralysis, Todd's Paresis
Todd's paralysis (Todd's paresis) is a transient localized weakness (usually hemiparesis), lasting seconds to minutes (exceptionally 24 to 48 hours), observed following a focal motor seizure or Jacksonian seizure originating in the central motor strip, or febrile convulsion, a phenomenon first described by RB Todd in 1854. The pattern and duration of post-ictal signs is quite heterogeneous. Aphasia is also described. A postictal "paralytic" conjugate ocular deviation may be observed after adversive seizures. Todd's paresis is of localizing value, being contralateral to the epileptogenic hemisphere.

The differential diagnosis of transient postictal hemiparesis includes stroke, hemiplegic migraine, and, in children, alternating hemiplegia.

References
Binder DK. A history of Todd and his paralysis. *Neurosurgery* 2004; **54**: 480-486

Kellinghaus C, Kotagal P. Lateralizing value of Todd's palsy in patients with epilepsy. *Neurology* 2004; **62**: 289-291

Rolak LA, Rutecki P, Ashizawa T, Harati Y. Clinical features of Todd's post-epileptic paralysis. *Journal of Neurology, Neurosurgery and Psychiatry* 1992; **55**: 63-64

Cross References
Hemiparesis; Seizures

Toe Walking
Toe walking, or cock walking, is walking on the balls of the toes, with the heel off the floor. A tendency to walk on the toes may be a feature of hereditary spastic paraparesis, and the presenting feature of idiopathic torsion dystonia in childhood.

Cross References
Dystonia

Torpor
- see OBTUNDATION

Torticollis
Torticollis (wryneck, cervical dystonia, nuchal dystonia) is a movement disorder characterized by involuntary contraction of neck musculature, involving especially sternocleidomastoid, trapezius, and splenius capitis. In the majority of cases (> 50%) this produces head rotation, but laterocollis, retrocollis, tremulous ("no-no") and complex

(*i.e.*, variable) forms are seen; antecollis is unusual. Contractions are usually unilateral, may be associated with local pain, and, as with other types of dystonia, may be relieved by a "sensory trick" (*geste antagoniste*).

Causes of torticollis include:
Idiopathic (the majority)
Secondary to acquired cervical spine abnormalities, trauma
Cervical spinal tumor
Tardive effect of neuroleptics

The treatment of choice is botulinum toxin injections into the affected muscles. Injections benefit up to 70-80% of patients, but need to be repeated every three months or so.

Cross References

Antecollis; Dystonia; Geste antagoniste; Laterocollis; Retrocollis

Tortopia

- see ENVIRONMENTAL TILT

Transcortical Aphasias

Transcortical aphasias may be categorized as either motor or sensory.

- *Transcortical motor aphasia* (TCMA):
  There is a dissociation between preserved repetition (*cf*. conduction aphasia) and impaired fluency, manifest as delayed initiation, even mutism, impaired lexical selection, and reduced capacity to generate unconstrained syntactic forms. TCMA is associated with pathology (usually infarction) in the supplementary motor area, superior to Broca's area (left lateral frontal cortex) or in subcortical structures including white matter projections and dorsal caudate nucleus; it has clinical similarities with Broca's aphasia.
- *Transcortical sensory aphasia* (TCSA):
  There is a dissociation between preserved repetition (*cf*. conduction aphasia) and impairments of spoken and written language comprehension without phonemic paraphasia. TCSA is associated with pathology (usually infarction) in the ventral and ventrolateral temporal lobe involving the fusiform gyrus and the inferior temporal gyrus, and posterior convexity lesions involving the posterior middle temporal gyrus and the temporo-occipital junction. It has similarities Wernicke's aphasia.

Some authorities prefer to label these conditions as extrasylvian aphasic syndromes, to distinguish them from the perisylvian aphasic syndromes (Broca, Wernicke, conduction); moreover, these syndromes are not "transcortical" in any literal sense.

Dynamic aphasia (*q.v.*) may be a lesser version of TCMA, in which there are no paraphasias and minimal anomia, preserved repetition and automatic speech, but reduced spontaneous speech. This may be associated with lesions of dorsolateral prefrontal cortex ("frontal aphasia") in the context of frontal lobe degeneration. There

may be incorporational echolalia, when the patient uses the examiner's question to help form an answer.

References

Alexander MP. Transcortical motor aphasia: a disorder of language production. In: D'Esposito M (ed.). *Neurological foundations of cognitive neuroscience*. Cambridge: MIT Press, 2003: 165-174

Alexander MP, Hiltbrunner B, Fischer RS. Distributed anatomy of transcortical sensory aphasia. *Archives of Neurology* 1989; **46**: 885-892

Boatman D, Gordon B, Hart J, Selnes O, Miglioretti D, Lenz F. Transcortical sensory aphasia: revisited and revised. *Brain* 2000; **123**: 1634-1642

Cross References

Aphasia; Broca's aphasia; Conduction aphasia; Dynamic aphasia; Echolalia; Paraphasia; Wernicke's aphasia

Transverse Smile

- see "MYASTHENIC SNARL"

Tremblement Affirmatif, Tremblement Negatif

- see HEAD TREMOR

Tremor

Tremor is an involuntary movement, roughly rhythmic and sinusoidal, although some tremors (*e.g.*, dystonic) are irregular in amplitude and periodicity. Tremors may be classified clinically:

- *Rest tremor*:
  present when a limb is supported against gravity and there is no voluntary muscle activation, *e.g.*, the 3.5-7 Hz "pill rolling" hand tremor of Parkinson's disease; midbrain/rubral tremor.
- *Action tremor*:
  present during any voluntary muscle contraction.
  Various subtypes of action tremor are recognized:
- *Postural tremor*:
  present during voluntary maintenance of a posture opposed by gravity, *e.g.*, arm tremor of essential tremor; 6Hz postural tremor sometimes seen in Parkinson's disease, which may predate emergence of akinesia/rigidity/rest tremor; modest postural tremor of cerebellar disease; some drug-induced tremors (including alcohol withdrawal, delirium tremens); tremor of IgM paraproteinemic neuropathy; wing-beating tremor of Wilson's disease.
- *Kinetic tremor*:
  present with movement, often with an exacerbation at the end of a goal-directed movement (intention tremor), *e.g.*, cerebellar/midbrain tremor (3-5Hz).
- *Task-specific tremor*:
  evident only during the performance of a highly-skilled activity, *e.g.*, primary writing tremor.

- *Isometric tremor*:
  present when voluntary muscle contraction is opposed by a stationary object, *e.g.*, primary orthostatic tremor (14-18Hz).
- *Psychogenic tremors*:
  these are difficult to classify, with changing characteristics; the frequency with which such tremors are observed varies greatly between different clinics; the coactivation sign (increase in tremor amplitude with peripheral loading) is said to be typical of psychogenic tremor.

EMG may be useful for determining tremor frequency, but is only diagnostic in primary orthostatic tremor.

Various treatments are available for tremor, with variable efficacy. Essential tremor often responds to alcohol, and this is a reasonable treatment (previous anxieties that such a recommendation would lead to alcoholism seem unjustified); alternatives include propranolol, primidone, topiramate, alprazolam, flunarizine, and nicardipine. In Parkinson's disease, tremor is less reliably responsive to levodopa preparations than akinesia and rigidity; anticholinergics, such as benzhexol, may be more helpful (but may cause confusion). Primary orthostatic tremor has been reported to respond to clonazepam, primidone, and levodopa. Cerebellar tremor is often treated with isoniazid, but seldom with marked benefit, likewise carbamazepine, clonazepam, ondansetron, limb weights; stereotactic surgery may be the optimum treatment if preliminary experimental data are confirmed.

References

Bain PG, Findley LJ. *Assessing Tremor Severity*. London: Smith-Gordon, 1993

Barker R, Burn DJ. Tremor. *Advances in Clinical Neuroscience & Rehabilitation* 2004; **4(1)**: 13-14

Deuschl G, Bain P, Brin M and an Ad Hoc Scientific Committee. Consensus statement of the Movement Disorder Society on tremor. *Movement Disorders* 1998; **13(suppl3)**: 2-23

Findley LJ, Koller WC (eds.). *Handbook of Tremor Disorders*. New York: Marcel Dekker, 1995

Cross References

Asterixis; Coactivation sign; Head tremor; Knee tremor; Parkinsonism; Vocal tremor, Voice tremor; Wing-beating tremor

Trendelenburg's Sign

Trendelenburg's sign is tilting of the pelvis toward the side of the unaffected raised leg in a unilateral superior gluteal nerve lesion.

Triparesis

- see SEQUENTIAL PARESIS

Trismus

Trismus is an inability to open the jaw due to tonic spasm or contracture of the masticatory muscles, principally masseter and temporalis, effecting forced jaw closure ("lockjaw").

Recognized causes and associations of trismus include:
Dystonia of the jaw muscles (*e.g.*, drug-induced dystonic reaction)
Generalized tonic-clonic epileptic seizure
Neuromuscular diseases: polymyositis, tetanus, nemaline myopathy, trauma to the muscles of mastication, rabies, strychnine poisoning
Infection in the pterygomandibular space
Metabolic disorders: Gaucher's disease (type II)
Central disorders: brainstem encephalopathy, multiple sclerosis, pseudobulbar palsy.

References

Lai MM, Howard RS. Pseudobulbar palsy associated with trismus. *Postgraduate Medical Journal* 1994; **70**: 823-824

Cross References

Dystonia; Pseudobulbar palsy

Trombone Tongue

Trombone tongue, or flycatcher tongue, refers to an irregular involuntary darting of the tongue in and out of the mouth when the patient is requested to keep the tongue protruded. This sign may be seen in choreiform movement disorders, such as Huntington's disease and neuroacanthocytosis, and in tardive dyskinesia.

Cross References

Chorea, Choreoathetosis; Impersistence; Milkmaid's grip

Trömner's Sign

Trömner's sign is flexion of the thumb and index finger in response to tapping or flicking the volar surface of the distal phalanx of the middle finger, held partially flexed between the examiner's finger and thumb. This is an alternative method to Hoffmann's sign ("snapping" the distal phalanx) to elicit the finger flexor response. As in the latter, it is suggestive of a corticospinal tract (upper motor neurone) lesion above C5 or C6, especially if unilateral, although it may be observed in some normal individuals.

Cross References

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

Trousseau's Sign

Trousseau described the signs and symptoms of tetany, including anesthesia, paresthesia, the *main d'accoucheur* posture, as well as noting that the latter could be reproduced by applying a bandage or inflating a cuff around the arm so as to impede circulation; the latter is now known as Trousseau's sign, and indicates latent tetany.

Trousseau also noted the concurrence of venous thrombosis and migrating thrombophlebitis with malignant disease, also referred to as Trousseau's sign; this may present with cerebral venous thrombosis.

References

Pearce JMS. Armand Trousseau, physician and neurologist. In: Pearce JMS. *Fragments of neurological history*. London: Imperial College Press, 2003: 528-547

Cross References
Achromatopsia; Chvostek's sign; Main d'accoucheur

Tullio Phenomenon

The Tullio phenomenon is the experience of vestibular symptoms and signs (vertigo, nystagmus, oscillopsia, postural imbalance, ocular tilt reaction, +/– skew deviation) on exposure to high intensity acoustic stimuli, presumed to be due to hyperexcitability of the normal vestibular response to sound, causing pathological stimulation of the semicircular canals and/or otoliths. This unusual phenomenon may be associated with perilymph leaks or a defect in the capsule forming the roof of the anterior semicircular canal. The sound sensitivity is probably at the level of the receptors rather than the vestibular nerve.

References
Watson SRD, Halmagyi GM, Colebatch JG. Vestibular hypersensitivity to sound (Tullio phenomenon). Structural and functional assessment. *Neurology* 2000; **54**: 722-728

Cross References
Nystagmus; Ocular tilt reaction; Oscillopsia; Skew deviation; Vertigo

"Tunnel Vision"

A complaint of "tunnel vision" may indicate constriction of the visual field. This may be observed with enlargement of the blind spot and papilledema as a consequence of raised intracranial pressure or with a compressive optic neuropathy. The visual field enlarges the further away from the eye the visual target used to map the field is held, hence there is in fact "funnel vision." In nonorganic visual impairment, by contrast, the visual field stays the same size with more distant targets (tunnel vision).

A tunnel vision phenomenon has also been described as part of the aura of seizures of anteromedial temporal and occipitotemporal origin. A closing in of vision may be described as a feature of presyncope.

Cross References
Aura; Blind spot; Hemianopia; Papilledema; Visual field defects

Two-Point Discrimination

Two-point discrimination is the ability to discriminate two adjacent point stimuli (*e.g.*, using a pair of calipers) as two rather than one. The minimum detectable distance between the points (acuity) is smaller on the skin of the finger tips (*i.e.*, greater acuity) than, say, the skin on the back of the trunk. Impairments of two-point discrimination may occur with dorsal column spinal cord lesions, in which proprioception (and possibly vibration) is also impaired. Cortical parietal lobe lesions may produce a cortical sensory syndrome of astereognosis, agraphesthesia, and impaired two-point discrimination.

Cross References
Astereognosis; Graphesthesia; Proprioception; Vibration

U

Uhthoff's Phenomenon

Uhthoff's phenomenon (symptom) is the worsening of visual acuity ("amblyopia" in Uhthoff's 1890 description) with exercise in optic neuritis, reflecting the temperature sensitivity of demyelinated axons (*i.e.*, reduced safety factor for faithful transmission of action potentials). The term has subsequently been applied to exercise and/or temperature related symptoms in other demyelinated pathways. It has also been described in the context of other optic nerve diseases, including Leber's hereditary optic neuropathy, sarcoidosis and tumor.

Evidence suggesting that Uhthoff's phenomenon is associated with an increased incidence of recurrent optic neuritis, and may be a prognostic indicator for the development of multiple sclerosis, has been presented.

Inverse Uhthoff sign, improved vision with warming, has been described.

References

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Cross References

Lhermitte's sign; Phosphene

Unterburger's Sign

Unterburger's test examines the integrity of vestibulospinal connections and attempts to define the side of a vestibular lesion. The patient is asked to march on the spot with the eyes closed (*i.e.*, proprioceptive and visual cues are removed); the patient will rotate to the side of a unilateral vestibular lesion (Unterburger's sign). The test is not very useful, particularly in chronic, progressive, or partially compensated vestibular lesions.

Cross References

Proprioception; Vertigo

Upbeat Nystagmus

- see NYSTAGMUS

Upper Motor Neurone (UMN) Syndrome

An upper motor neurone (UMN) syndrome constitutes a constellation of motor signs resulting from damage to upper motor neurone pathways, *i.e.*, proximal to the anterior horn cell. These may be termed "pyramidal signs," but since there are several descending motor pathways (*e.g.*, corticospinal, reticulospinal, vestibulospinal), of which the pyramidal or corticospinal pathway is just one, "upper motor neurone syndrome" is preferable. "Long tract signs" may be a more accurate term, often used interchangeably with "pyramidal signs." The syndrome may be variable in its clinical features but common elements, following the standard order of neurological examination of the motor system, include:

- *Appearance*:
  usually normal, but there may be wasting in chronic UMN syndromes, but this is usually not as evident as in lower motor neurone syndromes; contractures may be evident in chronically spastic limbs
- *Tone*:
  hypertonus, with spasticity, clasp-knife phenomenon and sustained clonus
- *Power*:
  weakness, often in a so-called pyramidal distribution (*i.e.*, affecting extensors more than flexors in the upper limb, and flexors more than extensors in the lower limb); despite its clinical utility, the term pyramidal is, however, a misnomer (see Weakness)
- *Coordination*:
  depending on the degree of weakness, it may not be possible to comment on the integrity of coordination in UMN syndromes; in a pure UMN syndrome coordination will be normal, but syndromes with both ataxia and UMN features do occur (*e.g.*, spinocerebellar syndromes, ataxic hemiparesis syndromes)
- *Reflexes*:
  limb hyperreflexia, sometimes with additional reflexes indicative of corticospinal tract involvement (Hoffmann's sign, Trömner's sign, crossed adductor reflex); Babinski's sign (extensor plantar response); cutaneous reflexes (abdominal, cremasteric) are lost.

The most reliable ("hardest") signs of UMN syndrome are increased tone, clonus, and upgoing plantar responses.

The clinical phenomena comprising the upper motor neurone syndrome may be classified as "positive" and "negative" depending on whether they reflect increased or decreased activity in neural pathways:

- Positive:
  Exaggerated stretch/tendon reflexes, flexor spasms
  Clonus
  Autonomic hyperreflexia
  Contractures

- Negative:
  Muscle weakness
  Loss of dexterity

These features help to differentiate UMN from LMN syndromes, although clinically the distinction is not always easy to make: a "pyramidal" pattern of weakness may occur in LMN syndromes (*e.g.*, Guillain-Barré syndrome) and acute UMN syndromes may cause flaccidity and areflexia (*e.g.*, "spinal shock").

References

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Cross References

Abdominal reflexes; Ataxic hemiparesis; Babinski's sign (1); Clasp-knife phenomenon; Clonus; Contracture; Cremasteric reflex; Hoffmann's sign; Hyperreflexia; Hypertonia, Hypertonus; Lower motor neurone (LMN) syndrome; Pseudobulbar palsy; Spasticity; Trömner's sign; Weakness

Urgency

- see INCONTINENCE

Urinary Retention

Although urinary retention is often urological in origin (*e.g.*, prostatic hypertrophy) or a side effect of drugs (*e.g.*, anticholinergics), it may have neurological causes. It may be a sign of acute spinal cord compression, with or without other signs in the lower limbs, or of acute cauda equina compression, for example with a central L1 disc herniation. Sometimes the level of the pathology is several segments above that expected on the basis of the ("false localizing") neurological signs. Loss of awareness of bladder fullness may lead to retention of urine with overflow.

A syndrome of urinary retention in young women has been described, associated with myotonic-like activity on sphincter EMG; this condition may be associated with polycystic ovary disease and is best treated with clean intermittent self-catheterization.

References

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Jamieson DRS, Teasdale E, Willison HJ. False localizing signs in the spinal cord. *BMJ* 1996; **312**: 243-244

Cross References

Cauda equina syndrome; "False localizing signs"; Incontinence; Myelopathy; Paraplegia; Radiculopathy

Useless Hand of Oppenheim

The deafferented hand or arm is functionally useless, and manifests involuntary movements due to severe proprioceptive loss. This was first described in multiple sclerosis by Oppenheim in 1911, and reflects plaques in the dorsal root entry zone of the relevant spinal cord segment(s).

References
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Cross References
Proprioception; Pseudoathetosis; Pseudochoreoathetosis

Utilization Behavior

Utilization behavior is a disturbed response to external stimuli, a component of the environmental dependency syndrome, in which seeing an object implies that it should be used. Two forms of utilization behavior are described:

- *Induced*:
  when an item is given to the patient or their attention is directed to it, *e.g.*, handing them a pair of spectacles which they put on, followed by a second pair, which are put on over the first pair.
- *Incidental* or *Spontaneous*:
  when the patient uses an object in their environment without their attention being specifically directed toward it.

Another element of the environmental dependency syndrome which coexists with utilization behavior is imitation behavior (*e.g.*, echolalia, echopraxia). Primitive reflexes and hypermetamorphosis may also be observed.

Utilization behavior is associated with lesions of the frontal lobe, affecting the inferior medial area bilaterally. It has also been reported following paramedian thalamic infarction.

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Cross References
Automatic writing behavior; Echolalia; Echopraxia; Frontal lobe syndromes; Hypermetamorphosis; Imitation behavior; Primitive reflexes

V

Valsalva Maneuver

The Valsalva maneuver is a simple test of autonomically-mediated cardiovascular reflexes, comprising forced expiration against resistance ("straining"), followed by release of the resistance and completion of expiration. The first phase produces impaired cardiac filling due to impaired venous return as a consequence of elevated intrathoracic pressure, with a fall in cardiac output and blood pressure, inducing peripheral vasoconstriction (sympathetic pathways) to maintain blood pressure. The second phase causes a transient overshoot in blood pressure as the restored cardiac output is ejected into a constricted circulation, followed by reflex slowing of heart rate.

In autonomic (sympathetic) dysfunction, reflex vasoconstriction, blood pressure overshoot and bradycardia do not occur. The latter may be conveniently assessed by measuring R-R intervals in a prolonged ECG recording, an R-R interval ratio between the straining and release phases of less than 1.1 suggesting impaired baroreceptor response.

Cross References

Orthostatic hypotension

Vegetative States

The vegetative state is a clinical syndrome in which cognitive function is lost, due to neocortical damage (hence no awareness, response, speech), while vegetative (autonomic, respiratory) function is preserved due to intact brainstem centres. Primitive postural and reflex limb movements may also be observed. The syndrome, also known as neocortical death, coma vigil, and the apallic syndrome, may be seen after extensive ischemic-hypoxic brain injury, for example following resuscitation after cardiac arrest, and needs to be distinguished from coma, akinetic mutism, and the locked-in syndrome. Persistent vegetative state (PVS) is defined by persistence of this state for > 12 months (UK) or > 6 months (USA) after brain trauma, or > 6 months (UK) or > 3 months (USA) following brain anoxia. The prognosis of PVS is poor, but occasional reports of very late recovery have appeared.

References

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Wade DT, Johnston C. The permanent vegetative state: practical guidelines on diagnosis and management. *British Medical Journal* 1999; **319**: 841-844

Zeman A. The persistent vegetative state: conscious of nothing? *Practical Neurology* 2002; **2**: 214-217

Cross References

Akinetic mutism; Coma; Locked-in syndrome

Venous Pulsation
- see RETINAL VENOUS PULSATION

Vernet's Syndrome
- see JUGULAR FORAMEN SYNDROME

Vertigo
Vertigo is an illusion of movement, a sense of rotation or of tilt, causing a feeling of imbalance or dysequilibrium. It is a subtype of "dizziness', to be distinguished from the light-headedness of general medical conditions (vasovagal attacks, presyncope, cardiac dysrhythmias). Vertigo is often triggered by head movement and there may be associated autonomic features (sweating, pallor, nausea, vomiting). Vertigo may be horizontal, vertical or rotatory.

Pathophysiologically, vertigo reflects an asymmetry of signaling anywhere in the central or peripheral vestibular pathways. Clinically it may be possible to draw a distinction between central and peripheral lesions: in the latter there may be concurrent hearing loss and tinnitus (reflecting vestibulocochlear (VIII) nerve involvement). Facial weakness (VII) and ipsilateral ataxia suggest a cerebellopontine angle lesion; diplopia, bulbar dysfunction and long tract signs are suggestive of a central pathology. Peripheral vertigo tends to compensate rapidly and completely with disappearance of nystagmus after a few days, whereas central lesions compensate slowly and nystagmus persists.

The clinical pattern of vertigo may gives clues as to underlying diagnosis:

Vertigo	**Peripheral**	**Central**
Acute Prolonged, spontaneous	Labyrinthitis Otomastoiditis Vestibular neur(on)itis Labyrinthine concussion Isolated labyrinthine infarct Vestibular nerve section Drug-induced	Brainstem/ cerebellum hemorrhage/ infarct/ demyelination
Recurrent, episodic	Ménière's disease (endolymphatic hydrops) Autoimmune inner ear disease (isolated, systemic) Perilymph fistula Migraine (rare) Epilepsy (rare)	Vertebrobasilar ischemia (with associated features)
Positional	Benign paroxysmal positional vertigo (BPPV)	4th ventricle lesions: multiple sclerosis Chiari malformation

(contd.)

(contd.)

Vertigo	Peripheral	Central
		Brainstem/ cerebellar tumors Spinocerebellar atrophy
Chronic	Vestibular decompensation/failure	Neurological disorder Psychogenic

All patients with vertigo should have a Hallpike maneuver performed during the examination.

Specific treatments are available for certain of these conditions. A brief course of a vestibular sedative (cinnarizine, Serc) is appropriate in the acute phase, but exercises to "rehabilitate" the semicircular canals should be begun as soon as possible in peripheral causes. In BPPV, most patients respond to the Epley maneuver to reposition the otoconia which are thought to cause the condition (canalolithiasis). Brandt-Daroff exercises are an alternative. Cawthorne-Cooksey exercises are helpful in vestibular decompensation or failure.

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Cross References

Ataxia; Caloric testing; Facial paresis; Hallpike maneuver, Hallpike test; Hennebert's sign; Illusion; Nystagmus; Vestibulo-ocular reflexes

Vestibulo-Ocular Reflexes

The vestibulo-ocular reflexes (VOR) are a physiological mechanism which generates eye rotations that compensate for head movements, especially during locomotion, so stabilizing the retinal image on the fovea. VORs depend upon the integrity of the connections between the semicircular canals of the vestibular system (afferent limb of reflex arc) and oculomotor nuclei in the brainstem (efferent limb). Loss of vestibular function, as in acute bilateral vestibular failure, causes gaze instability due to loss of VORs, causing the symptom of oscillopsia (*q.v.*) when the head moves. As well as vestibular input, compensatory eye rotations may also be generated in response to visual information (pursuit-optokinetic eye movements) and neck proprioceptive information; anticipatory eye movements may also help stabilize the retinal image.

VORs are also useful in assessing whether ophthalmoplegia results from a supranuclear or infranuclear disorder, since in the former the restriction of eye movement may be overcome, at least in the early stages, by the intact VOR (*e.g.*, the supranuclear gaze palsy in the vertical plane in progressive supranuclear palsy).

VORs are difficult to assess in conscious patients because of concurrent pursuit-optokinetic eye movements, and because rotation of the head through large angles in conscious patients leads to interruption of VORs by vestibular nystagmus in the opposite direction (optokinetic nystagmus). The head impulse test (*q.v.*) may be used to test VORs in conscious patients, for example those with vertigo in whom vestibular failure is suspected. VOR may also be assessed using a slow (0.5-1.0 Hz) doll's head maneuver while directly observing the eyes ("catch up" saccades may be seen in the absence of VOR), measuring visual acuity (dynamic visual acuity, or illegible E test; dropping two to three lines on visual acuity with head movement *vs.* normal if VOR impaired), and ophthalmoscopy (optic disc moves with head if VOR abnormal).

In unconscious patients, slow phase of the VORs may be tested by rotating the head and looking for contraversive conjugate eye movements (oculocephalic responses, doll's head eye movements) or by caloric testing. VORs are lost in brainstem death.

Another important element of VOR assessment is suppression or cancellation of VOR by the pursuit system during combined head and eye tracking. VOR suppression may be tested by asking the patient to fixate on their thumbs with arms held outstretched while rotating at the trunk or sitting in a swivel chair. VOR suppression can also be assessed during caloric testing: when the nystagmus ceases with fixation, removal of the fixation point (*e.g.*, with Frenzel's glasses) will lead to recurrence of nystagmus in normals but not in those with reduced or absent VOR suppression. VOR suppression is impaired (presence of nystagmus even with slow head movements) in cerebellar and brainstem disease.

References

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Leigh RJ, Brandt T. A reevaluation of the vestibulo-ocular reflex: new ideas of its purpose, properties, neural substrate, and disorders. *Neurology* 1993; **43**: 1288-1295

Cross References

Caloric testing; Coma; Doll's eye maneuver, Doll's head maneuver; Hallpike maneuver, Hallpike test; Head impulse test; Ocular tilt reaction; Oculocephalic response; Oscillopsia; Supranuclear gaze palsy; Vertigo

Vibration

Vibratory sensibility (pallesthesia) represents a temporal modulation of tactile sense. On this ground, some would argue that the elevation of vibration to a "sensory modality" is not justified. Vibratory sensibility is easily tested using a tuning fork (128 Hz). This assesses the integrity of rapidly adapting mechanoreceptors (Pacinian corpuscles) and their

peripheral and central connections; the former consist of large afferent fibers, the latter of ascending projections in both the dorsal and lateral columns. The classification of both vibration and proprioception as "posterior column signs," sharing spinal cord and brainstem pathways, is common in neurological parlance (and textbooks) but questioned by some. Instances of dissociation of vibratory sensibility and proprioception are well recognized, for instance the former is usually more impaired with intramedullary myelopathies.

Decrease in sensitivity of vibratory perception (increased perceptual threshold) is the most prominent age-related finding on sensory examination, thought to reflect distal degeneration of sensory axons.

References

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Cross References

Age-related signs; Myelopathy; Proprioception; Two-point discrimination

Visual Agnosia

Visual agnosia is a disorder of visual object recognition. The term derives from Freud (1891), but it was Lissauer (1890), speaking of *seelenblindheit* (psychic blindness), who suggested the categorization into two types which continues to be used:

- *Apperceptive visual agnosia*:
  A defect of higher order visual perception leading to impaired shape recognition, manifested as difficulty copying shapes or matching shapes, despite preserved primary visual capacities, including visual acuity and fields (adequate to achieve recognition), brightness discrimination, color vision and motion perception (indeed motion may facilitate shape perception; see Riddoch's phenomenon). Reading is performed with great difficulty, with a "slavish" tracing of letters which is easily derailed by any irrelevant lines; such patients may appear blind.
- *Associative visual agnosia*:
  An impairment of visual object recognition thought not to be due to a perceptual deficit, since copying shapes of unrecognized objects is good. The scope of this impairment may vary, some patients being limited to a failure to recognize faces (prosopagnosia) or visually presented words (pure alexia, pure word blindness).

Visually agnosic patients can recognize objects presented to other sensory modalities. Clinically, apperceptive visual agnosia lies between cortical blindness and associative visual agnosia.

Apperceptive visual agnosia results from diffuse posterior brain damage; associative visual agnosia has been reported with lesions in a variety of locations, usually ventral temporal and occipital regions,

usually bilateral but occasionally unilateral. Pathological causes include cerebrovascular disease, tumor, degenerative dementia (visual agnosia may on occasion be the presenting feature of Alzheimer's disease, the so-called visual variant, or posterior cortical atrophy), and carbon monoxide poisoning.

A related syndrome which has on occasion been labeled as apperceptive visual agnosia is simultanagnosia (*q.v.*), particularly the dorsal variant in which there is inability to recognize more than one object at a time. Associative visual agnosia has sometimes been confused with optic aphasia (*q.v.*).

References

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Riddoch MJ, Humphreys GW. Visual agnosia. *Neurologic Clinics* 2003; **21**: 501-520

Cross References

Agnosia; Alexia; Cortical blindness; Optic aphasia; Prosopagnosia; Riddoch's phenomenon; Simultanagnosia; Visual form agnosia

Visual Disorientation

Visual disorientation refers to the inability to perceive more than a fragment of the visual field at any one time; it is sometimes characterized as a shifting fragment or island of clear vision. There may be difficulty fixating static visual stimuli and impaired visual pursuit eye movements.

Visual disorientation may be demonstrated by sitting directly opposite the patient and asking them, while looking at the bridge of the examiner's nose, to reach for the examiner's hand held up in the peripheral field of vision. Once contact is made with the hand, the examiner holds up the other hand in a different part of the field of vision. Individuals with visual disorientation will find it hard to see the hand and will grope for it, sometimes mistakenly grasping the examiner's clothing ("tie sign") or face.

Visual disorientation is secondary to, and an inevitable consequence of, the attentional disorder of dorsal simultanagnosia (*q.v.*), in which the inability to attend two separate loci leads to impaired localization. It may be a feature of Alzheimer's disease; indeed, sometimes it may be the presenting feature, but there are usually signs of more generalized cognitive problems (*e.g.*, impairment of episodic memory).

References

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

Cross References

Simultanagnosia; Visual agnosia

Visual Extinction

Visual extinction is the failure to respond to a novel or meaningful visual stimulus on one side when a homologous stimulus is given simultaneously to the contralateral side (*i.e.*, double simultaneous stimulation), despite the ability to perceive each stimulus when presented singly.

Cross References
Extinction; Neglect

Visual Field Defects

Visual fields may be mapped clinically by confrontation testing. The exact pattern of visual field loss may have localizing value due to the retinotopic arrangement of fibers in the visual pathways: any unilateral area of restricted loss implies a pre-chiasmatic lesion (choroid, retina, optic nerve), although lesions of the anterior calcarine cortex can produce a contralateral monocular temporal crescent. Bilateral homonymous scotomata are post-chiasmal in origin; bilateral heteronymous scotomata may be seen with chiasmal lesions.

Topographically, typical visual field defects are:

Retina: monocular visual loss, altitudinal field defects; central or centrocecal scotoma, ring scotoma
Optic nerve: central or centrocecal scotoma; junctional scotoma of Traquair
Optic chiasm: bitemporal hemianopia; junctional scotoma
Optic tract: homonymous hemianopia, usually incongruous
Lateral geniculate nucleus: homonymous hemianopia, usually incongruous
Optic radiations: homonymous hemianopia, usually congruous; quadrantanopia
Visual cortex: homonymous hemianopia, usually congruous; quadrantanopia; cortical blindness

References

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Trobe JD, Acosta PC, Krischer JP, Trick GL. Confrontation visual field techniques in detection of anterior visual pathway lesions. *Annals of Neurology* 1981; **10**: 28-34

Cross References
Altitudinal field defect; Hemianopia; Junctional scotoma, Junctional scotoma of traquair; Macula sparing, Macula splitting; Quadrantanopia; Scotoma

Visual Form Agnosia

This name has been given to an unusual and highly selective visual perceptual deficit, characterized by loss of the ability to identify shape and form, although color and surface detail can be appreciated, but with striking preservation of visuomotor control (*i.e.*, a pattern of deficits inverse to those seen in optic ataxia). This reflects selective damage to the ventral ("what") stream of visual processing in the lateral occipital area, while the dorsal ("where") stream remains intact, yet its workings are not available to consciousness.

References

Goodale MA, Milner AD. *Sight unseen. An exploration of conscious and unconscious vision*. Oxford, OUP, 2003

Cross References

Agnosia; Optic ataxia; Visual agnosia

Visual Grasp Reflex

- see SACCADES

Visuopalpebral Reflex

- see BLINK REFLEX

Vocal Tremor, Voice Tremor

Vocal or voice tremor is a shaking, quivering, or quavering of the voice. It may be heard in:

Essential tremor
Cerebellar disorders
Spasmodic dysphonia/laryngeal dystonia
Parkinson's disease
Motor neurone disease.

The pathophysiology is uncertain but may relate to rhythmic contractions of the cricothyroid and rectus abdominis muscles.

Cross References

Dysphonia; Tremor

Von Graefe's Sign

Von Graefe's sign is the retarded descent of the upper eyelid during movement of the eye from the primary position to downgaze; the lid "follows" the eye. This may be termed "lid lag," although some authorities reserve this term for a static situation in which the lid is higher than the globe on downgaze. Von Graefe's sign is seen in thyroid ophthalmopathy.

Cross References

Lid lag; Pseudo-von Graefe's sign

Vorbereiden

- see GANSER PHENOMENON

VOR Suppression

- see VESTIBULO-OCULAR REFLEXES

Vulpian's Sign

- see PREVOST'S SIGN

W

Waddling Gait
Weakness of the proximal leg and hip girdle muscles, most often of myopathic origin, impairs the stability of the pelvis on the trunk during walking, leading to exaggerated rotation with each step, an appearance likened to the waddling of a duck. In addition, the hips may be slightly flexed and lumbar lordosis exaggerated. Neurogenic causes include spinal muscular atrophy and Guillain-Barré syndrome.
Cross References
Myopathy

"Waiter's Tip" Posture
Lesions of the upper trunk of the brachial plexus (Erb-Duchenne type) produce weakness and sensory loss in the C5 and C6 distribution, typically with the arm hanging at the side, internally rotated at the shoulder with the elbow extended and the forearm pronated: the "waiter's tip" posture, also sometimes known as the "porter's tip" or "policeman's tip."
Cross References
Plexopathy; Radiculopathy

Wallenberg's Syndrome
- see LATERAL MEDULLARY SYNDROME

Wall-Eyed
- see EXOTROPIA; INTERNUCLEAR OPHTHALMOGPLEGIA

Warm-Up Phenomenon
Easing of muscle stiffness with repeated contraction, the warm-up phenomenon, is reported by many patients with myotonia congenita (Thomsen's disease, Becker's disease), in contrast to the situation in paramyotonia.
Cross References
Myotonia; Paramyotonia

Wartenberg's Sign
- see CORNEOMANDIBULAR REFLEX

Wartenberg's Swing Test
Wartenberg's swing test is used to assess limb and trunk rigidity (*cf.* Wartenberg's pendulum test, used to measure spasticity, *q.v.*). With the patient standing, the examiner holds the shoulders and gently shakes backward and forwards, the two sides out of phase. Normally the passive arm swing induced by this movement will be out of phase with the

trunk movements, but in rigidity the limbs and trunk tend to move *en bloc*. Passive swinging of the wrist or elbow joint may also be performed to assess rigidity.

Cross References

Parkinsonism; Rigidity; "Rolex" sign; Spasticity

Wasting

Wasting refers to a thinning of the musculature, also known as atrophy or, if of neurogenic origin, amyotrophy.

Wasting may be a consequence of disorders of:

- Muscle (myopathies, dystrophies)
- Peripheral nerve (more so in axonal than demyelinating peripheral neuropathies)
- Anterior horn cells (*e.g.*, motor neurone disease).

Wasting may occur in chronic upper motor neurone syndromes (*e.g.*, chronic hemiplegia) but is not as evident as in lower motor neurone syndromes where wasting may appear subacutely (over a few weeks).

Wasting may also be seen in general medical disorders associated with a profound catabolic state, *e.g.*, cancer cachexia, uncontrolled heart failure, liver cirrhosis, renal failure.

Cross References

Amyotrophy; Atrophy; Lower motor neurone (LMN) syndrome; Upper motor neurone (UMN) syndrome

Weakness

Weakness is an objective loss of muscle strength. This is conveniently quantified or rated using the MRC grading system:

5 = normal power
4 = active movement against gravity and resistance
3 = active movement against gravity
2 = active movement with gravity eliminated
1 = flicker or trace of contraction
0 = no contraction (paralysis).

However, this is not a linear scale; grade 4 often becomes subdivided into 4–, 4, and 4+ (or even 5–) according to the increasing degree of resistance which the examiner must apply to overcome activity. It is also important to assess what effort the patient is making to comply with the testing; "apparent weakness" or "pseudoparesis" may be shorthand for lack of patient effort. Sudden "giving way" of muscle contraction may be an indicator of this. Nonuniform resistance may also be due to pain (algesic pseudoparesis). Testing records only the best forced maximal contraction, and should not develop into an unseemly trial of strength between patient and examiner. Accepting all these difficulties, it should be acknowledged that the grading of weakness, like all clinical observations, is subject to some degree of observer bias.

The pattern of muscle weakness may suggest its anatomical origin. So-called "pyramidal weakness" (*i.e.*, affecting upper limb extensors more than flexors, and lower limb flexors more than extensors), suggests an upper motor neurone lesion (corticospinal pathways). However, there is no evidence that pure lesions of the pyramidal tracts produce this picture: pyramidotomy in the monkey results in a deficit in fine finger movements, but without weakness. Moreover, a similar pattern of weakness may be observed in lower motor neurone disorders, such as Guillain-Barré syndrome. Coexistent wasting suggests muscle weakness is of lower motor neurone origin, especially if acute, although wasting may occur in long-standing upper motor neurone lesions. Weakness with minimal or no muscle wasting may be nonorganic, but may be seen in conditions, such as multifocal motor neuropathy with conduction block.

References

Aids to the Examination of the Peripheral Nervous System. London: HMSO, 1976

Cross References

Collapsing weakness; Hyperreflexia; Lower motor neurone (LMN) syndrome; Upper motor neurone (UMN) syndrome; Wasting

Weber's Test

Weber's test is one of the tuning fork tests, which may be used to confirm a conductive component in unilateral or asymmetric hearing loss. The vibrating tuning fork is put on the middle of the forehead and the patient asked in which ear it is heard; this depends entirely upon bone conduction (BC). Hence the sound localizes to the side of a conductive hearing loss (where bone conduction is greater than air conduction, BC > AC), and away from the side of a sensorineural hearing loss.

References

Miyamoto RT, Wynne MK. Approach to the patient with hearing loss. In: Biller J (ed.). *Practical neurology* (2nd edition). Philadelphia: Lippincott Williams & Wilkins, 2002: 206-226

Cross References

Rinne's test

Wernicke's Aphasia

Wernicke's aphasia is the classical "receptive aphasia," in distinction to the "expressive aphasia" of Broca, although this classification is problematic since there are concurrent "expressive" problems in Wernicke's aphasia.

Considering each of the features suggested for the clinical classification of aphasias (see Aphasia), Wernicke's aphasia is characterized by:

- *Fluency*: fluent speech with phonemic and semantic paraphasias and paragrammatism (inappropriate use of syntax); "empty speech" with few verbs and nouns; prosody usually preserved; at worst, flowing speech (logorrhea) devoid of semantic meaning (jargon aphasia, semantic aphasia); automatic speech is often bet-

ter preserved than spontaneous, *e.g*., counting, days of week, overlearned phrases ("I'm fine").

- *Comprehension*: impaired auditory comprehension (*sine qua non*; "word deafness"); impaired reading comprehension probably also required (not specifically discussed by Wernicke).
- *Repetition*: impaired.
- *Naming*: severely impaired (anomia) and not aided by cueing (*cf*. Broca's aphasia).
- *Reading*: usually impaired, with numerous paralexic errors, and impaired reading comprehension (*cf*. pure word deafness).
- *Writing*: similarly affected.

There may be associated anxiety, with or without agitation and paranoia, and concurrent auditory agnosia.

The neuroanatomical substrate of Wernicke's aphasia has been a subject of debate. Wernicke placed it in the posterior two-thirds of the superior temporal gyrus and planum temporale (Brodmann area 22), but more recent neuroradiological studies (structural and functional imaging) suggest that this area may be more associated with the generation of paraphasia whereas more ventral areas of temporal lobe and angular gyrus (Brodmann areas 37, 39 and 40) may be associated with disturbance of comprehension. A correlation exists between the size of the lesion and the extent of the aphasia. A similar clinical picture may occur with infarcts of the head of the left caudate nucleus and left thalamic nuclei.

The differential diagnosis of Wernicke's aphasia includes delirium and schizophasia.

References

Binder JR. Wernicke aphasia: a disorder of central language processing. . In: D'Esposito M (ed.). *Neurological foundations of cognitive neuroscience*. Cambridge: MIT Press, 2003: 175-238

Wise RJS, Scott SK, Blank SC, Mummery CJ, Murphy K, Warburton EA. Separate neural subsystems within "Wernicke's area." *Brain* 2001; **124**: 83-95

Cross References

Agnosia; Agraphia; Alexia; Anomia; Aphasia; Broca's aphasia; Jargon aphasia; Logorrhea; Paraphasia; Pure word deafness; Schizophasia; Transcortical aphasias

"Wheelchair Sign"

The so-called "wheelchair sign" describes patients with parkinsonism who take to using a wheelchair early in the course of their disease, usually because of falls. Early falls are a feature of progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome) rather than idiopathic Parkinson's disease.

Cross References

Parkinsonism; "Rocket sign"

Wing-Beating Tremor

Wing-beating tremor is absent at rest but develops when the arms are extended, hence a postural tremor. It is said to be typical of Wilson's disease.

Cross References

Tremor

Winging of the Scapula

Winging of the scapula, or scapula alata, is a failure to hold the medial border of the scapula against the rib cage when pushing forward with the hands. It is most easily observed by asking the patient to push or press against a wall or the examiner's hand while observing the scapula which lifts away from the posterior chest wall.

Winging of the scapula may be a consequence of weakness of the serratus anterior muscle, usually due to a neuropathy of the long thoracic nerve of Bell, but sometimes as a consequence of brachial plexus injury or cervical root (C7) injury. It may also be of myopathic origin, as in facioscapulohumeral dystrophy.

Weakness of trapezius, particularly the middle trapezius muscle, may also cause winging of the upper part of the scapula, more prominent on abduction of the arm, when the superior angle of the scapula moves farther from the midline. Hence spinal accessory (XI) nerve palsy enters the differential diagnosis.

Witzelsucht

Witzelsucht refers to excessive and inappropriate facetiousness or jocularity, a term coined in the 1890's for one of the personality changes observed following frontal (especially orbitofrontal) lobe injury. This phenomenon may overlap with those described as moria or emotional lability.

Cross References

Emotionalism, Emotional lability; Frontal lobe syndromes; Moria

Woltman's Sign

Woltman's sign denotes slow-relaxing, or "hung-up," tendon reflexes. These are most commonly seen in the context of untreated hypothyroidism, but have also been recorded in other situations, including treatment with β-blockers, diabetes mellitus, and complete heart block. The phenomenon is sometimes labeled pseudomyotonia because of its superficial resemblance to the slow muscle relaxation of myotonia, but electrophysiological testing does not show myotonic discharges.

Chorea may result in apparently "hung-up" reflexes, perhaps due to a choreiform jerk after muscle relaxation.

The mechanisms underlying Woltman's sign are uncertain: changes in basal metabolic rate and in muscle fibre types (selective loss of fast twitch fibers) have been suggested.

References

Larner AJ. Normalisation of slow-relaxing tendon reflexes (Woltman's sign) after cardiac pacing for complete heart block. *British Journal of Clinical Practice* 1995; **49**: 331-332

Cross References
Chorea, Choreo athetusis; Myotonia; Pseudomyotonia

"Wrestler's Sign"

This name has been given to the excessive effort in irrelevant muscle groups accompanied by prominent verbal grunts in patients with apparent ("functional") weakness. It may coexist with intermittent voluntary effort, collapsing weakness, co-contraction of agonist and antagonist muscles and inconsistency in clinical examination (*e.g.*, inability to lift leg from couch when recumbent, but able to stand and walk).
Cross References
Collapsing weakness

Wrist Drop

Wrist drop describes a hand hanging in flexion due to weakness of wrist extension. This results from radial nerve palsy, either in the axilla or spiral groove of the humerus ("Saturday night palsy"). Distal lesions affecting branches of the posterior interosseous branch of the radial nerve may produce more circumscribed deformity, such as weak extension of metacarpophalangeal joints ("finger drop," "thumb drop").

Writer's Cramp

Writer's cramp, or graphospasm, is a focal dystonia of the hand in which dystonic posturing is induced specifically by writing: this is the most common task-specific dystonia. The involuntary movements may eventually make it impossible to write with the dominant hand. Learning to write with the opposite hand may only be a partial solution, since it too may become affected. Muscle fatigue may make writing more legible. Writer's cramp is much commoner than primary writing tremor as a cause of writing difficulty.

Botulinum toxin injections may be of benefit if relatively few muscles are affected. There may be an associated carpal tunnel syndrome.
References
Sheehy MP, Marsden CD. Writer's cramp – a focal dystonia. *Brain* 1982; **105**: 461-480
Cross References
Dystonia; Fatigue; Tremor

Wrong-Way Eyes

- see PREVOST'S SIGN

Wry Neck

- see TORTICOLLIS

X

Xanthopsia

Xanthopsia is a visual disturbance characterized by excessive perception of yellow colors (literally "yellow vision"). It may be associated with use of various drugs including digoxin (especially if levels are toxic), thiazides (especially chlorothiazide), sulphonamides, and barbiturates. The mechanism is uncertain, but one possibility is that this is a partial form of achromatopsia, affecting one color more than others.

It has been suggested that the artist Vincent van Gogh (1853-1890) may have suffered from xanthopsia as a consequence of digitalis toxicity, accounting for the bright yellows in many of his later canvases.

References

Critchley M. Acquired anomalies of color perception of central origin. *Brain* 1965; **88**: 711-724

Cross References

Achromatopsia

Xerophthalmia, Xerostomia

Xerophthalmia, dryness of the eyes, and xerostomia, dryness of the mouth, due to impaired secretion from the lacrimal glands and the salivary glands respectively, often occur together. This may reflect autonomic dysfunction, as for example in Lambert Eaton myasthenic syndrome, or be due to autoimmune disorders, such as Sjögren's syndrome.

Cross References

Facilitation; Orthostatic hypotension

Y

Yawning

Yawning is an arousal reflex thought to be generated in the brainstem reticular formation to counteract brain hypoxia; it may precede vasovagal syncope. Excessive or pathological yawning (chasm) is compulsive, repetitive yawning not triggered by physiological stimuli, such as fatigue or boredom. Known associations of yawning include:

Encephalitis
Seizures
Multiple sclerosis
Tumors of the 4th ventricle, frontal lobes
Electroconvulsive therapy
Postthalamotomy
Drugs (valproate, imipramine)
Neuroleptic withdrawal
Parkinson's disease, progressive supranuclear palsy, restless legs syndrome, pseudobulbar palsy of motor neurone disease

Although the mechanisms are uncertain, yawning may represent a disturbance of dopaminergic transmission. Levodopa may help.

References

Leonhardt M, Abele M, Klockgether T, Dichgans J, Weller M. Pathological yawning (chasm) associated with periodic leg movements in sleep: cure by levodopa. *Journal of Neurology* 1999; **246**: 621-622

Williams DR. The yawning reflex: an upper motor neuron sign in amyotrophic lateral sclerosis. *Neurology* 2000; **55**: 1592-1593

Cross References

Parkinsonism; Sighing

Yo-yo-ing

Yo-yo-ing is a form of dyskinesia experienced by patients with idiopathic Parkinson's disease who have been treated for several years with levodopa preparations, in which there are sudden and unpredictable swings between hypokinesia/akinesia ("off" state; freezing) and severe hyperkinesia ("on" state), sometimes known as the "on-off phenomenon." Yo-yo-ing is difficult to treat: approaches include dose fractionation, improved drug absorption, or use of dopaminergic agonists with concurrent reduction in levodopa dosage.

Cross References

Akinesia; Dyskinesia; Hypokinesia

Z

Zooagnosia

The term zooagnosia has been used to describe a difficulty in recognizing animal faces. This may be observed as a component of prosopagnosia. In one case, this deficit seemed to persist despite improvement in human face recognition, suggesting the possibility of separate systems for animal and human face recognition; however, the evidence is not compelling.

References

Assal G, Favre C, Anderes J. Non recognition of familiar animals by a farmer: zooagnosia or prosopagnosia for animals. *Revue Neurologique (Paris)* 1984; **140**: 580-584

Cross References

Agnosia; Prosopagnosia