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

Cross References
Lhermitte’s sign; Phosphenes

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
Urgency

- 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

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
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).
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.

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

Cross References
Automatic writing behavior; Echolalia; Echopraxia; Frontal lobe syndromes; Hypermetamorphosis; Imitation behavior; Primitive reflexes