III./12.2. Disorders of consciousness

In neurology, clear consciousness is defined as the brain’s ability to react (arousal or alertness) and the adequate use of the contents of memory and to think (awareness), which allow the adaptation of the individual to the environment and are the basis of cognition. Awareness depends on alertness.

From the clinical point of view, disorders of consciousness are divided into two groups: disorder of arousal (reduced arousal, sleep-like state) and disorder of awareness (inadequate thinking and behavior).

Arousal is a prerequisite of awareness. It is defined as the ability to wake up from sleep when stimuli are applied, leading to eye opening and behavioral changes. Awakening is indicated by desynchronization on the EEG (decreased amplitude and faster background activity). The anatomical basis of arousal is the intact functioning of the ascending reticular activating system (ARAS). ARAS is composed of the pontomesencephalic reticular formation, the monoaminergic networks of the diencephalon, and the intralaminar and medial nuclei of the thalamus. The centers of these systems according to the type of neurotransmitter involved are the following: cholinergic system- nucleus basalis Meynerti, noradrenergic- locus ceruleus, serotoninergic- raphe nuclei, histaminergic- posterior part of hypothalamus.

In a state of reduced arousal, the patient appears to be sleeping and doesn’t react to awakening stimuli in a normal way. The mildest form of reduced arousal is somnolence, where verbal stimuli are enough to generate an awakening reaction, but without stimuli the patient falls asleep again. In a more severe form, which is called stupor, verbal stimuli are hardly effective in awakening the patient, but patients may open their eyes to painful stimuli. Withdrawal reactions to painful stimuli are slow and usually inappropriate. Unconscious patients (coma) cannot be awakened, and their posture is usually abnormal (decorticate or decerebrate posture). The term coma I is used if brainstem reflexes are still preserved, and coma II if they are lost.

In disorders of awareness, the central nervous system’s global function is impaired. Signs of focal cognitive deficit (e.g. alexia, aacalculia) are not a part of this condition. In these states, the patient’s arousal is normal, but the content of memory is disturbed, inaccessible or their use is inappropriate.

Clinical forms include:

- persistent vegetative state
- akinetic mutism
- confusion and delirium

In persistent vegetative state (synonyms: decorticate state, apallic syndrome; permanent vegetative state is used when it lasts longer than 30 days), the rostral brainstem remains intact, so the patient’s thermoregulation (hypothalamus), the sleep-wake cycle, cardiorespiratory and other visceral functions, and the endocrine system are functioning. Patients seem to be awake, their eyes are open, scanning eye movements and fixation are absent, attention
cannot be aroused, but eye movement reflexes are elicitable (e.g. vestibuloocular reflex). Causes of a vegetative state include an extensive functional or structural impairment of the cerebral cortex (e.g. global cerebral ischemia caused by circulatory arrest, severe metabolic conditions like hypoglycemia, renal- and hepatic failure, postconvulsive state, Wernicke’s encephalopathy, final stages of cortical dementias), extensive white matter damage (diffuse axonal injury after head trauma), and bilateral damage to the thalamus.

In akinetic mutism, the patient is awake, but mute and lies without moving. Mutism is not caused by aphasia, as signs indicating preserved comprehension are usually present, and paralysis is excluded by normal withdrawal reactions to painful stimuli. In the majority of cases, frontal release signs are seen (sucking reflex, bulldog reflex, palmar grasp reflex). This frequently unrecognized syndrome is caused by the bilateral interruption of connections between the supplementary motor area, the cingular region and the midline nuclei of the thalamus. Akinetic mutism may be caused by jet bleeding (rupture of an anterior communicating artery aneurysm), frontobasal contusion, bilateral ischemia in the territory of the anterior cerebral artery (Willis-circle variants), subfalcial herniation, occlusive hydrocephalus, butterfly tumors growing across the corpus callosum into the prefrontal lobes, and tumors of the third ventricle.

In confusion, attention and thinking are impaired, the patient is slow and vague, thinking is incoherent, but there are no signs suggestive of delusions. A typical example of confusion is seen after an epileptic seizure (postconvulsive state or tenebrosity).

In the earlier definition of delirium, abnormal perception (illusions and hallucinations) was considered to be the main psychopathological symptom, and the patient’s intense restlessness and abnormal behavior was explained by this. According to the current definition, the disorder of attention and awareness is the core symptom of delirium, and the disorder of perception is no longer considered as a diagnostic criterion. According to this modified definition, confusion is also a form of delirium, but it is useful in clinical practice to distinguish between the two states on the basis whether abnormal perception is present or not.

Therefore, a delirious patient cannot focus, change or fix attention, and due to the cognitive dysfunction she/he is disoriented and thinking is incoherent. Delirium usually evolves quickly (within a few hours or 24 hours at the most), and during its course diurnal fluctuation of intensity is characteristic. Symptoms deteriorate at night or in an environment lacking stimuli, and inversion of the sleep/wake cycle is also typical. Autonomic instability, mostly excitatory symptoms (sweating, tachycardia, unstable blood pressure), is frequently present. In addition to metabolic disorders and drug or alcohol related causes, delirium may be a complication of right hemispheric lesions (at the parieto-occipito-temporal junction).

Locked-in syndrome is not a disorder of consciousness, but it is frequently misdiagnosed by superficial examiners, since the patient is unable to communicate neither in verbal nor in a non-verbal way, because the voluntary innervation of the facial muscles and the extremities is lost. The site of the lesion is the base and the tegmentum of the pons on both sides, thus the corticospinal tract, the pontine horizontal gaze centers, and the facial nuclei are damaged. In most cases, locked-in syndrome is caused by the occlusion of the
basilar artery, but the rapid correction of hyponatremia leading to central pontine myelinolysis may also become manifest in this syndrome. As ARAS is not injured, the patient’s sleep-wake cycle is intact, they perceive visual and acoustic stimuli, and some form of communication may be achieved by blinking, using “yes or no” questions.