Symptoms and Syndromes

15 Hepatic encephalopathy

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It seems that even **Hippocrates** may have recognized a relationship between liver disease and brain disorder when he noted: *"depravedness of the brain arises from phlegm and bile"*. • One can also quote **Shakespeare**, who wrote in Twelfth Night (1605): *"I am a great eater of beaf but I believe that does some harm to my wit."* • Such a form of **"meat intoxication"** was first described by N.V. Eck (1877) as being the cause of a subsequent neurologic syndrome in dogs with a portacaval shunt, which had been fed on a diet of meat.

► **Cerebral symptoms in the wake of liver diseases** were already described by F.Th. Frerichs (1861) and H.I. Quincke (1899) as well as over the following decades by E. Pollak (1927), H.-J. Scherer (1933), G. Zillig (1947), V. Gaustad (1949), R.D. Adams et al. (1949), J.M. Walshe (1951), S. Sherlock et al. (1954), E.A. Davidson et al. (1956) (23), F. Erbsloeh (1958, 1974), G.A. Martini (1975) (63), and others.

Such **terms** as "hepatargy", "leucoencephalopathy", "shunt encephalomyelopathy" and "encephalomyelopathy" were put forward to classify these central nervous disorders. • The term **portosystemic encephalopathy** was introduced by S. Sherlock (1954). Changes in the central nervous system which occur after a long course of disease were also termed "chronic hepatoportal encephalopathy". In line with neuropsychiatric definitions, preference is given to the term "hepatic (portosystemic) encephalopathy".

1 Definition

Encephalopathy is defined as a pathological non-inflammatory brain disease resulting from heterogeneous pathological effects, which involve various neurological and/or psychic symptoms. In itself, this term says nothing about the aetiopathogenesis, nor about the respective regions of the brain affected.

Hepatic encephalopathy (HE) is defined as the totality of all cerebral dysfunctions which can occur during the course of serious − acute or chronic − liver disease. The neurological and mental symptoms, which as a rule are potentially reversible, can be witnessed to varying degrees of intensity and in different combinations, so that it is possible to subdivide hepatic encephalopathy into several well-defined grades of severity or distinct stages. Clinical symptomatology ranges from moderate neuropsychiatric disorders through to coma.

1.1 Causes of encephalopathy

Encephalopathy can be triggered by **(*1.*) degenerative causes** (cerebral sclerosis, arterial hypertension, diabetes, etc.), **(*2.*) hypoxaemia** (chronic cardiac insufficiency, constrictive pericarditis (2), respiratory insufficiency, chronic anaemia, etc.), **(*3.*) metabolic causes** (enzymopathies, endocrine disorders, hypokalaemia, hyponatraemia, acidosis, alkalosis, exsiccosis, hypercapnia, paraproteinosis, etc.), **(*4.*) traumatization** (Friedmann's vasomotor syndrome, boxer's encephalopathy, etc.), **(*5.*) toxic causes** (alcohol, lead, organic solvents, bismuth, mussels contaminated with domoic acid, burn-related toxins, bilirubin [= kernicterus], vidarabine, uraemia, infectious diseases, liver insufficiency, etc.), and **(*6.*) cerebral causes** (intracranial space-occupying lesions, alcohol withdrawal syndrome, manic depression, schizophrenia, etc.).

All in all, more than 50 possible kinds of damage are known to cause encephalopathy. This large number of aetiological possibilities has to be considered when setting up the differential diagnosis, especially since symptomatology, therapy and prognosis are always determined by the respective underlying cause.

1.2 Impaired consciousness

Consciousness is defined as the totality of mental and emotional processes perceived as current events in combination with an awareness of the subjectivity of experience. • **Quantitatively impaired consciousness** is the collective term for disturbances in awareness (= restricted vigilance). This condition is characterized by a limited readiness to show interest and concern, restricted attentiveness and impaired elements of consciousness. The activity of the ascending reticular system of the reticular formation is compromised, a condition which is generally accompanied by changes in the electroencephalogram (EEG). Moreover, disorders or loss of function are displayed in the area of the cerebral hemispheres and/or their afferent pathways, which are responsible for the subjective sensation of consciousness. • Differentiation is made between **four types** of clouded consciousness (W. Hacke, 1986):

1. Disorientation: A slight degree of impaired of consciousness without clouding, yet with impediment to and deceleration of mental capacity and memory, compromised perception and reactivity (transitional psychosis).

2. Somnolence: Disorientation with additional, abnormal sleepiness, though arousal is still possible, and there can be memory gaps. • **Lethargy** is deemed to be the tendency to sleep without interruption, with greatly reduced mental/emotional responsiveness and absence of arousal reaction to normal stimuli.

3. Sopor: A condition resembling deep sleep with short-term attempts at orientation when being addressed, orderly movements of defence in reaction to pain − yet with no capability to react spontaneously.

4. Coma: Impaired consciousness of the most serious nature with long periods of profound unconsciousness and unresponsiveness to any form of address through to the absence of pain reaction and reflexes. • The term **stupor** is used to denote a disease with total absence of physical and/or mental activity due to loss of drive, yet with full consciousness. Amimia, lack of spontaneity, unresponsiveness to external stimuli or attempts at establishing contact, and mutism are witnessed. • Stupor is also deemed to be unconsciousness of cerebro-organic origin, as for example in a serious transitional syndrome; it may be regarded as a physically substantiated, non-specific psychosis.