THE APALLIC SYNDROME
SYMTOMATOLOGY AND PROGNOSIS

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The apallic syndrome is a diffuse progressive disorder of the cerebrum, affecting the cortex and white matter It may appear in patients with a primary cerebral disorder arising from intrinsic diseases of neurons and glia (PLUM and POSNER 1972) . Examples are senile or presenile brain atrophy, diffuse brain sclerosis, subacute sclerotic leukencephalitis, the severest from of multiple sclerosis and the MACHIAFAVABIGNAMI syndrome. The apallic syndrome occuring in a progressive process of the cerebrum is an irreparable end-state.

On the other hand the apallic syndrome appears in patients with an acute or subacute secondary disorder of the brain. The disorder may affect the total cerebrum or may interrupt the ascending and descending pathways by a local lesion in the midbrain region. In many disorders, a combination of both is observed. Examples are cerebral hypoxia of different origin edema caused by allergic reaction diffuse encephalitis, gas embolism, acute or subacute metabolic disorders, exogenous intoxications, traumtic brain injuries as well as tumors or circulatory disturbances in the midbrain region, subarachnoid hemorrhage with tamponde of the basal cisterns. In the majority of events described in this group the diffuse brain edema is the cause of supratentorial brain shift and downward displacement of the hemispheres and the basal nuclei followed by tentorial herniation and lesion of the oral brain stem.

According to the clinical course of primary or secondary brain disorders, the apallic syndrome develops in different sequences. In patients with acute traumatic brain injuryor with acute metabolic brain disorder, phases of midbrain syndrome are followed by a tran-

sition stage to the apallic syndrome. In subacute progressive metabolic brain disorders the disintegration shows signs of the phases which are usually seen in the remission phase after acute brain damage. Table 1 demonstrates the different stages in the derelop ment and the remission of an apallic syndrome.

There are three characteristic clinical pictures of the transition stage to the apallic syndrome in cases with acute brain stem involvement demonstrating the phases of an acute midbrain syndrome (GERSTENBRAND 1967).

1.1. Coma prolonge. The most remarkable signs at this stage are: Continued deep unconsciousness, lack of any reaction to external stimuli, subsideration of spontaneous streching spasms, only accentuated by pain stimuli, extension of extremities, trunk and neck, beginning flexed position of the arms at the end of the stage, high tension of muscles, increased tendon reflexes, easily evoked Babinski reflexes, divergent eye balls, but no longer in fixed position, slightly accnetuated brain stem reflexes, spontaneous Chewing mechanisms. This stags may last 3-7 days befor changing into the "Parasomnia" stage.

1.2. Parasomnia. Typical signs at this stage are. Unconscious ness, closed eyes, no reaction to external stimuli, mass movements of the upper extremities and simultaneous increase of stretching of the legs after pain stimuli, flexed position, high muscle tone beginning to decrease, enhanced tendon reflexes, exaggerated pyramidal signs, diminished divergence of eye balls, increased brainstem reflexes, stabilization of vegatative reactions, augmented automatic chewing with increase to external stimuli, snout reflex is postive. The parasomnia stage lasts 3-5 days and is followed by the state of akinetic mutism.

1.3. Akinetic mutism. The characteristic signs are: Opening of the eyes from short to extended periods, No blinking reflex; vegetative hypertension shifted to the sympathetic tense state, periods resembling sleep, slight flexing tendency in body's posture, increased asymmetric hypertensive neck reflex, mass movements

of the arms and legs afterpain stimuli, hypertension of muscles, increased pyramidal signs resulting in a flexing movement. Increased brain stem reflexes, sympathetic tonic condition, different primitive motor patterns, like tactil or oral adjustment mechanism followed by spontaneous chewing, lip closing, appearence of snout reflex, bulldog roflex, asymmetric tonic neck reflex, glabellar reflexes, BABKIN and CHOTZKO reflex.

The third phase of transition stage lasts 2-5 days and followed by the overall picture of the apallic syndrome.

2. The full stage of the apallic syndrome is defined by the following symptoms: Coma vigil with sleep-awake regulation controlled by exhaustion, eye , with following, movements, threating reflex absent, blinking lex presents, no reaction to external stimuli, only mass mo ents of the extremities to pain stimuli, accompanied by a sympathetic reaection, no emotional reaction, divergent position of the eyes, different size of pupils, brainstem reflexes exaggereted, increase of masseter tone and reflex, anemia, extended trunk, slight flexion in the elbow and knee joints, adduction of upper and lower extremites, increased muscle tone "rigidespasicity" (MARABAYASHI 1962), hyperreflexia or diminished reflexes caused by periarticular ossificative motor patterns, sucking, chewing automatism, gnathing of teeth, observation of grasping, reflex BABKIN-,GALANT-, asymmetric adn symmetric tonic neck reflexes, disinhibition of autonomic functions, signs of sympathetic tone regulation shift to parasympathetic regulation during sleep.

3.1. The remission stage of an apallic syndrome is charaeterized by reintegration of higher brain functions and a decrease in the activity of the primitive motor patterns.

3.2. The signs of remission are the disappearance of chewing sucking automatism, a change in the sleep-awake rhythm to day-nignt regulation, emotional reactions in response to pain stimuli.

3-3 In the further course of remission optical fixation and

a differentiation of emotional reaction can be observed.

3-4 Convergence reaction of the bulbi and further diminishing of the divergent position of the eye balls, grasping of subjects, recognition of well known persons,smiling reaction, voluntary movements especially in the fingers occur in the phase. At the end of this period the patient responds to very simple orders.

3.5. The following phase is marked by the symptomatology of the KLUVER-BUCY-TERZIAN-DALLE ORE syndrome. The main symptoms are: Grasping objects and bringing to mouth, biting and swallowing, change of emotional reactions to positive affects, euphoric mood, hypersexuality with hetero and homo sexual tendencies, lack of fear lack of memory, bulimia, progressive development of voluntary movements, slightly increased muscle tone, & symptome of parkinson like syndrome.

3-6 The next step during remission is the WERNICKE-KORSAKOFF syndrome. Significant signs of emotional lability can be found. The less of recent memory induces confabulation. Speech and other high brain functions have aleady progressed and voluntary movements have been normalized. The symptoms of local brain lesion can be clearly determined now. This phase lasts up to 1 year.

3-7 During further recovery, the phase of psychoorganic syndrome occurs. The main symptoms are disturbance of attentiveness and recent memory, narrowing of thought and perseveration.

3.8 At last, defect stage may occur in the development of an apallic syndrome. Some groups of symptoms can be prefiled.

a. Predominance of an organic dementia combined with special emotional disinhibition.

b. Predominance of spastic symptoms with pseudobulbar-

C. Predominance of cerebellar symptomatology.

d. Predominance of an extrapyramidal hyperkinetic sympto-

This symptomatology may be combined with symptoms of matology.

uni-or multifocal superimposed brain lesions and with symptoms of

spastic hemiparesis or aphasia etc.

The exitus of a patient after an acute brain damage may be caused by an irreversible breakdown of the circulatory system o. by a renewed secondary midbrain syndrome followed by amedullary syndrome - Death of the apallic patients usually occurs during the full stage, or during the early period of remission stage. The prognosis of survial is favorable after entry in the KLUVER-BUCY TERZIAN-DALLEGRE syndrome. The prognosis in patients with an apallic syndrome caused by a metabolic disorder depends on the further course of the metabolic disease and the effectiveness of therapy. The full stage of an apallic syndrome shows a remarkable uniform symptomatology independent of its etiology. It may be explained by restriction of the brain functions to mesodiencephalic level. The remission of an apallic syndrome as well of the acute disintegration of brain functions to an apallic syndrome show a sys tematic sequence in their development. The apallic syndrome is a final stat in patients with chronic progressive brain disorder. An acute brain lesion may cause areversible apallic syndrome with or without the development of defect stage.

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