Recent Developments in the Rehabilitation of Patients in Vegetative State/Apallic Syndrome – Introduction

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The first case of a patient with a traumatic apallic syndrome was described in 1899 by Rosenblath, without naming this state apallic syndrome. He delineated the syndrome and the neuropathological findings of the 15 year old tightrope-walker, A. Geissler, who fell down from a height of 4 meters. 245 days after this accident the patient died, showing signs of a full stage of an apallic syndrome. The neuropathological findings, Rosenblath listed, were the same, which are found in apallic patients.

Kretschmer described 1940 a patient with a full stage of an apallic syndrome developed after acute virus encephalitis followed by an almost complete remission. In 1967 Gerstenbrand gave the first description of the traumatic apallic syndrome based on 74 patients (38 recovered). Due to a lack of knowledge of European literature, Jennet and Plum 1972 published a descriptive statement of the same syndrome calling it "persistent vegetative state after brain damage, a syndrome in search for a name". The attribute "persistent" had to be withdrawn later, because some of the patients reached various remission phases.

"Apallic" - Pallium, the Latin word for the old Greek overcoat - brought in medical nomenclature by Kretschmer 1940, describes a loss of all cerebral functions except the brainstem, without correlation to an anatomical damage of the brain and with the principal possibility of recovery. Vegetative state depicts the similar neurological state taking the vegetative disturbances as the leading symptom, but misses most of the other symptoms seen in the full stage of an apallic syndrome.

The neuropathological findings in the brain of patients with an apallic

syndrome/vegetative state are different from one patient to another. In patients with traumatic apallic syndrome the brain lesions are dependent on the direction and the force of the impact to the skull (Grcevíc 1988; Birbamer, Gerstenbrand, Grcevic,1999) leading to primary damages in the cerebral cortex, white matter, brainstem and cerebellum (Jellinger,1979; Kinney and Samuels,1994, Adams et. al 2000). Secondary traumatic brain lesions occur as a result of perifocal edema in the penumbra of primary lesions, or after a global brain edema (hypoxic, hypoxaemic etc.) developing local and/or diffuse tissue damage in cortical and subcortical areas. In the midbrain region local lesions due to tentorial herniation can be found. Secondary brain lesions can also develop due to compression in the irrigation area of cerebri posterior and the thalamo-geniculata. After venous tailback local lesions can be observed mainly in the brainstem (Gerstenbrand,1967; Jellinger,1979). Nearly identical neuropathologic patterns can be found in apallic patients after encephalitis or multilocular brain infarctions.

After hypoxia the neuropathological findings show laminar necrosis in cortical regions. Local posthypoxic lesions are found in thalamus, striatum and hippocampus (Jellinger,1979; Kinney and Samuels,1994). Similar findings to the traumatic apallic syndrome can be seen due to tentorial herniation with compression, of the cerebri posterior and its irrigation field, especially in the thalamo-geniculata with laminar necrosis in the occipital lobe and local lesion in the thalamus (Rumpl,Gerstenbrand 1991; Graham et al. 1995, Jellinger 1999).

Pathophysiologically the brain functions in the apallic syndrome/vegetative state are reduced to the meso-diencephalic level. This functional status can be compared to the meso-pontine Anencephalus described by Monnier and Willi (1947). A comparison to the brain functions of a newborn may be made.

There are four different ways an apallic syndrome/vegetative state can generate. Two of them are following an acute damage to the brain or the brainstem. After a progressive diffuse brain disease (Creutzfeldt-Jakob disease, Alzheimer disease, etc.) an apallic syndrome/vegetative state is developing as a final state. Special drug treatment can produce passagere apallic symptoms.

After an acute severe brain damage (traumatic, encephalitis, extra and intracranial hemorrhage, acute subarachnoidal hemorrhage, acute multilocular brain infarctions), as well as hypoxia (cardiopulmonary arrest, strangulation, nearly drowning, etc.) and endogenous or exogenous Intoxications, obligatory symptoms of an acute midbrain syndrome (Gerstenbrand and Lücking,1971) /midbrain-upper pons stage (Plum and Posner,1980) in few cases an acute bulbar brain syndrome (Gerstenbrand and Lücking,1971) medullary stage (Plum and Posner,1980) is passing followed by a transitory stage to the apallic syndrome (Gerstenbrand 1967, 1974,1994) (see Fig. 1).

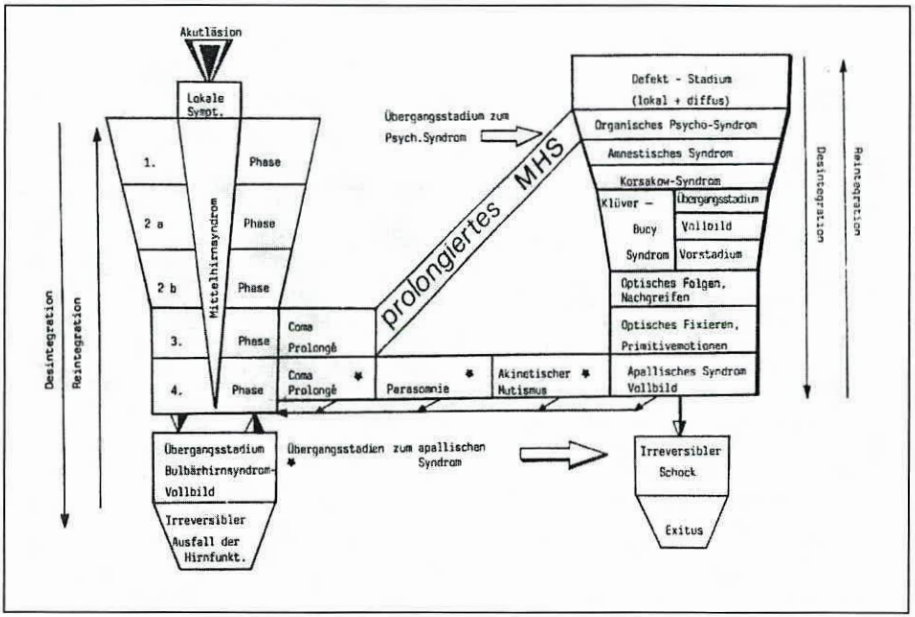

Figure 1: Schema of the development of an apallic syndrome with initial, transistory, full state and remission.

A direct lesion in the midbrain and pontine region due to local hemorrhage, encephalitis, tumor etc. induces initial symptoms of the midbrain-pontine lesion followed directly by the apallic syndrome/vegetative state.

After a progressive diffuse brain process like Creutzfeldt-Jakob disease, Alzheimer disease, Pick's atrophy, Chorea Huntington disease, chronic exogenous intoxication (Minamata disease), chronic endogenous intoxication (Coma hepaticum, etc.) the apallic syndrome/vegetative state is a final state without the possibility of any remission (Gerstenbrand 1972, 1999).

A passagere form of apallic symptoms can be observed after high potent neuroleptic drugs like Majeptil (Bruck and Gerstenbrand,1967)

The main symptoms of an apallic syndrome are coma vigile, disturbance of sleep/wake rhythm, no registration and no contact to the sorrounding, optomotoric dysfunction, typical position of body and extremities (embryonic posture), muscle tonus disturbance as rigidospasticity, primitive motor patterns (Kretschmer,1953) and vegetative dysregulation. Neurophysiological examinations (EEG, evoked Potentials, etc.) are showing typical abnormalities. Neuroimaging methods indicate more or less severe structural lesions in brain, brainstem and cerebellum (Gerstenbrand,1999; Jellinger,1999).

In an apallic syndrome after an acute brain damage the initial stage

with acute midbrain symptoms, the transitory phase and the full stage is developing during 10 to 18 days (Gerstenbrand 1967, 1977, 1999). In some cases a prolonged midbrain syndrome can be observed with partial apallic symptoms (Gerstenbrand and Rumpl,1992) (Fig.1). In 80 % of the patients, remission can be registrated, passing eight different phases (Fig.1) (Gerstenbrand 1967, 1977, 1999). In the phases I to II, the patient starts to build up contact to his surrounding, parallel the coma vigile is diminishing (Gerstenbrand 1967,1977,1999). The phases III to V are characterized by Klüver-Bucy symptoms with typical "higher organized" motor patterns and sexual disinhibiton. In the following three phases the contact to the surrounding is continuously getting more, disturbances of higher and highest brain functions are more profiled (Gerstenbrand and Lücking,1977). The neurological symptoms induced by primary and secondary local and regional lesions are covered by the apallic symptoms during the full stage, and will be found later in the remission stage. Local brain lesions like hemiparesis, optomotoric disturbances and other midbrain symptoms first can be seen in remission phases I and II together with primitive emotional reactions and increase of awareness. In the further course different local symptoms like Parkinson signs, thalamic and cerebellar symptoms, first signs of aphasia etc., as well as symptoms of regional lesions like frontal and temporal lobe symptoms can be observed. The severity of neurological and neuropsychological deficits have great influence on the further outcome of an apallic patient. Only in 15 to 20 % there is no remission. In every phase, mostly in phase I and II, the remission course can stop. But it is possible, that after a certain time (weeks to months), the remission course continues again. When remission has reached the phase of Klüver-Bucy the further prognosis is favourable (Formisano, Gerstenbrand,1993; Formisano et al,1995; Gerstenbrand, 1999).

In the outcome after an apallic syndrome/vegetative state a defectless, a minor defect state, moderate defect state, severe defect and severest defect state can be differentiated. 30 % of the patients who achieve remission can be resocialized, 20 % of patients show moderate defects with a different grade of invalidity, 30 % of patients have severe neurological defects needing nurse care. 20 % of patients with severest neurological defects die in the first two years mostly in full stage of the apallic syndrome or in the first remission phases (Gerstenbrand 1972,1977,1999 Formisano et al,1995). A patient in full stage of an apallic syndrome or in early remission can survive up to five and more years without any change of his state (Gerstenbrand, 1999).

The remission course can be complicated by tertiary lesions like polyneuropathy, encephalopathy, pontine myolinolysis and myelopathy. Quarterly lesions like hydrocephalus occlusus, brain abscess, etc. can interrupt the remission course. Complications like periarticular ossifications, joint contractures, decubitus etc. can influence the remission in some cases in

a severe manner. The primary brain lesions depend on the impact of the force to the skull, and cannot be influenced. On the other hand, secondary brain lesions following an acute brain damage can be minimized by early treatment on intensive care unit. In a high percentage, tertiary lesions, using modern treatment, can be prevented, and should therefore be called iatrogenic.

Quartery lesions and complications need a sufficient and contionous control (cerebral CT, x-rays etc.) and early interventions.

In the therapy of an apallic syndrome, the demand is, that every patient after an acute brain or brainstem injury has to be treated in a special rehabilitation centre. Such a centre needs special equipment, as well as experienced medical, paramedical and nursing staff. This centre should have 10 to 15 beds, in a nearby area to a department for Neurorehabilitation. The first obligation for the treatment of apallic patients is to start in any case with the enthusiasm to cure the individual patient. For every patient an individual rehabilitation program, exactly time-tabled, has to be planned and has to be adapted during the course of the remission in correlation to the patients improvement.

The main points of the therapeutic program are special drug treatment, special modified physiotherapy, specialized stimulating therapy, early beginning of logopedia and ergotherapy, and last not least early beginning of a resocialisation program using the therapeutic community (Gerstenbrand and Hoff,1968). In details the following points have to be listed.

- Special nursing system with regular changing of the patients position, high-calory dietary with 2500-3500k cal/d (Gerstenbrand, Rossi 1972; Gerstenbrand,1999), basal nursing stimulation, etc.
- Drug treatment with substances for stimulation of brain metabolism and for blood circulation (piracetam, CDP-Cholin, etc.), antiparkinson medications, beta-blockers to reduce the high noradrenalin level (Hörtnagl et al. 1980).
- Physiotherapy with tonus regulating reflexes (asymmetric and symmetric tonic neck reflexes), passive movements of the extremities.
- Development and building up spontaneous motoric activities initiated with Klüver-Bucy motor patterns (grasping reflexes, etc.).
- Stimulation therapy (Gerstenbrand 1992,1999; Hildebrandt,2002), antigravity stimulation (verticotherapy with bedside wheelchair sitting, standing-board), pressure massage and vibration massage of sole, haptic stimulation including basal nursing stimulation, later visual and acoustic stimulation (Posner 1978) stimulation of the olfactory sense.
- Ergotherapy starting as soon as possible.
- Logopedia initiated with the growling reactions.
- Resocializing strategies in cooperation with family members, friends

and profession colleagues and with the help and support of social workers (Gerstenbrand Hoff 1968).

Current and exact evaluation of apallic patients including clinical, neurophysiological and radiological examinations have to be made every 3 weeks. Daily a monitoring list has to be filled up (Gerstenbrand, Lackner, Lücking 1977). If, after 6 months, no remission course is observed, clinical deficits and eminent structural damages of the brain and midbrain as well as tertiary lesions are found, or if the remission course stops in phase I or II, the prognosis is unfavourable, a transfer to a special nursing home for apallic patients has to be considered. The relatives have be to informed in all details. In complicated cases a consilium with the cooperating specialists is necessary.

In the special nursing home for apallic patients a "continous stimulating care" is the basic condition.

Body positional program, high caloric dietary scheme, special physiotherapy and stimulating treatment (verticotherapy, etc.) has to be provided. Such a special nursing home for apallic patients should have around 20 beds and should be in a nearby area and in close contact to the special centre for apallic patients.

Summary

In 80 % of patients with an apallic syndrome after an acute cerebral injury remission can be observed. By using modern treatment, 30 % of these patients can be resocialized. With a consequent treatment in the acute state, followed by a continuously adapted and with an individual therapy program, the percentage of this group could even be higher. An early start of the rehabilitation program is an obligate demand as well as the most optimistic attitude towards every patient. Every patient with an apallic syndrome has to be treated in a special centre. A definite prognosis even using modern diagnostic methods can not be made in the first six months.

Even after 12 months a remission can be observed (Gerstenbrand and Lücking, 1977), a once stopped remission can proceed. Patients with an apallic syndrome without an advance in their remission course, have to be transferred to a special nursing home centre for apallic patients with a continous stimulating care.

In Central, East and South parts of Europe no discussion is possible about withdrawal of nutrition and liquid supply, a practical way of euthanasia.

Generally patients with a traumatic-apallic syndrome and after encephalitis do have a better outcome compared to patients after hypoxia. Patients with an apallic syndrome as a final stage after a diffuse progressive brain disease, also need a special care and must not get rid off.

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Reprinted from:

NEUROLOGICAL REHABILITATION

Proceedings of the 3rd World Congress

Venice (Italy), April 2-6, 2002

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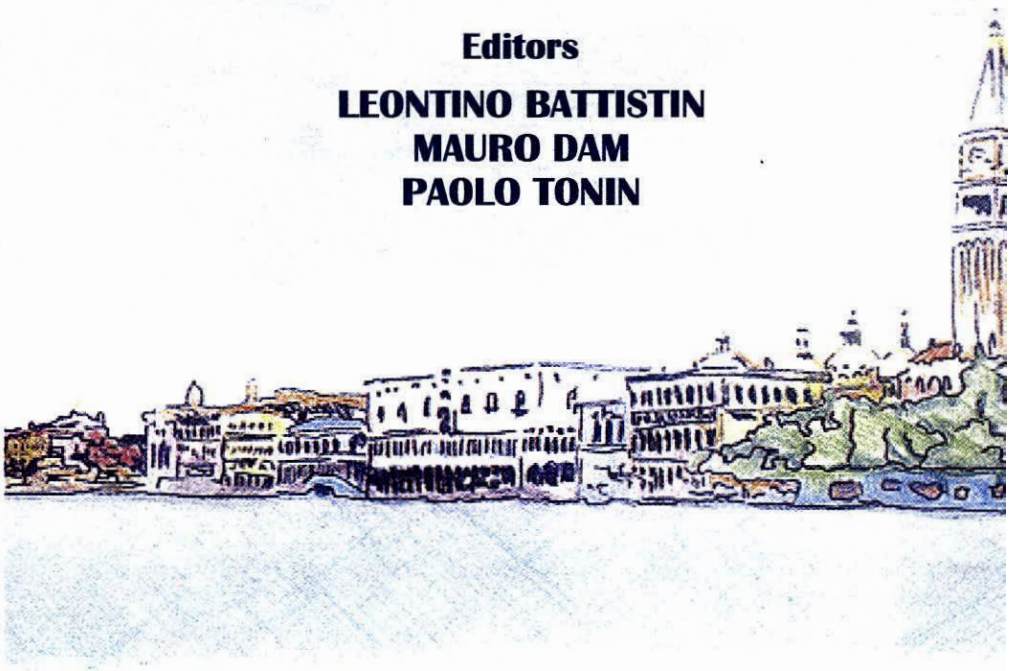

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