2. The Symptomatology of the Apallic Syndrome

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Introduction

From the type of motivating disease and the course of the first observed cases of an apallic syndrome described by Kretschmer (1940), the conclusion can be drawn that the concept "apallic" was conceived by its creator as a term denoting a functional disturbance of the cerebrum and was not intended to describe the existing brain damage as a morphologic substrate. Thus in the apallic syndrome a functional dynamism exists and in some cases remission is possible.

The apallic syndrome can develop in two different ways:

1. Following an acute process of the brain which affects the total cerebrum or which interrupts the ascending and descending pathways by a local lesion in the midbrain region, or a combination of both. The latter constitutes the most severe acute brain damage. To this group belong cerebral hypoxydosis of different origins (strangulation, obstruction of the trachea, anesthesia accidents, cardiac arrest, heart operation, etc.); brain edema caused by allergic reaction, etc.; diffuse encephalitis; gas embolism; metabolic disturbances (hepatic coma, uremic coma, etc.); prolonged insulin shock; exogenous intoxication (carbon monoxide, mercury poisoning, etc.); and brain injury as well as processes of the midbrain region such as tumors, circulatory disturbances (upper basilar syndrome), and subarachnoidal hemorrhage with tamponade of the basal cisterns and incidents after stereotactic operations. In the majority of events mentioned in this group the diffuse brain edema is the cause of a mass movement in the supratentorial cavity followed by tentorial herniation and secondary lesion of the oral brain stem. In this form of apallic syndrome a remission is possible, but the full syndrome can remain until the death of the patient without the slightest tendency towards remission.

2. A diffuse progressive process of the cerebrum affecting the cortex, or the white matter, or both together. Neurologic diseases which can cause an apallic syndrome in this manner are, for example, senile or presenile brain atrophy, diffuse brain sclerosis, subacute sclerotic leukencephalitis, the severest forms of multiple sclerosis and the Marchiafava-Bignami syndrome. The apallic syndrome occurring in a progressive process of the cerebrum is an irreparable end-state.

In accordance with the above description, the apallic syndrome develops in certain stages. In the acute processes of the brain an initial stage is obligatory, showing the different phases of the acute midbrain syndrome, in rare cases the acute bulbar brain syndrome, followed by a transient stage.

In the remission stage several phases of the reintegration of the higher brain functions of the brain, the same phases as in the remission stage occur in a particular sequence, according to the disintegration of the higher brain functions, but only in reverse.

The symptomatology of the apallic syndrome in all its stages can be divided into the symptom categories of the disturbance of consciousness (activity and content of consciousness) and vigilance; the reaction to external stimulation; the emotional reaction; the oculomotor function; the motor function of the face, throat, and body; the body posture; the primitive motor patterns; and the autonomic functions.

The Symptomatology of the Full Stage

The symptomatology of the full stage of the apallic syndrome is as follows:

1. Coma vigile with sleep-awake regulation controlled by exhaustion. The patient is awake but has no conscious activity nor any conscious capacity. The apallic patient lies with eyes open, not noticing events around him (Fig. 1). He can neither focus nor follow optically. The threatening reflex cannot be evoked, but the blinking reflex is present. This awake state becomes independent of time and is interrupted by periods of sleep, which are characterized by their depth. During the awake state the autonomic functions are in a sympathetic tonus, during the sleep periods in a parasympathetic tonus.

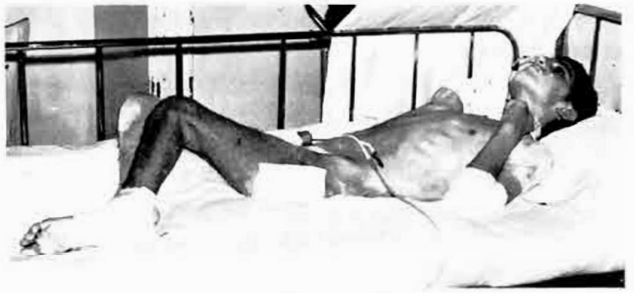

Fig. 1. Traumatic apallic syndrome, full stage (Patient A.D., age 17 years, male, 7 months after accident). Coma vigile, flexion position of extremities with contractions of main joints, signs of marasmus, decubital ulcers

2. The patient shows no reaction to external stimulation. Pain stimuli cause, after a period of latency, mass movements of the extremities corresponding to the holokinetic movements of the newborn (Lesny, 1965), accompanied by a stress reaction (enlarging of the pupils, increase in breathing and pulse rate as well as increase in blood pressure).

3. Emotional reactions are not present.

4. The eyes are in a divergent position and are of different size; convergent reaction is not evocable. Eye movements are slow and saccadic and are mostly evoked by vestibular stimuli. The oculocephalic reflexes (doll-head phenomena) are diminished; the vestibulo-ocular reflex (cold calorization) shows a tonic reaction. Size of the pupils varies according to the autonomic tonus. The light reaction is diminished and tonic; the pain reaction (cilio-

5. The tonus of the masseter muscle is increased, especially in apallic cases after brain hypoxia; the masseter reflex is also increased. There is a significant amimia. Swallowing movements occur only as automatisms together with chewing automatism and are considerably slowed.

6. The posture of the extremities shows a slight flexion in the elbow and knee joints, an adduction of upper and lower extremities, a fist position, and a plantar flexion in the foot angle. The trunk is extended (Fig. 1). In some cases the flexed position is seen; in other cases a stretch position of the legs can be observed. Sometimes the posture of the asymmetric tonic neck reflex with deviation of the head to one side and a stretched position of the chin extremities and flexion of the other side can be observed. In young children with an apallic syndrome an abduction posture of the upper extremities is possible. The muscle tonus of the extremities and the trunk is increased and shows signs of spasticity and of rigidity ("rigido-spasticity": Narabayashi, 1962). There is a hyperreflexia of all tendon reflexes. However, in some cases the tendon reflexes are diminished because of contraction of the joints, periarticular ossification, as well as a highly increased muscle tonus. The pyramidal signs can be evoked. In some cases the escape reflex can be evoked in the lower extremities, in rare cases overlapping onto the other side or from the legs to the arms. The escape reflex is always accompanied by sympathetic reaction corresponding to a stress reaction.

7. Primitive motor patterns. The primitive motor patterns have a pathognomonic value in the apallic syndrome. These motor actions can be divided into different groups, according to their functional basis, for instance, the motor patterns of the oral sense, the grasping reflexes, the mental reflexes, and the fright reaction. Moreover, the position and posture reflexes can be observed. In cases of an apallic syndrome of the primitive motor patterns of the oral sense, the sucking-chewing automatism appears; in others only the sucking component appears.

Gnashing of teeth can be an equivalent of chewing automatism. This motor pattern may occur spontaneously or can be evoked by specific or unspecific stimulations.

The oral adjusting mechanisms can be divided into tactile and optic forms. In the full stage of an apallic syndrome only the tactile form can be elicited; this is accomplished by perioral stimuli.

Sometimes the tactile adjusting oral mechanisms are followed by sucking movements, similar to the lick-sucking, and by the bulldog reflex. During a sustained bulldog reflex the lick-sucking can continue. In rare cases the tactile adjusting oral mechanisms are followed by magnet reaction.

The snout reflex can be elicited in most cases of apallic syndrome.

In single cases the motor pattern of breast searching can be observed. This motor pattern occurs particularly in cases of progressive brain damage or after a diffuse encephalitis.

In some cases an intensive yawning is observed. The yawning motion may be repeated several times. It is similar to the yawning of the brain-damaged newborn.

The grasping reflexes are divided into the phasic and the tonic grasping reflexes (Wieser, 1957). The tonic grasping, which is evoked by proprioceptive flexion of the fingers, can be followed by a flexion of the elbow joint and by an adduction of the whole arm. An intensive tonic grasping reflex enables one to pull the patient up. It is possible to intensify the motor patterns of the oral sense by the grasping reflexes. In rare cases the Chodzko

The so-called mental reflexes—mental because of the contraction of the mental muscles—first described by Marinesco and Radovici (1920) are obligatory in the apallic syndrome. Different types are known, the most common being the palmomental reflex, elicited by scratching the palm with a blunt object. Not only the homolateral mental muscle is contracted but also the reflexogenous zones often spread to the whole arm, sometimes to the trapezius muscle. The pollicomental reflex can be elicited by intensive scratching of the top of the pollux. Other types belonging to the mental reflex group are the cygomatico-, mento-, labio-, and corneomental reflexes, as well as some others.

The same pathophysiologic basis as in the mental reflex group can be observed in the Babkin reflex, elicited by simultaneous intensive pressure of both palms, followed by an opening of the mouth sometimes combined with an attenuated stretch position of the trunk. This motor pattern can be found in every premature baby and in cases of an anencephalus. Very rarely is the Lesný reflex, the one-sided Babkin reflex, observed.

Of the group of fright reflexes, the glabellar reflex, the head retraction, and the total-fright reflex are most often observed.

In some cases climbing movements of the legs—also called tread movements—appear. The Galant reflex is also very seldom observed. (After scratching of the lateral trunk the homolateral hip moves towards the stimulated side.)

From the position and posture reflexes the asymmetric and symmetric tonic neck reflexes are elicited in nearly every case in a more or less intensive form. Sometimes the posture reflexes can be found from one extremity to the other, or at the head. These reflexes usually occur latently. It must be pointed out that these reflexes are of great importance for early physiotherapy in the so-called method of tonus-regulating reflex therapy.

The support reaction can be found only in some cases and nearly always as support reaction in the flexion position. These reflexes can also be used in early physiotherapy.

8. The disinhibition of the autonomic functions has a distinct influence on the condition as well as on the further course of an apallic patient. As mentioned above, during the awake state all signs of a sympathetic tonus regulation may be found shifting over after the entrance of the sleeping period in a parasympathetic regulation level. The period of the predominance of the sympathetic tonus regulation has a tendency towards chronification of an emergency reaction. This tendency towards a sympathetic tonus regulation state may explain the necessity for hypercaloric nutrition in apallic patients. In connection with this regulation of the autonomic system, complications such as marasmus, bedsores, and so on may be observed.

In the course of autonomic dysregulation the incontinence of bladder and rectum must be cited. Of special importance is the diminishing resistance to infections of various kinds. Most cases have a cystitis. The overaction of some glands producing hypersalivation, ointment face, and sweating yield parallels a Parkinson symptomatology which is enforced by the presence of akinesia and amimia. Therefore, Parkinson symptomatology was found to be a part of the apallic syndrome (Gerstenbrand, 1967).

As already mentioned in the introductory remarks, the apallic syndrome can develop in two different ways. In both ways, in the apallic syndrome occurring (1) after a severe acute brain lesion, as well as (2) in a progressive process of the cerebrum, a fixed course towards a syndrome can be determined. In the apallic syndrome, as an end state of a pro-

functions, corresponding to the distintegration of the cerebral function (Jackson, 1958). The course of disintegration of the higher brain functions is directly comparable to the reintegration of the brain activity during the remission stage of an apallic syndrome resulting in severe brain damage.

In cases of an apallic syndrome following an acute brain damage, of different etiology, an initial stage and a transition stage appear before the full stage. The initial stage and the transition stage are described in Chapter 3.

The Remission Stage

During the remission stage of an apallic syndrome due to acute damage of the brain, reintegration of the higher brain functions appears, together with a decrease in activity of the primitive motor patterns which become integrated into the increasing voluntary movements, and a normalization of the disinhibited autonomic functions. During the course of the remission, considered from the functional point of view, two periods may be differentiated: (1) the stage of establishing contact with the surroundings, coupled with an increased mental awareness, as well as with a reintegration of the turning-towards movements (*Zuwendungsmotorik*), and (2) the second stage during which the remaining disinhibition symptoms disappear and voluntary motor as well as higher brain functions reappear.

The clinical course of the remission stage can be divided into several phases. The beginning of the remission is indicated by the emergence of primitive emotional reactions in response to pain stimulus, corresponding to a primitive alarm reaction and accompanied by an emergency reaction. Moreover, a change in the sleep-awake rhythm influenced by day-night regulation, as well as a disappearance of the chewing-sucking automatism can be observed. These first signs of remission can occur either after several days of a lasting full stage or after some months. The duration of this phase can vary from several days to several months or may become a final one.

In the further course of remission optical fixation occurs (Fig. 2). The patient turns towards objects brought into his visual field. At the same time a differentiation of the emotional reaction can be observed. Besides the primitive alarm reactions during nursing care

Fig. 2. Traumatic apallic syndrome, remission stage, second phase (Patient A.Z., age 29

and injections, displeasure reactions are elicited. The change in the sleep-awake regulation progresses. This development parallels a progressive disappearance of the coma vigile. This phase may also last for a varying length of time but can be the end stage of the remission.

During the progression of the remission the apallic patient shows optic following together with deviation of the eyes, later of the head, with initial convergence reaction of the bulbi and diminishing of the divergent position of the eyeballs, as well as grasping of objects put in front of him. The emotions show positive effects with slight signs of recognition of special persons and the first signs of a smiling reaction (Fig. 3). The first voluntary movements occur, usually in the fingers. At the end of this phase the patient responds to very simple orders (opening of the mouth, closing of the eyes, etc.). In this phase the latest symptoms of superimposed focal lesions of the cerebrum and sometimes of the cerebellum may be found (hemiparesis, etc.).

The following phase is marked by the symptomatology of the Klüver-Bücy-Terzian-Dalle Ore syndrome. The main symptoms are grasping and bringing to the mouth every object in the surroundings, followed by biting and swallowing (Fig. 4); change in the

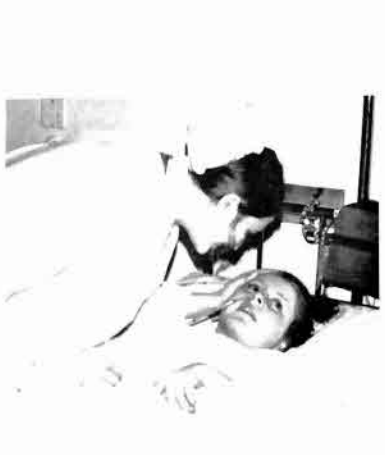

Fig. 3

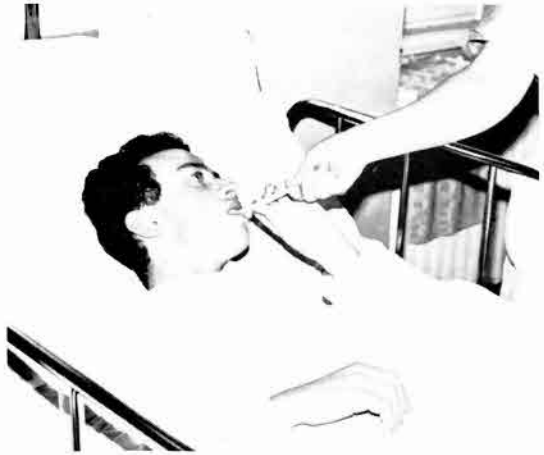

Fig. 4

Fig. 3. Traumatic apallic syndrome, remission stage, beginning Klüver-Bucy phase (Patient A.X., age 12 years, female, 10 months after accident). Adulation reaction (*Schmeichelreaktion*)

Fig. 4. Traumatic apallic syndrome, remission stage, Klüver-Bucy phase (Patient N.D., age 20 years, male, 20 days after accident). Shown object grasped immediately and put into mouth, followed by sucking and biting movements

emotional reactions to positive effects; establishment of an euphoric mood; hypersexuality with hetero- and homosexual tendencies and lack of shame reactions during masturbation; and lack of fear reactions sometimes accompanied by the appearance of rage reactions, bulimia, and by a lack of memory. The primitive motor patterns show a further differentiation and some disappear (Babkin reflex, Chodzko reflex, posture reflexes, etc.). The

The symptomatology described above is completely different from that of the apallic syndrome and could be explained by the rising of the functional niveau to the higher level of the disinhibited limbic system. It must be emphasized that in a number of cases only a partial symptomatology of the Klüver-Bucy-Terzian-Dalle Ore syndrome can be observed in this remission phase.

During remission the symptomatology of a Korsakoff syndrome occurs. In the meantime, having started in the first phase, the redevelopment of speech and other higher brain functions has progressed and voluntary movements have been normalized. Significant signs of emotional lability can be found. In some cases psychotic decompensation may appear during this phase. The symptoms of a local brain lesion can now be clearly determined. In rare cases this phase can last up to 1 year.

During further recovery the phase of psychoorganic syndrome occurs followed by a defect stage.

The Defect Stage

In the defect stage groups of symptoms can be profiled:

1. Predominance of an organic dementia combined with a special emotional disinhibition.
2. Predominance of spastic symptoms with pseudobulbar-paralytic symptomatology
3. Predominance of cerebellar symptomatology
4. Predominance of a Parkinson symptomatology
5. Predominance of an extrapyramidal hyperkinetic symptomatology

This symptomatology, typical for the defect stage of the apallic syndrome, may be combined with the symptoms of uni- or multifocal superimposed brain lesions and with the symptoms of a spastic hemiparesis, aphasia, etc.

It should be emphasized that in rare cases an apallic syndrome can follow the course of a transient syndrome without exhibiting defect symptoms, or the defect symptoms may be very slight.

The exitus of an apallic patient after acute brain damage may be caused by an irreversible breakdown of the circulatory system, or by a renewed secondary midbrain syndrome followed by a bulbar brain syndrome. Death of the apallic patients in the former group usually occurs during the full stage, or during the early period of the remission stage. After entry into the Klüver-Bucy-Terzian-Dalle Ore phase the prognosis for survival is favorable.

The Apallic Syndrome as a Sequel to a Diffuse Progressive Brain Process

In the progressive cerebral processes followed by an apallic syndrome as a final state, the apallic symptomatology develops correspondingly to the disintegration of the higher brain functions involving the remission stage, but in reverse. It can be presumed that, due to progress in intensive care treatment, the number of apallic patients of this etiology will

The clinical course of the progressive loss of brain function to the apallic level is described in Chapter 5. In order to give a rough idea of the course of development, it should be mentioned that during the first period of the clinical course psychiatric symptoms predominate. These symptoms can occur either in the different forms of the exogenic reaction types as described by Bonhoeffer (1910) in a sequence of different pictures, or with the psychoorganic syndrome according to E. Bleuler (1969). In most cases a Korsakow symptomatology appears followed by a Klüver-Bucy-Terzian-Dalle Ore syndrome as has been described, for example, by Pilleri (1961a). The further course to the full stage corresponds on the whole to the first period of the remission.

The full stage of an apallic syndrome as a sequel to a progressive process of the cerebrum shows for the most part the same symptomatology as an apallic syndrome after acute brain damage without local symptoms of a brain stem lesion.

Conclusions

The full stage of an apallic syndrome, independent of its etiology, is remarkably uniform and may be explained by a restriction of the brain functions to the mesodiencephalic level. The remission of an apallic syndrome, as well as the disintegration of the brain functions to an apallic syndrome, show in their development a systematic sequence. A difference is found, however, in the remission course caused by superimposed brain lesions. While in cases with a progressive brain process the apallic syndrome is a final state, the full stage of an apallic syndrome caused by an acute brain lesion can be reversible to a defect stage, in some cases without clinical symptoms as a transient syndrome without defect (*defektfreies Durchgangssyndrom*).

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