1. Clinical Picture and Problems in Terminology

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The clinical picture of the apallic syndrome was occasionally described in casuistic studies at the end of the nineteenth century. No special attention was given to the clinical picture, and it was described in a rather incidental way. The first description was probably given by Rosenblath (1899), in which a 15-year-old tightrope walker, after toppling from a high wire, fell immediately into a coma and developed stretch cramps. After 2 weeks the patient became strangely awake and this state did not change until the death of the boy 8 months after the accident. It was not until 1940 when Kretschmer—based on his own clinical observations but without knowledge of the older descriptions—recognized the particular importance of the symptom complex. He called it apallic, since in his opinion the condition occurs as a result of extensive disturbances of the pallium in its entire functioning.

Although identical clinical pictures were later described in other terms which will be discussed below, Kretschmer's term has largely prevailed despite some difficulties[1]. While there are some objections, the authors are very much in favor of retaining the expression apallic syndrome to identify the *clinical* picture. The term is being used extensively and gives some recognition to Kretschmer's original effort. In addition, the term can be easily translated (French: *syndrome apallique*, Italian: *sindrome apallico*, German: *apallisches Syndrom*). However, some misunderstandings have occurred. A misspelling—e.g. "aphalic syndrome"—might suggest a loss of penis; the Italian translation could be retranslated into "syndrome without testicles." These rather remote possibilities for error have not proven to be a serious obstacle to the usage of the term. Furthermore, it should not be used solely in its literal Latin meaning ("derobed") and thus be applied only to diseases that are caused by extensive damage to the cortex.

Philosophical objections of this kind do not often lead to scientific discoveries. It should be emphasized again that, in the authors' opinion, the term applies only to the clinical picture and that the pathogenesis, at this point, has not yet been clearly established—the connection between cortex and brain stem may be interrupted anywhere: e.g. pallium, white substance, midbrain. Furthermore, it is common practice to identify the etiology by adding a special term (for example, traumatic apallic syndrome).

Clinical Picture

As we noted in Kretschmer's original study, the clinical description of the apallic syndrome is based primarily on the report of a case of panencephalitis subacuta (Pette) published in

[1] After Kretschmer the expression "apallic syndrome" was again taken up by a study group

1940 by Conrad and Delbrügge, as well as on the case of a brain injury caused by an oblique sagittal shot through both hemispheres. But Kretschmer (1940) pointed out expressly that the same syndrome can be caused by several other diseases such as "lues cerebri concentrated mainly in the cerebrum," disseminated encephalomalacic focus, severe arteriosclerosis of the brain, etc. The "severe full stage" did not often occur.

Kretschmer's clinical description of the condition is so appropriate that we cannot add much to his comments. We will therefore quote the original description from Kretschmer's paper:

> The patient is prostrate, awake, with his eyes open. He either stares straight ahead or his eyes travel back and forth without understanding or being able to focus on anything. Attempts to attract his attention are unsuccessful or only slightly successful. Talking to the the patient, touching him or showing him objects does not result in any sensible response. The reflexive movements of flight and defense are missing. Sometimes even the reflexive return to the basic position, that is, the position of optimum relaxation, is lacking. A normal person usually returns to the basic position from an accidental position or one that is no longer used or necessary for a specific purpose, or is inefficient or uncomfortable.
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> The patient may therefore be in a position accidentally assumed that can be active or passive. This behavior can be based either on the inability to react sensibly to a stimulus or on the disturbance of a primary impulse. In contrast, the elementary irradiation of undigested and uncurbed outside stimulation may increase enormously. In spite of the fact that the patient is awake he can neither talk, recognize, nor perform sensible patterns of behavior that he had once learned. Certain elementary vegetative functions such as swallowing still exist. In addition, the familiar basic instincts such as sucking and grasping appear. . .
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> The syndrome differs from the coma in that there is no clouded consciousness (control of sleeping and waking). It differs from the dementia in that the latter results in a quantitative reduction in the function of the cerebrum, whereby the functional capacity is retained to a certain degree. However, the syndrome described here results in a blockage of the functional capacity of the cerebrum, so that in an ideal situation it might equal a panagnosis plus panapraxia.

Important Individual Phenomena:

1. The particular way in which consciousness is disturbed—coma vigile (see below).
2. Suspension of the sleeping and waking rhythm normally regulated by local time. The sleeping and waking phases are regulated instead by normal tiring. We can distinguish between waking and sleeping phases but they are very irregularly distributed over the 24-h day.
3. The lack of emotional reactions.
4. Torso and extremities are in a stretched positon.
5. Slow, pendular movements of the eyes or lack of coordination of the eye movements.
6. Appearance of primitive motor patterns.
7. The integrating systems of the vegetative functions become uninhibited.

We have seen from Kretschmer's (1940) description that the apallic syndrome basically occurs in two different forms (Gerstenbrand, 1967):

1. By an acute process which leads to the loss of the entire cerebral function. This loss may be caused by damage to the cortex, to the brain stem, or to the connecting systems at various levels. It occurs through local processes in the upper brain stem resulting from tumors, circulatory disturbances, or tentorial strangulation. Or it occurs through hypoxidosis of the various etiology, metabolic disturbances of different origin, gas embolism, acute encephalitis, edema of the brain caused by an allergy, brain injury, etc. In these cases the apallic syndrome may be a terminal or transitional syndrome.

2. By a graduated breakdown of the cerebral function which can affect cortex and marrow either separately or simultaneously. The clinical picture of the apallic syndrome caused by

processes of this kind (senile or presenile atrophy of the brain, diffuse sclerotic leukencephalitis, multiple sclerosis, Marchiafava-Binami syndrome, etc.) is terminal.

The development of the apallic syndrome to full stage, caused by graduated processes in the cerebrum, shows phases that can be clinically differentiated. These phases may be regarded as the disintegration of the cerebrum functions to the level of the mesencephalon. During the remission phases we can again differentiate the same kinds of phases as in the graduated disintegration. Reintegration, however, in the remission process, can come to a halt at a certain phase and that phase may remain as a defect syndrome.

Explanations of Terminology

Since a commonly known and recognized expression for the clinical picture of the apallic syndrome was lacking for a long time, the same clinical pictures have been described by various terms. The following is a list of all known terms brought into relation with the clinical picture of the apallic syndrome given above:

1. Coma vigile: The most familiar term is coma vigile, widely used in French literature. In a number of studies this term was the full stage, but the descriptions involved other clinical pictures as well. In the descriptions by Mollaret and Goulon (1959) we recognize rather a sopor. Furthermore, the term coma vigile is really a contradiction in itself, since the familiar connotation of "coma" actually excludes the possibility of being awake. Yet it is this incompatibility of contrasting symptoms in the apallic syndrome–no object content of consciousness and yet being awake–that appear to make the expression so fitting. Since the apallic syndrome includes so much more than only this strange form of changed consciousness, it is not suitable to accept coma vigile as a synonym for the apallic syndrome as a whole. Gerstenbrand (1967) has therefore used coma vigile only to describe the state of consciousness in the apallic syndrome since it is often necessary in a given context to emphasize this side of the entire symptom complex. It is suggested that the use of this term be retained.

2. Parasomnia: This expression was created by Jefferson (1944) to find a better name for that particular type of disturbed consciousness following "normal" brain traumas in which the patient can, for example, react verbally when called but falls immediately back into a kind of sleep. It is important to Jefferson that the patient does not react to stimulation with a response that will assist in analyzing the problem (his definition of consciousness). In accordance with the usual nomenclature, parasomnia can mean a state of clouded consciousness which may extend from slight somnolence to coma, whereas in the apallic syndrome the "skalar" consciousness is still intact. Among the three cases briefly reported by Jefferson, therefore, an apallic syndrome is not present and yet the term parasomnia has been used occasionally to describe the state of consciousness in the apallic syndrome.

A further objection to the usage of the expression parasomnia is the fact that before Jefferson it was used to describe the various forms of troubled sleep (anxiety dreams, somnambulism, spasms when going to sleep).

3. Hypersomnia: Even the old and common term hypersomnia has occasionally been used to identify an apallic syndrome. Façon et al. (1958), employing the term *hypersomnie prolongée*, described a female patient aged 78 whose condition showed, at least occasionally,

an apallic syndrome during the 3 years of her illness which occurred after occlusion of the arteria basilaris. French (1952) also talks of hypersomnia in a case which corresponds to an apallic syndrome. The usage of the term hypersomnia does not correspond to the familiar connotation of this expression (Michaelis) and thus should not be applied. The authors, however, might again have wanted to emphasize the particular posttraumatic state of sleep known as parasomnia.

4. Akinetic Mutism: The inhibition of all motor functions, including speech, gesticulation, and facial expressions, is the central symptom of the akinetic mutism, conceptualized and solely based on the phenomena by Cairns et al. (1941). However, according to the description by these authors, the patients can be fully awake and remember everything well. The condition can be entirely functional; patients with akinetic mutism may eventually get up, go to the toilet, and return afterwards to the akinetic mutism (e.g., Case 3, Sutter et al., 1959). A similar situation arose in the first case described by Cairns et al. (1941) of an epidermoid cyst of the third ventricle, in which the akinetic mutism disappeared within a few minutes every time the cyst was drained.

We are dealing, therefore, with a syndrome all on its own which Jefferson (1944) already regarded as being separate from parasomnia. According to him, the difference lies above all in the particular tendency of parasomnia patients to sleep, which does not occur in the akinetic mutism. It is also clear that there is actually no close relation between the akinetic mutism and the apallic syndrome when one looks at their respective full stages. But since apallic patients may for a long time be akinetic and mute, it is not surprising to find the symptoms of the apallic syndrome described in case histories of akinetic mutism. There is one case of an apallic syndrome among the cases of akinetic mutism and occlusion of the A. vertebralis or basilaris described by Cravioto et al. (1960). In one case by Lhermitte et al. (1963) the cause of the apallic syndrome was a thrombosis of the A. basilaris. Vitale (1964) describes the syndrome following an unforeseen event during the stereotactic operation of a patient with Huntington's chorea. The clinical pictures described by Umbach and Riechert (1963) following stereotactic elimination in the basal ganglia also correspond to a large degree to the symptoms of the apallic syndrome. Maspes and Marossero (1957) observed an apallic syndrome when vessels in the area of the thalamus had been damaged. The female patient described by Hermann and Sulat (1959) obviously exhibited the symptoms of the apallic syndrome to some extent. This also seems true of the case described by Grotjahn (1936) more than 30 years ago. Finally, Bruck and Gerstenbrand (1967) were able to show that a case of akinetic mutism caused by high doses of phenotiazine also presented the symptoms of the apallic syndrome.

5. Posttraumatic Encephalopathy: A large number of cases of the traumatic apallic syndrome have been described with this term, for example by Jellinger (1965), Jellinger et al. (1963), Chavany et al. (1955), Trillet (1949, 1961), Dechaume et al. (1962), and others. Osetowska (1964), assuming a morphologic point of view, called the cases of traumatic apallic syndrome *leukoencéphalopathie œdémateuse posttraumatique.* Kramer (1964), on the other hand, uses the expression "progressive posttraumatic encephalopathy" to describe a case which clinically corresponds to the apallic syndrome. The morphologic analysis, however, showed a transition to the *coma dépassée* already described by Mollaret et al. (1959).

6. Decerebration Syndrome or Decerebration Rigidity: We can easily understand why an apallic syndrome is occasionally described by this term (see, for example, Mumenthaler,

1961) since "brain stem spasms" are part of the clinical picture. Kretschmer (1940) showed interest in this question in his original paper and recognized that, although there is a certain similarity between the apallic syndrome and decerebration rigidity, the clinical pictures are not identical. There seems to be a tendency to apply the term decerebration rigidity to the early stages of the central brain syndrome (Sutter et al., 1959) and apallic syndrome to the psychotic changes. Hubach and Poeck (1964), who refer to a traumatic decerebration, demonstrate in a schematic diagram to what extent the development of both partial syndromes can run parallel to or separate from each other. The related expression "decortication" has only rarely been used for cases of the apallic syndrome (Nyström, 1960).

7. Coma Prolongé: The term *coma traumatique prolongé*, which is used frequently in French literature, encompasses more than the apallic syndrome. It can apply to all clinical pictures in which patients survive in a coma for at least 3 weeks (Vigouroux et al., 1964). But many cases of apallic syndrome have been described as *coma prolongé* for lack of a proper term. This fact not only shows up in the synopsis of the remission phases by Vigouroux et al. (1964), which indicates distinct parallels with phases of the traumatic apallic syndrome according to Gerstenbrand (1967), but frequently in case histories we can see that, whenever the *coma traumatique prolongé* is mentioned, an apallic syndrome is actually meant.–Fau (1956), Girard et al. (1963), Le Beau et al. (1958), Leson (1960), Reymond and Fau (1956), Wertheimer and Allegre (1953), Sutter et al. (1959). In the English literature the same expression can be found, translated as "prolonged unconsciousness" (for example, French, 1952). In Russian literature the term "prolonged state of coma" (Ivanoff, 1962; Mägi, 1965 and 1968) is sometimes applied to the apallic syndrome.

8. Posttraumatic Catatonia: This expression has occasionally been used as a synonym for the apallic syndrome (for example, Jellinger et al., 1963). Sutter et al. (1959), however, wanted to create a general term for all traumatic brain damage which is followed by catatonic stupor. Of the four cases described, only the first can be considered to be an apallic syndrome. The authors even deduce the therapeutic consequences of the treatment from the expression (see Case 3 involving a posttraumatic twilight state). Since heterogeneous clinical pictures were mentioned under this term in the original study, we fear that further use of the term posttraumatic catatonia might lead to misunderstanding.

9. Stupeur Hypertonique Posttraumatique: Fischgold and Mathis (1959) have used this term to describe a clinical picture corresponding to apallic syndrome.

10. Lucid Stupor is used by Ajuriaguerra et al. (1953) to describe a clinical picture corresponding to the apallic syndrome caused in their case by a tumor in the mesodiencephalic area.

11. Severe Dementia Following Brain Injury. In pathologic-anatomic casuistics Sabina Strich (1956) describes typical apallic syndromes as "severe dementia following head injury." These two terms are synonymous to that extent. The author did not go into details of clinical symptomatology in her later papers. Apparently her term has not been used elsewhere.

12. Démence Progressive avec Cachexie: This is the term used by Gruner (1965) for the clinical picture of a traumatic apallic syndrome which corresponds largely to the classical description.

13. Vita Reducta was introduced by Masshoff (1963a and b) and Neuhaus (1963) and is sometimes discussed in connection with the apallic syndrome. According to the definition, it is clinically related to a "deep coma, or rather a state which follows a deep coma"

(Kubicki, 1963, 1965). Schliack (1963), Schliack and Kubicki (1964) have already pointed out that the clinical picture of the vita reducta may change to an apallic syndrome. The symptoms of the vita reducta, which may briefly be described as complete loss of reflexes, poor muscle tone, and breakdown of the vegetative functions (Kubicki, 1965), show no parallels to the apallic syndrome.

This collection of the numerous terms which have hitherto been used makes visible again the usefulness of the term "apallic syndrome". But also from clinical, pathophysiologic, and therapeutic points of view this appears most useful as long as the view is not anatomic but clinical, the apallic syndrome being a total but functional interruption of the cerebral functions.

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