Prevalence of persistent vegetative state/apallic syndrome in Vienna

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The aim of the project was to survey the hospital prevalence of apallic syndrome in a federal state in Austria at an exact point in time. To achieve this, a point prevalence study was carried out on 28 November 2001 in the Vienna region. The central element was a questionnaire, which provided an exact recording of the patients' condition. An additional preliminary task was to check all the discharge diagnoses in the hospitals of the Vienna Hospital Association (Wiener Krankenanstalten Verbund) between 1996 and 2000 according to the ICD-9 diagnoses for apallic syndrome. These data should serve to cross-check the recorded results. All hospitals ($n = 48$) and nursing facilities ($n = 44$) in Vienna were included in this investigation. As the aim of the study was to record the prevalence of apallic syndrome in the population of Vienna, four patients of the group with full-stage apallic syndrome ($n = 36$) were excluded as they were from other federal states. In total, 32 hospital patients who met the clinical criteria for apallic syndrome (full-stage) were recorded. The point prevalence of apallic syndrome was 1.9/100 000 inhabitants. As a result of this study, the exact survey of hospital prevalence of apallic syndrome could be found. As a consequence, the Viennese government has developed a rehabilitation concept for patients with apallic syndrome in Vienna.

Introduction

As a result of improvements in the acute treatment of traumatic and/or hypoxic damage to the brain, there is an increased number of patients who have been diagnosed with apallic syndrome in acute hospitals or in nursing facilities. Caring for such patients is an intensive process, both in terms of time and cost, and this must be considered in future health planning. The prevalence of a disease gives the basis of any such plans (Häussler, 1999).

Very little has been published on the prevalence of apallic syndrome (Higashi *et al*., 1977, 1981; Sato *et al*., 1978; Minderhoud and Braakman, 1985; Hellema, 1991; Tasseau *et al*., 1991; MSTF o PVS, 1994). The stated frequencies vary depending on aetiology, the time of description and the varying patient populations. Additionally, the differing nomenclature compounds problems (Wade, 2001). In the European continent the preferred term is 'apallic syndrome', whereas the Anglo-American term is 'persistent vegetative state' (PVS).

The term 'apallic syndrome' dates back to a description by Kretschmer in 1940 (Kretschmer, 1940). Following on from this, the course of this syndrome was described by Gerstenbrand in 1967 (Gerstenbrand, 1967). Jennet and Plum (1972) established the term 'vegetative state'. In 1994, a manual was published by the Multi-Society Task Force on PVS in order to establish a common approach to the naming and diagnosis of this form of illness (MSTF o PVS, 1994). These two terms are defined by the same clinical symptoms (complete unawareness, sleep–wake cycles, perseveration of hypothalamic and brain stem autonomic function). In both cases the diagnostic assessment is to be arrived at by using existing criteria. The patient must not be in a conscious state.

A sleep–wake pattern must be detectable and it should be possible to register vegetative functions of the brain (Gerstenbrand, 1977; MSTF o PVS, 1994; Wade and Johnston, 1999).

Method

Based on the works of Gerstenbrand (1977), Peters and Gerstenbrand (1977), Aichner *et al*. (1982), Berek *et al*. (1993) and of the MSTF o PVS (1994) and Practice Parameters (Summary Statement) (1995), a concept for the recording of patients with apallic syndrome (full-stage) was drawn up. It is necessary to devide between full-stage and remission stage in apallic syndrome, because only the full stage is equal to the persistent vegetative state. In the remission stage of the apallic syndrome the re-integration of higher brain functions

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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. So the important diagnostic point of complete unawareness disappeared (Gerstenbrand, 1977). The same criteria also applies to the diagnostic term minimal consciousness. Consciousness was defined by James (1890) as awareness of the self and the environment (MSTF o PVS, 1994). Consequently, a patient with minimal consciousness has no complete unawareness.

The central element used in our survey was a questionnaire, which provides an exact recording of the patients' condition. The first section of the questionnaire recorded general data such as age, gender and the aetiology of apallic syndrome. The second section provided a description of the neurological condition of the patient. The criteria and general symptoms which are necessary to diagnose apallic syndrome were subdivided in this section. In the third part, the following scoring systems were used: Glasgow Coma Scale, Glasgow Outcome Scale, Edinburgh 2 Coma Scale, Barthel Score (Mahoney and Barthel, 1965; Teasdale and Jennett, 1974; Jennet and Bond, 1975; Shigioura *et al*., 1983) (Fig. 1). These scales were selected because of their frequent use in assessing patients with apallic syndrome.

An additional preliminary task was to check all the discharge diagnoses in the hospitals of the Vienna Hospital Association (Wiener Krankenanstalten Verbund) between 1996 and 2000 according to the ICD-9 diagnoses for apallic syndrome (Table 1); 80% of all hospital and nursing facilities of Vienna are included in the Wiener Krankenanstalten Verbund. These data should serve to cross-check the surveyed results. Between 16 and 21 adults and up to nine children per year were reported to suffer from apallic syndrome. From these data it is possible to determine a median of 22 newly diagnosed cases (adults and children) of apallic syndrome per year.

In order to attain a complete recording of all patients diagnosed with full-stage apallic syndrome, the following procedure was chosen. It was decided that on a specific day, all patients with the full-stage of an apallic syndrome, who at this point in time were accommodated in acute wards or in nursing facilities, should be recorded by means of a point prevalence study. The date chosen was 28 November 2001 in Vienna.

A month before this date, all medical directors of the hospitals ($n = 48$) and the heads of nursing facilities ($n = 44$) were informed of the forthcoming investigation including the date on which the study should take place. A form was included with the question: 'In your ward or department, how many patients have been in a coma (loss of consciousness) for 14 or more days?'

The question was deliberately phrased so as to record all patients with apallic syndrome, even if they were diagnosed by doctors without specialist expertise in this field of neurology. It was assumed that other similar forms of disease would be reported, but this way all patients with apallic syndrome would certainly be included.

The registration period was set from 8.00 hours on 28 November 2001 to 8.00 hours on 29 November 2001. All patients reported were clinically assessed by a single member of the team (Ch.S.) between 29 November 2001 and 1 December 2001.

Results

In total, there was a response rate of 96% from all the medical and nursing institutions in Vienna: acute wards (90%, $n = 37$); nursing facilities (100%, $n = 44$) (Table 2). Eighty-nine patients in Viennese hospitals and nursing facilities filfilled the criteria.

Of the 89 patients registered, 78 (88%) were examined in the reporting department. Eleven patients (12%) could not be examined for the following reasons: one patient was discharged the day after being registered; one examination was refused by an external doctor of a department; three patients were taking part in a therapy workshop on the day of the planned examination; two patients had already died by the day of the examination; two patients were on day release; and two patients were being transferred on the day of the examination.

Of the group of patients examined ($n = 78$), 36 persons (46%) displayed full-stage apallic syndrome. A further 9% ($n = 7$) showed remission stages (1–3) of apallic syndrome. Two patients being treated in intensive care wards could not be assessed because of their heavy sedation and mechanical ventilation and 33% ($n = 26$) showed other forms of clinical syndrome.

Despite detailed prior information, there were seven (9%) incorrect reports. In these cases (where patients were being looked after in nursing facilities) the possibility of apallic syndrome as a transition stage during a long period of hospitalization was used as a reason to register the patient. Most patients of this group could easily be contacted and responded adequately to the examiner's questions. The diagnosis of apallic syndrome, even of its remission stages, could be ruled out in these cases following consultation of the medical histories.

In the entire group of patients examined, there were 43 men and 35 women (Table 3), and a modal age of between 50 and 59 years was recorded in 17 people in this category.

Questionnaire used for the acertainment of patients with apallic syndrome in Vienna (translated from German)

Point prevalence study vienna
Apallic syndrome and similar conditions

(Study of the number of patients with a coma lasting longer than 14 days in Vienna's hospitals)

Date of the examination:		Examiner:	

ID:

Hospital:		Department:	Ward:
Number:		Initials:	Date of birth:
Gender:		Previous hospitalization:	
M	F		

Diagnosis:

Cause:	Date of the acute event:
Previous illnesses and risk factors:	Any other information: (ventilation etc.)

	YES	NO
Sedative medication: (Neoroleptics, Benzo.)	☐	☐

Diagnostic criteria for Apallic Syndrome:

	YES	NO
Lack of awareness	☐	☐
Sleep–awake pattern	☐	☐
Random eye movement	☐	☐
Eye tracking	☐	☐
Eye fixation	☐	☐
Eyeballs in diverged position	☐	☐
Motoric primitive patterns	☐	☐
Vegetative reaction	☐	☐
Automatisms (Chew, Suck, Swallow)	☐	☐
Flexion of the UE	☐	☐
Flexion of the LE	☐	☐
Tonus increased on UE	☐	☐
Tonus increased on LE	☐	☐
Reflexes intensified on UE	☐	☐
Reflexes intensified on LE	☐	☐
Pyramidal signs UE	☐	☐
Pyramidal signs LE	☐	☐

Scores:

GCS	GOS	E2CS	Barthel

Figure 1 Questionnaire used for the ascertainment of patients with apallic syndrome in Vienna (translated from German).

Table 1 Key to ICD-9 diagnosis used

348.9	Apallic syndrome
800.7	Open/closed skull fracture with apallic syndrome
801.7	Skull fracture with apallic syndrome
803.7	Other fractures of the skull bones with apallic syndrome
804.7	Multiple fractures with involvement of the skull or of the face with other bones with apallic syndrome
851.5	Cerebral contusion with apallic syndrome
852.5	SAB with apallic syndrome
854.5	Intracranial injury with apallic syndrome

Table 2 Acknowledged replies categorized according to administration (total number of institutions written to = 82)

Vienna General Hospital ($n = 1$)	100%
Hospitals in Vienna Hospital Association ($n = 13$)	85%
UnfallKH (accident and emergency ward), rehab. centres, private hospitals and hospitals affiliated to religious orders in Vienna ($n = 24$)	96%
Geriatric centres in Vienna Hospital Association ($n = 4$)	100%
Private nursing homes in Vienna ($n = 40$)	100%
In total there is a response rate of 96% from Viennese medical institutions and nursing facilities.	

Table 3 Distribution according to age and gender in the total group surveyed

	Age (years)								
Gender	< 30	30–39	40–49	50–59	60–69	70–79	80–89	90–99	Total
Females	2	5	7	8	6	5	2	0	35
Males	5	7	7	9	6	6	2	1	43
Total	7	12	14	17	12	11	4	1	78

As the aim of the study was to record the population-based prevalence of apallic syndrome, four patients of the group with full-stage apallic syndrome ($n = 36$) were excluded as they originally lived in other federal states.

On the study day, there were seven persons, living with their family at home, who were entitled to the highest level of financial support as stated by the administrative authorities in Vienna. According to the criteria used to allocate this provision, it was possible that some of the patients in this group had full-stage apallic syndrome. However, a survey of this group was not possible for data protection reasons.

From this point on, only residents of Vienna ($n = 32$) will be discussed who were diagnosed with full-stage apallic syndrome and who were recorded as inpatients in public or private medical institutions in Vienna on the day of the study.

Full-stage apallic syndrome

Men ($n = 21$) outnumber women ($n = 11$) (Table 4). There were 31 adults and one child. The most frequent age group represented was 50–59 years. Thirteen patients (41%) were being treated in the acute care units of hospitals at the time of the study. Nineteen patients were accommodated in nursing facilities. The duration of stay in acute care units or nursing facility had a wide range: the shortest duration was 14 days and the longest 3260 days (Table 5).

According to aetiological criteria there were two patients in acute units with conditions of a traumatic origin contrary to 11 patients of non-traumatic origin [three cardiac arrests with cardio-pulmonal resuscitation (CPR), two myocardial infarctions, two strokes, one hypovolemic shock, one status asthmaticus, and two cerebral haemorrhages].

In the nursing facilities, similar conditions could be observed. Five patients (26%) were admitted following trauma and 14 (74%) with conditions of a non-traumatic origin [four myocardial infarctions, three cardiac arrests with CPR, three intoxications, two drownings and two cerebral haemorrhages).

Clinical examinations

The clinical examinations were divided into two sections. The first section included clinical signs, whose existence or non-existence are imperative for diagnosing apallic syndrome. In the second section, general neurological symptoms observed in the patients were described.

In the first section, 100% of the patients showed lack of awareness. In 87% of the patients the eye balls were in a diverged position and 56% displayed random eye

Table 4 Age and gender distribution and prevalence (n/100 000) in the group of patients with apallic syndrome (full-stage)

	Age (years)								
Prevalence	< 30	30–39	40–49	50–59	60–69	70–79	80–89	90–99	Total
Gender									
Females (1.4)	1[a]	2	2	4	1	0	1	0	11
Males (2.4)	3	5	2	4	2	3	2	0	21
Age	0.7	1.6	1.0	2.4	3.4	2.4	2.2	0	1.9

[a]Child between 1 and 2 years old.

Table 5 The duration of stay in acute care units or nursing facilities

Days after insult	Number of patients
28	3
112	5
224	3
365	4
>365	17
Total	32

movement. In 100% of the patients the presence of a sleep–wake pattern was observed. No patient in the group displayed eye fixation or the capability of eye tracking. In addition, 84% of the patients displayed the clinical form of vegetative reactions and 75% showed automatisms (chewing, sucking) as described in this context by Gerstenbrand (1977).

The second section of general neurological changes referred to tonus, reflexes and pyramidal signs. Sixty-six per cent of patients displayed flexion in the upper extremities, and 44% in the lower extremities. Increased tonus could be deteceted in the arms of 91% of the patients and in the legs of 87%. In comparison, the reflexes of the upper extremity had increased more often (25%) than those of the lower extremity (19%). Positive pyramidal signs could be detected on the legs of 28% of the patients (Rossollimo Reflex) and on the arms of 19% (Hoffman Reflex).

Scores

The median of the Glasgow Outcome Score was 2. This corresponds to the level of persistent vegetative state. The median of the Glasgow Coma Scale among the examined patients was 8 (range 5–8). The Edinburgh 2 Coma Scale showed a median of 7 points (range 7–9). The median of the Bartel Früh Reha scores was 0.

Prevalence of apallic syndrome in Vienna

At the time of the investigation the population of Vienna was comprised 1 620 170. The use of the exclusive criteria as described above, results in a group of 32 patients with full-stage apallic syndrome. From these data the resulting hospital prevalence is 1.9/100 000 inhabitants.

Discussion

In order to comprehend the problem of apallic syndrome, it is essential to have access to the exact prevalence data. In the literature, the prevalence rates range widely. This seems to depend on differences of the definition of vegetative state as well as on study inclusion criteria (Jennett, 2002). Higashi *et al.* (1977) published the results of their observations on patients with apallic syndrome in a Japanese province. In this work not only patients who had full-stage apallic syndrome were recorded but also its course and the collective remission stages were described. From this work Schönle (1999) calculated a prevalence of 2.5/100 000, although Higashi *et al.* (1977) did not allude to this exact statistic. In another study from Japan, Sato *et al.* (1978) found a prevalence of 2.5/100 000. In European reports (the Netherlands, France) the prevalence rates range between 0.5 and 1.4/100 000 (Minderhoud and Braakman, 1985; Hellema, 1991; Tasseau *et al.*, 1991). Only a very few studies deal with the problem of children in a vegetative state. Ashwal *et al.* (1992,1994) described a prevalence between 2.4 and 2.9/100 000 in children in his studies. Most of the authors did not stratify by age when calculating the prevalence (Jennett, 2002). The MSTF o PVS (1994) reported that the number of adult patients was between 10 000 and 25 000 and of 4000–10 000 children in the USA. These figures were ascertained from the details of the most up-to-date literature of the time.

Considering that the results of our Vienna study resemble a rate of 1.9/100 000, it can be shown that the results produced by other authors point in the same direction.

The criteria described both by Jennet and Plum (1972) and by Gerstenbrand (1967) for recording patients with apallic syndrome proved to be excellent parameters for the differential diagnosis against other forms of diseases. Thus 100% of the patients showed a lack of awareness, 100% fatigue-limited sleep–awake pattern and 87% displayed diverged eye balls. In contrast, signs of remission such as eye fixation and eye tracking are exclusive criteria. None of the 32 patients displayed these features.

A shift in aetiology, from traumatic lesions to non-traumatic origin, could be observed. However, a trend cannot be concluded from this as the number of patients is too small. In the group with non-traumatic aetiology, the largest number of patients in total were those with cardiac diseases (six myocardial infarction, six cardiac arrests), followed by patients with vascular disease (four cerebral haemorrhages, two strokes).

The number of men outweighs that of women in the recorded group. The ratio of men to women is 2:1. For both, the modal average age group recorded was 50–59 years.

From the data in this study, hospital prevalence of apallic syndrome in Vienna is found to be 1.9/100 000 inhabitants. There are several weak points in our study which have to be considered carefully. For example, seven potential patients were in a home environment

and could not be examined because of data protection reasons. This group could represent a number of patients with apallic syndrome, raising a question about the validity of the survey numbers. For that reason, a follow-up of our hospital point prevalence study will take place in November 2003. Another shortcoming may be the reluctance of some of the cooperating hospitals to report patients and to provide access to the patients for clinical follow-up. This may have led to underestimation of the prevalence. We tried to increase the willingness for cooperation by *a priori* informing the heads of the wards and nursing facilities and keeping them informed about the results of the survey. A better coordination between these departements should be developed. If a patient is suspected of apallic syndrome, he will be sent to a department, specialized for assessment. There it will be decided whether the patient is to be sent to a neurological rehabilitation ward or to a nursing facility. This project should optimize the treatment and improve the outcome, because dealing with patients with apallic syndrome is an ethical as well as an economical problem. On the one hand, each patient should get the best treatment for his disease, and on the other, it is necessary to employ economical criteria to keep the capacity of our health system at high standards.

The Viennese Health Authorities have incorporated our findings into its forecast for medical provisions for the year 2003. This plan indicates the number of beds provided for each patient group in acute wards, and rehabilitation centres and nursing facilities.

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