Executive and Cognitive Functioning Pattern Differentiate Between Active and Arrested Normal Pressure Hydrocephalus

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Abstract

The problem of executive and cognitive functions in patients with normal pressure hydrocephalus (NPH) was investigated in the study. Executive and cognitive function parameters were assumed to be among factors that may differentiate the clinical pattern in NPH. Two major indicators of executive functioning, i.e. flexibility and productivity of thinking, were assessed in neuropsychological examination using the Trail Making Test (TMT), Verbal Fluency Test (COWAT), and the Wisconsin Card Sorting Test (WCST). Participants in the study were 18 patients with NPH divided using a set of diagnostic criteria into two subgroups: with idiopathic active hydrocephalus (ACT) or with arrested hydrocephalus (ARR). Executive and cognitive functioning patterns were found to differentiate between the two NPH subgroups. Namely, patients diagnosed with active hydrocephalus (who qualify for shunt implantation surgery) tended to present lower levels of verbal fluency in all semantic categories, which suggests a decreased productivity of thinking. Regarding the patients’ performance on the TMT, no intergroup differences in psychomotor speed and task concentration were found in both parts of the test, A and B. Although this differentiating tendency was quite marked, it turned out to be statistically insignificant. Besides, ACT patients’ performance on the WCST was significantly inferior on two measures: (1) they committed more non-perseverative errors (which indicates their chaotic way of working on the test and the occurrence of random responses) and (2) displayed lower ability of “learning to learn” (which suggests their impaired flexibility of thinking). These aspects of executive and cognitive function, with productivity and flexibility of thinking first and foremost, seem promising as additional prognostic indicators to consider in patient selection for shunt implantation.

ABBREVIATIONS

CSF: Cerebrospinal Fluid; NPH: Normal Pressure Hydrocephalus

INTRODUCTION

The diagnostics and management of the central nervous system (CNS) abnormalities may present a considerable challenge not only for beginners, but also for specialists in the arts of medicine or neuropsychology. One of serious CNS conditions is enlargement of the ventricular system, frequently called hydrocephalus. Its etiology is not quite explained, and clinical presentation including cognitive dysfunctions remains unclear [1]. The problem of the ventricular system pathology is most interesting the more so that despite recent developments in radiological imaging as well as in many other functional methods and tests, hydrocephalus continues to be a major diagnostic and therapeutic problem.

Normal pressure hydrocephalus (NPH) is the brain ventricular abnormality interesting from the perspective of both its origin and clinical pattern. Hydrocephalus results from a suddenly increased intracranial pressure (ICP), frequently without any tangible cause. Such a rise in the CSF pressure generally decreases during the next two days, attaining a relatively stable level within the normal range. However, due to changes in the intracranial parameters the ventricular system remains enlarged [2].

One of the forms of normal pressure hydrocephalus (NPH) is described as “arrested” or “compensated” hydrocephalus, diagnosed in cases where cerebral edema due to a high ICP is appropriately treated and controlled. Hydrocephalus can be stopped or “arrested” when any further rise in the CSF pressure is prevented, its level gradually returns to the normal range and becomes relatively stabilized. While a dilatation of the ventricular system may be evidenced by neuroimaging scans, no changes in cognitive function can be seen, and since neither cognitive impairments, nor neurological deficits are observable, there is no

need for treatment implementation. However, deterioration may occur over time, including not only impaired cognitive function (e.g., clouding of consciousness or language disorders), but also neurological deficits, with increased muscle tension among other symptoms [3].

Clinical practice and numerous studies suggest that arrested hydrocephalus can be activated by an acute rise in the intracranial pressure following an excessive CSF build-up due to its flow blockage or “arrest”. Persons with arrested hydrocephalus may develop over time a set of clinical symptoms characteristic of “active” hydrocephalus; then they qualify for surgical implantation of a shunt device. It is still unclear why such a cerebral decompensation occurs. The phenomenon is the more interesting (and significant) that in many cases the patient’s arrested hydrocephalus will probably never be activated [1].

Many studies conducted so far on normal pressure hydrocephalus (NPH) indicate that symptoms characteristic of this condition comprise cognitive impairments, gait disorders, and urinary incontinence (although the latter symptom has been more recently considered as not necessarily present in the NPH clinical pattern). These data are consistent with the NPH clinical description proposed by Hakim and Adams in 1965 [4,5]. However, current studies [6] conducted worldwide, also in Poland, more and more often indicate that NPH not always presents with all the symptoms, that its clinical pattern of cognitive impairments is not clear-cut, and that patients with this condition do adapt to changes in their intracranial parameters [7-9].

The treatment of choice for symptomatic hydrocephalus consists in the cerebrospinal fluid (CSF) drainage from the ventricles of the brain using a shunting device. Appropriate selection of patients for shunting is a major challenge as the clinical presentation of hydrocephalus is rather ambiguous, cognitive impairments may be elusive, symptom severity varies, and besides, the presence of gait disorders and urinary incontinence is not limited to hydrocephalus, but may be seen also in other comorbid conditions.

Neuropsychological assessment may be a significant part of the diagnostic procedure to identify patients who qualify for shunt implantation. Using tests and tasks sensitive to particular neuropsychological parameters it is possible to assess the patient’s cognition in various domains, including their allocentric and egocentric orientation, awareness of deficits, attention, memory, learning, reasoning, visuospatial abilities, verbal fluency, calculation/ mathematical skills, as well as motor skills and emotional status [2]. Deficits found in some cognitive domains may possibly have a prognostic value in pre-selection for shunting in hydrocephalus patients [7].

Since the ventricular system enlargement may lead to dysfunction of the frontal lobe structures, especially in the prefrontal area, characteristic impairments of executive functioning may be also hypothesized to occur. The focus of this study is on the executive function specificity as an additional prognostic indicator in pre-selection of patients for shunting.

Executive functioning is characterized by its volitional nature, by abilities to plan and perform purposeful and effective action which requires also the ability to monitor the course of behavior and utilize feedback, i.e., to learn from errors.

Executive functions as compared to cognitive processes are much less investigated and analyzed in the literature. In consequence, they are not so well understood and may be variously interpreted by different authors [10], even though executive functioning constitutes a vital aspect of human behavior.

While subtle cognitive deficits may be at first unobservable, executive dysfunctions evidently affect human functioning from the onset. Since they disorganize all the aspects of behavior, executive function disorders cannot pass unnoticed. Impairment of two self-regulation dimensions: productivity (related to spontaneous initiation of activity and striving to attain an intended goal) and flexibility (related to set-shifting ability) can be expected to result in attention management deficits, as set-shifting is necessary for attention regulation. Without appropriate action planning neither conscious intention to act nor monitoring of the course of action are possible [10].

Our preliminary research results presented at the Neurosurgical Conference in Pułtusk [9] allow us to hypothesize that patients with active normal pressure hydrocephalus (NPH) differ from those with arrested NPH in their executive functioning levels, especially as regards mental flexibility and productivity.

### MATERIALS AND METHODS

#### Participants

Participants in the study were 18 in patients diagnosed with NPH due to ventricular enlargement, with no comorbid conditions or expansive processes of the CNS. The patients’ health status enabled them to undergo a complete neuropsychological evaluation. Two groups of patients were distinguished on the grounds of their medical history, dynamics of their symptoms and course of illness, results of the neurological examination, detailed assessment of cognitive functions, and infusion test results (only the latter parameter is presented in what follows).

The first group comprised 6 patients (five women and a man) aged 25-70. The levels of CSF absorptive capacity as assessed by the lumbar infusion test ranged from 12 to 16 mmHg/ml/min, which indicates an abnormal and sustained rise in CSF pressure, or reduced absorptive capacity. All these patients qualified for a shunting device implantation. This group was diagnosed with active hydrocephalus, and denoted in what follows by “ACT”.

In the second group of 12 patients (7 women and 5 men aged 38-69) the results of the lumbar infusion tests ranged from 6 to 8 mmHg/ml/min. Therefore, the patients did not qualify for shunting. They were diagnosed with arrested hydrocephalus and denoted in the following by “ARR” [11].

Age could not be used as a group selection criterion, first and foremost because the population of persons with enlarged ventricles of the brain is most differentiated. It should be noted however, that numerous clinical observations suggest no relationship between NPH patients’ age and the pattern of their cognitive functions.
Assessment instruments and procedure

Three tests were used in the assessment of executive and cognitive function:

1. The Trail Making Test (TMT) from the Halstead-Reitan Neuropsychological Battery, to evaluate the ability to shift attention from one criterion to another [10,12,13].

2. Verbal Fluency Test (Controlled Oral Word Association Test (COWAT), a part of the Halstead-Reitan Neuropsychological Battery. The test measures fluency of word production, assumed to require preserved memory as well as language and executive function [14].

3. The Wisconsin Card Sorting Test (WCST) developed by Grant and Berg, in the Polish adaptation by Jaworowska [12,15]. Parameters scored in the WCST include: (1) Percent perseverative errors (PE), a measure of response rigidity; (2) Percent non-perseverative errors (NPE) indicating inattentive, chaotic task performance; (3) Conceptual level responses (CLR), indicating the ability of category formation and maintenance as well as utilization of new information; (4) Categories completed (CC), a measure of efficacy of thinking (mental productivity); (5) Trials to achieve the first category (TtFC), a measure of concept formation efficacy. (6) Another important parameter in the WCST interpretation is learning efficacy (LE, “learning to learn”), a measure of flexibility of thinking [10,12,15].

The above tests were preceded by a clinical interview and administered in the same order: the Trail Making Test (TMT, Part A and B), Verbal Fluency Test (COWAT), and the Wisconsin Card Sorting Test (WCST). The duration of neuropsychological assessment ranged from 60 to 70 minutes. Participation in the study was voluntary and written informed consent was obtained from each participant.

RESULTS

Since the sample sizes were small and unequal, the chi-square test was conducted which confirmed that the study groups did not differ significantly and could be regarded as equivalent. One-way analysis of variance (ANOVA) was used to analyze intergroup differences in the patients’ performance on all tests in terms of above-described parameters.

The patients’ performance on the Trail Making Test is illustrated in Figure (1) (test completion time) and (Figure 2) (number of errors).

As can be seen in Figure (1), the average performance time on both parts of the TMT did not differentiate between the two groups, although patients with active hydrocephalus (group ACT) tended to take slightly more time to complete the tasks.

On the other hand, the mean number of errors (Figure 2) was higher in the group with arrested hydrocephalus (ARR) the difference was statistically not significant, but the tendency can be clearly seen.

The Verbal Fluency Test scores, (i.e. mean number of words produced in each of the three semantic categories and in the phonemic category), are shown in Figure (3).

Again, while intergroup differences were not statistically significant, there was an unambiguous tendency for group ACT to score lower in each of the verbal fluency categories.

Figure (4-6) illustrate the scores on the Wisconsin Card Sorting Test (WCST). Two parameters: percent perseverative errors (PE) and percent non-perseverative errors (NPE) are presented as percentages in Figure (4).

Percent conceptual level responses (CLR - total number of correct responses occurring in consecutive runs of three or more:}
the total number of trials in the test) is presented as percentages in Figure (5).

Patients with active hydrocephalus (ACT) as compared to those with arrested hydrocephalus (ARR) had more perseverative errors (PE, indicating mental rigidity) and conceptual level responses (CLR, suggesting difficulty with cognitive set maintenance and shifting), but these intergroup differences did not reach the level of statistical significance. However, the opposite is true as regards non-perseverative errors (NPE, indicating an inattentive, chaotic task performance), significantly more often committed by ACT patients. Thus, patients with active hydrocephalus probably present with a more chaotic work style.

WCST average scores for mental productivity and flexibility are shown in Figure (6).

As can be seen, there are no significant intergroup differences regarding either the number of trials to achieve the first category (indicator of productivity) or the number of categories completed (CC), so performance levels in these dimensions were similar in both patient groups. On the other hand, as regards the ability of “learning to learn”, i.e. mental flexibility as measured by the efficacy of creating mental representations of WCST categories, ACT patients scored significantly lower than did those with ARR. Thus, patients with active hydrocephalus turned out to have a significantly lower flexibility of thinking.

**DISCUSSION**

There is no doubt nowadays that enlargement of intracranial fluid spaces in persons with normal CSF pressure does not have to result in global mental deterioration [6]. This opinion was held also by Hakim himself [4], the first person to describe the cluster of symptoms associated with ventricular enlargement. Among symptoms of cognitive disorders he included psychomotor retardation, attentional difficulties, memory impairment and akinetic mutism. According to some authors, certain functions, with language above all, may remain quite intact [16]. Specific cognitive deficits characteristic of normal pressure hydrocephalus that might differentiate this condition from other CNS disorders such as Alzheimer’s disease, are more and more popular as the focus of research [17]. Besides, it seems most important to seek and distinguish such cognitive impairments that might have a prognostic value in selection of NPH patients for shunt implantation. The search is difficult, since NPH patients do not constitute a homogeneous group and their general cognitive functioning is differentiated [8]. Nevertheless, many studies and clinical observations suggest that impairment of visuospatial processes, but above all - marked deficits of verbal learning or new verbal information processing may be particularly sensitive indicators for the differential diagnosis of persons with active hydrocephalus who qualify for shunting surgery [1,7]. Moreover, in many studies patients diagnosed with arrested hydrocephalus (who do not qualify for shunting) turned out to present in the neuropsychological assessment a different profile of deficits than those with active hydrocephalus. Namely, their cognitive functioning was unimpaired, and both verbal learning and visuospatial skills were intact. Some arrested hydrocephalus patients were reported to manifest only selected cognitive deficits, including impaired memorization of new information, lower ability to modify their behavior due to new experiences, or tendency to act based on immediate memory traces [9]. Since such difficulties, typical of executive dysfunction (and particularly of working memory impairment), were observed also in clinical examination, an attempt was made in our earlier study at a more detailed analysis of these deficits [18].

In the present study executive function patterns were analyzed in two groups of NPH patients: with active or arrested hydrocephalus (while the former qualify for shunting, the latter

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![Figure 4](image4.png) **Figure 4** WCST percentages of perseverative errors (PE), non-perseverative errors (NPE) in NPH patients with active (ACT) or arrested (ARR) hydrocephalus.

![Figure 5](image5.png) **Figure 5** WCST percentages of conceptual level responses (CLR) in NPH patients with active (ACT) or arrested (ARR) hydrocephalus.

![Figure 6](image6.png) **Figure 6** WCST – indicators of categories completed (CC) productivity (PR) and flexibility (FL) of thinking in NPH patients with active (ACT) or arrested (ARR) hydrocephalus.
do not). The focus of the study was on thinking and reasoning processes as a part of executive function, and especially on their two dimensions: flexibility and productivity of thinking.

Regarding the patients’ performance on the TMT, no intergroup differences in psychomotor speed and task concentration were found in both parts of the test, A and B. So both groups of patients, with active NPH or arrested NPH, worked at a similar pace and manifested similar attention-focusing abilities. However, the two groups tended to differ in their ability of attention shifting from one criterion to the other (set-shifting) and in their flexibility of thinking. Patients with arrested NPH tended to commit more errors on Part B of the test which suggests that their ability for flexible set shifting was lower than that of active NPH patients. Although this differentiating tendency was quite marked, it turned out to be statistically insignificant. It cannot be excluded that statistical insignificance of these intergroup differences might be due to the small sample size and considerable individual differences between the patients, especially in the ARR group. Such differentiation seems quite obvious, as in many cases the cause of hydrocephalus was unknown, and the length of time between the onset and diagnosis of ventricular enlargement (i.e. the time for the brain’s adaptation to changed intracranial parameters) was not controlled in the sample. Considering the lack of statistically significant intergroup differences in all the three analyzed variables (psychomotor speed, task concentration and attention shifting) the diagnostic sensitivity of the TMT to different types of hydrocephalus seems to be limited.

On the Verbal Fluency Test patients with active hydrocephalus scored lower than did those with arrested NPH, namely, the former recalled and produced fewer words in each of the semantic categories. This result suggests that active NPH patients (who qualify for shunt surgery) as compared to those with arrested NPH (in whom shunting device placement is not recommended) have poorer semantic memory, display lower motivation to perform the task and slower working speed; in consequence, their productivity of thinking is lower. Thus, lower verbal fluency or decreased word finding ability may be regarded as a useful prognostic indicator in selection of patients for shunt implantation surgery.

In the Wisconsin Card Sorting Test (WCST) marked, statistically significant differences between the two groups of NPH patients were found as regards the number of non-perseverative errors (NPE) and “learning to learn” scores. As regards the remaining indicators, no statistically significant intergroup differences were noted even if one group seemed superior to the other. Thus, the two groups under study can be assumed to be similar in terms of rigidity of thinking or mental flexibility, ability to keep in mind the current criterion (cognitive set maintenance), to utilize new information and previous experiences, as well as concept formation skills. On the other hand, evident intergroup differences were found regarding the attentive, organized (non-chaotic) way of functioning and efficacy of creating mental representations of categories, i.e. the ability of “learning to learn” (mental flexibility). Patients with active NPH turned out to be less capable of action planning or designing a plan (especially in tasks where achievement of intermediate goals was required), and of set-shifting from one task to another. They also displayed a more marked rigidity of thinking and tendency to perseveration. The difficulties observed in active NPH patients are a manifestation of thinking impairment, affecting both flexibility (above all) and productivity of thinking processes. The level of cognitive flexibility in patients with normal pressure hydrocephalus may be assumed to be a promising prognostic indicator useful for selection of patients to shunting.

The study results suggest also that executive function deficits may serve as prognostic indicators of enlarged ventricular system activation, to identify patients who qualify for shunt surgery.

Since additional recommendations to shunting device implantation are being continuously sought, including biochemical markers [19] and Doppler ultrasonography [20], the present study results seem useful. Neuropsychological assessment of not only cognitive processes but also of executive function in patients with ventricular enlargement may provide an additional indicator for differentiation between the types of hydrocephalus [21]. This would allow for a more specific diagnosis, and more accurate selection of patients for shunt surgery. However, this issue requires further investigation.

Continuation of research concerning NPH patients seems warranted not only to seek additional neuropsychological recommendations to shunting, but also to investigate causes of this condition. Despite the continually growing body of knowledge about normal pressure hydrocephalus gained from many clinical and experimental studies, identification of causes of hydrocephalus, as well as the diagnostics and management of this condition still remain a major challenge for neurosurgeons, neurologists and neuropsychologists.

CONCLUSION

The number of patients studied may raise doubts about the reliability of studies. However, above results, clinical practice and the fact of publication of our preliminary report suggest that executive functioning pattern can be relevant for the diagnosis of the form of normal pressure hydrocephalus. It is also possible that one of the aspects of executive functions can be an additional criterion of patients’ selection for shunt surgery.

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