Review article

Cognitive disorders in children's hydrocephalus

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Background and objective: Hydrocephalus is defined as an increase of volume of cerebrospinal fluid in the ventricular system of the brain. It develops as a result of cerebrospinal fluid flow disorder due to dysfunctions of absorption or, less frequently, as a result of a rise in its production. Hydrocephalus may lead to various cognitive dysfunctions in children.

Materials and methods: In order to determine cognitive functioning in children with hydrocephalus, the authors reviewed available literature while investigating this subject.

Results: The profile of cognitive disorders in children with hydrocephalus may include a wide spectrum of dysfunctions and the process of neuropsychological assessment may be very demanding. The most frequently described cognitive disorders within children’s hydrocephalus include attention, executive, memory, visual, spatial or linguistic dysfunctions, as well as behavioral problems.

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1. Introduction

Hydrocephalus (from Greek: hydro – water; cephalus – head) is frequently defined as an increase of volume of cerebrospinal fluid in the ventricular system of the brain and it can be caused by various factors. However, there is no clear definition of this disorder, mainly due to the multiple factors resulting in hydrocephalus and various research approaches [1]. Hydrocephalus is accompanied by increased brain ventricular size and, if the cranial sutures are not closed yet, also the patient’s head circumference can increase [2]. It is estimated that this condition occurs in 0.8 cases in each 1000 live births [3].

Hydrocephalus develops as a result of cerebrospinal fluid (CSF) flow disorder due to the dysfunctions of absorption or, less frequently, as a result of a rise in its production [4]. The cerebrospinal fluid is formed in the choroid plexus, mainly in the lateral brain ventricles. It is a colorless fluid consisting mostly of water. It chiefly acts as a buffer protecting the brain and medulla oblongata from the results of head injuries among others. It also helps to maintain constant pressure within the skull and reduces the actual brain weight [5]. The influence of an increased ventricular system on developing brain is a really complex issue, as it results in increase of intracranial pressure and the displacement of the neighboring brain structures [2]. In patients with hydrocephalus and in the experimental animal models the following changes in the brain structures were observed: disfigurements or decomposition of the cerebral blood vessels; pressure against the caudate nucleus; extension, narrowing and shifting of the...
corpus callosum; also, the axons and myelin sheaths within the white matter are also affected by the lesions, especially in the paraventricular areas, and the neurons within the brain cortex [4].

Hydrocephalus is traditionally classified as congenital and acquired or communicating and non-communicating. The latter division was created as a result of early studies of hydrocephalus when the non-invasive imaging techniques were not known yet. Dandy and Brackfan, conducting their research on hydrocephalus in the 1920s, had at their disposal merely the lumbar puncture, ventricular puncture and radiological images. Therefore, they conducted a study consisting of a puncture of a brain ventricle and introducing a pigment into it, after which they made a lumbar puncture. If the pigment was present during the lumbar puncture, hydrocephalus was classified as communicating, if not—it was classified as non-communicating (obturative). In the 1960s a hypothesis was formulated maintaining that at the background of each type of hydrocephalus there is an obstacle in the flow of the cerebrospinal fluid, thus the division into communicating and non-communicating hydrocephalus seems illegitimate [1].

1.1. Causes of hydrocephalus

Amongst the most frequent causes of hydrocephalus are intraventricular hemorrhages, rachischisis, aqueduct stenosis, meningitis, Arnold–Chiari malformation, brain tumors and the Dandy Walker syndrome [6].

Rachischisis occurs as a result of neural tube closure failure. This defect can develop as a result of a non-formation of the back part of the vertebral arches (spina bifida occulta), meningocele or myelomeningocele. The number of children with rachischisis has considerably decreased since the time of folic acid supplementation in pregnant women, and yet myelomeningocele still remains the most frequent cause of congenital hydrocephalus [7]. More than 80% of children with this defect also have the type II Arnold–Chiari malformation, that is the deformation of the brain stem and cerebellum which blocks the flow of the cerebrospinal fluid, as a result of which hydrocephalus occurs [8].

Intraventricular hemorrhages as a consequence of premature birth and low birth weight are the most frequent perinatal causes of hydrocephalus. Majority of researchers believe that hemorrhages of this type occur in about 40% newborn babies with the birth weight below 1500 g, and in many cases the brain ventricles experience a post-hemorrhagic extension [9]. The hemorrhage leads to destruction of the brain structures and can result in the formation of porencephalic cysts with the accompanying neurological deficits which depend mainly on the hemorrhage and cysts location and size [2].

Before the introduction of the treatment consisting of the implantation of the pressure valve disposing of the excess CSF outside the cranial cavity, the death rate in children with hydrocephalus was high and amounted to 45–53% [10]. Only 38% of the children who survived managed to achieve a normal intelligence level [11], and 17% of children were still dependent on their guardians. Another 3% of children achieved independence but they were not capable of employment in their adult lives. When pressure valves were introduced the death rate dropped to about 15%. What is interesting, after the introduction of the new treatment, 27% of the persons, that is 10% more than before introducing the valves, are fully dependent, which may be caused by the decrease in the death rate of the children severely burdened with the disease, who would not be able to survive without surgical treatment. The number of people who are independent but incapable of employment rose to 16%. On the other hand, the percentage of people capable of employment was doubled, with 20% before the valve surgeries to about 42% after the revolution in the treatment of hydrocephalus [10]. Clinical and experimental research indicate positive effects of introducing valves such as the reduction of intracranial pressure, reduction of pathologically extended brain ventricles and improved cognitive functioning [2]. However, the treatment is not flawless. The occurrence of infections soon after the surgery is estimated at the level of 5–8% and it increases to 15–30% depending on the time that has passed since the surgery. As a result of infections and the need to adjust the valves, their corrections (revisions) are used [12].

There are numerous factors causing hydrocephalus in children. That is why the patients’ cognitive profile is not clear-cut and can vary depending on the causes, type, age of the disease onset, low birth weight, treatment with valves or its lack, age at which the valve was implanted, number of valve corrections and the occurrence of complications such as infections. That is why the author has made an attempt to clarify the nature of cognitive disorders, which most frequently occur in the children with this disorder.

2. Intelligence

Acquired and congenital hydrocephalus can affect both the level of intellectual functioning of children and its profile. It appears from the research that since the introduction of pressure valves about 1/3 of the children have achieved results below the norm in the tests measuring intelligence (IQ equal to or lower than 69), another 1/3 of the results can be placed within the range where the results (IQ of 70–84) are low or very low. On the other hand, the remaining persons achieve the IQ results above 85. At the same time various types of cognitive disorders and problems with learning can also be observed in the last group of patients mentioned [13]. It is commonly assumed that children with hydrocephalus do better in verbal rather than in non-verbal tests [7,14–17], although not all researchers agree with this [18,19]. Despite the divergences in researches concerning linguistic intelligence measured in IQ tests, the children with myelomeningocele and hydrocephalus, in spite of no significant reduction of general linguistic skills, can have problems with pragmatics and reading comprehension [20–22].

2.1. Executive function and attention

Executive functions are skills that enable one to manifest behaviors that are independent and focused on achieving a particular aim [23]. This happens owing to the application of strategic thinking or planning and the ability to switch
attention as well as controlling the correctness of performing given actions. In this area, severe disorders in children with hydrocephalus were detected in the scope of planning and visual and spatial organization. Children with hydrocephalus achieved results in range of about 1 standard deviation from the norm lower, while healthy children achieved results that were close to 1 standard deviation from the norm higher. Also the phonemic verbal fluency is significantly lowered [8]. The results of the tests aimed at the measurement of executive functions (such as, e.g. Wisconsin Card Sorting Test, Tower of London) appear to be lowered in children with hydrocephalus after the treatment with a valve. However, due to the lack of reduced direct indicators of executive functions disorder, it is suggested that the lower results achieved in these tests can be interpreted as attention system dysfunctions [24]. The results obtained by Vachha and Adams additionally indicate deficits in controlling the selected strategies of learning verbal material. In this study the children with hydrocephalus did not take into account the criterion of the word weight in the test designed and tried to remember each of them, while the children in the control group remembered as many words of higher weight as possible, which indicated their selection of a more effective strategy [25].

The children with hydrocephalus that occurred as a result of rachischisis can have problems performing test of the “paper-pencil” type, such as for instance WISC-III subtests, aimed at the examination of attention functions. They mainly indicate deficits in the scope of resistance to distractors in comparison to the children with rachischisis and without hydrocephalus. However, such differences could not be detected with the application of computerized tests, examining the durability and focusing of attention and impulsiveness or also in the case of applying The Stroop Test for research, that checked the susceptibility to distraction [26]. In addition, children with hydrocephalus seem to be also more susceptible to auditory distraction in the course of problem solving. They also find it more difficult to transfer attention than to focus and maintain attention on a given object [6].

2.2. Memory

Studies focusing on memory processes in children with hydrocephalus have not brought any clear-cut results so far. While some researchers do find the presence of memory disorders in the studies conducted by them [15,27], others are of the opposite opinion [14]. The studies indicating the occurrence of such disorders stress that the disorders occur within the retrieval of the verbal contents in persons with standard IQ [27], and also stress the deficits in the scope of visual memory but without perception disorders in the Benton Test [15]. In turn, another study proves the occurrence of disorders in short-term and long-term memory, both visual-spatial and verbal ones, in comparison to the control group [8]. Ann Scott and colleagues suggest that children with hydrocephalus, regardless of its etiology, manifest memory deficits [28], while children with hydrocephalus combined with myelomeningocele prove to have a lower memory span in comparison to the control group [25]. Also the explicit memory, defined as conscious memory of past events and experiences, becomes handicapped [29].

2.3. Visual and spatial functions

Reduced effectiveness of performing complex visual and spatial tasks is a frequent issue encountered in children with hydrocephalus [14,15,27]. It is suggested that this can be caused by dysfunction of the posterior areas of the brain. Also, deficits have been noted in the scope of the tasks which should be performed with both hands, which in turn can be related to deformations within the cerebellum [7]. In one of the studies parents were asked to define the frequency of the occurrence of vision/recognition disorders in their children undergoing treatment due to hydrocephalus. In this study the answers of the parents of 52 children with hydrocephalus and 192 children from the control group were taken into account. The results indicated the occurrence of vision disorders of the cognitive background in 27 children with hydrocephalus, and in 16 of them more than one type of disorder were observed. Those children had problems in the scope of: recognizing shapes (23%), simultaneous perception (21%), perceiving movement (14%), recognizing colors (14%), topographic orientation (5 children) and with recognizing faces (3 children) [30]. Other authors [31,32] also wrote about disorders in recognizing shapes in children with hydrocephalus (agnosia). Another researchers indicate the relatively good functioning of the children with hydrocephalus caused by rachischisis and myelomeningocele in the scope of performing tasks checking recognition of objects. The children examined by them, however, had problems with matching objects according to their length. Skills such as separating one type of objects form the other ones, recognizing objects presented under a different angle, access to the representation of well-known objects and naming them remained undisturbed [33].

2.4. Language disorders

In the case of hydrocephalus one most frequently talks about disorders typically related to the right brain hemisphere due to the well documented visual and spatial deficits visible in the studies of intelligence. However, linguistic disorders are also observed, especially within the discourse and pragmatics. Despite the lack of global dysfunctions in the scope of speech in children with hydrocephalus, their speech is not devoid of disorders in the particular linguistic domains [2]. In comparison to healthy children, they significantly underachieve in the scope of understanding words representing earlier learned notions and in the tasks checking the ability to use speech in social situations [22]. On the other hand, in the course of telling stories sick children tend to omit semantic structural relations between the story elements [6]. Some researchers suggested the occurrence of a “cocktail party syndrome” in patients with hydrocephalus, characterized by loquacity, maintaining the correct articulation and fairly rich vocabulary. At the same time the content of the utterance was defined as poor in meaning. This syndrome also consisted of perseveration and reduction of the distance to the interlocutor, which could remind of the behavior of a person with the frontal syndrome. The cocktail party syndrome was considered as especially prominent in patients with hydrocephalus originated as a result of rachischisis, and its occurrence was estimated at 28–41% in this group of patients [34]. However, it is believed
nowadays that the cocktail party syndrome is mainly a manifestation of reduced intellectual skills [8].

2.5. Behavioral disorders

There are relatively few reports concerning behavioral disorders in children with hydrocephalus. Still, before the treatment with pressure valves was introduced, Hagberg and Sjogren suggested that nearly 2/3 of children with hydrocephalus, with or without the confirmed mental handicap, had various problems with their behavior [35]. In turn, another study indicates that problems with behavior, particularly those related to attention disorder and hyperactivity are chiefly characteristic of the children with hydrocephalus and mental handicap [36]. In the opinion of the parents of 67% of the children with hydrocephalus, they have various behavioral problems. On the other hand, teachers indicate this type of disorder in 39% of the sick children, and the opinions of the parents and teachers are closest in respect of hyperexcitability. It is worth pointing one’s attention to divergences in the parents’ estimation of behavioral disorder in relation to the intellectual functioning. Among the children with the IQ below 70, about 90% had behavioral problems in comparison to 50% of the children with the IQ above 69 and 12% with the IQ above 85. Teachers estimated that about 2/3 of the children with the IQ below 70 and 1/5 of the children with the IQ above 70 had behavioral problems. In the same study it was found that 13% of the examined children manifested autistic features which strongly correlated to learning problems [37]. In the population study autism and autistic spectrum disorders were detected in 25% of the children with hydrocephalus. However, the researchers did not take into account the children with myelomeningocele and those prematurely born. The occurrence of additional diseases, such as epilepsy or cerebral palsy, was more common in this study among the children manifesting autistic features [38]. The spread of hydrocephalus in children with autistic spectrum disorder (ASD) in Finland is estimated at 3% [39], and 2% in England [40]. This result is much higher than 1 in 1000 in the general population [3].

3. Functioning of the children with hydrocephalus in adult life

Despite the introduction of early treatment of children with hydrocephalus, the disease can affect their functioning in adult life. In one of the studies the researchers decided to examine the level of intelligence in persons aged 30-41, who had been treated for their hydrocephalus in childhood. That was not a numerous group – the study included 25 patients. Three people had their IQ below the norm, 75% within the norm, and three persons obtained the results above the average [41]. In another study, in which 456 patients with hydrocephalus participated, primarily examined at the age of 55.6 months, they were examined again when they were about 24 years. In the case of 17.8% of the persons examined, the last revision of the pressure valve was carried out after they turned 20, and 22 of those people had never had to undergo a repeated surgery. 13 patients examined in their childhood died before the follow-up, and in the case of five persons their death occurred for reasons related to the valve. Among the people examined 41.4% participated in a standard, complete learning curriculum, and 3.3% completed education at an ordinary school prematurely. About 30% of the patients required education at special education schools, and the next 15% of the people examined required a certain extent of assistance at an ordinary school. 26 persons did not participate in any form of school education. The issues related to the employment of people with hydrocephalus were also examined. About 25% of them had a normal job, 17.7% obtained employment in sheltered conditions, and 23.9% were not employed. The unemployment rate among the people whose health allowed them to work amounted to 24.8% in comparison to 16.3% among the healthy persons up to the age of 25 in a given region of the country. The most frequent issues related to the functioning of the patients included motor problems (46.5%), 13% of the examined required assistance with walking, and 13% moved on a wheelchair. Cognitive disorders were equally frequent and occurred in 47.6% of the patients [42]. Frequent problems of adults with hydrocephalus acquired in childhood also include chronic headaches, occurring even in 40% of the patients [43] and depression in as many as 45% of the patients [44].

In one of the studies particular attention was drawn to the quality of life of adults without mental handicap (IQ above 70), treated in their childhood due to hydrocephalus without rachischisis. No significant differences were found between the study group and the control group, and thus no global deterioration of the life quality among adults treated in their childhood for their hydrocephalus. On the other hand, persons with co-existing epilepsy and/or cerebral palsy had worse results than both the control group and persons with hydrocephalus not accompanied by other diseases. Majority of the study participants differed from the control group in respect of the evaluation of their intellectual efficiency/memory and the skills significant for their every-day functioning [45].

4. Tests applied in the neuropsychological evaluation of children with hydrocephalus

Hydrocephalus among children is the cause of numerous cognitive problems. However, so far no test or test battery designed specifically for their needs, checking all the cognitive domains that can be weakened due to the disease, have been created. Among the most frequently applied tools in the case of the cognitive evaluation of children with hydrocephalus one can enumerate: Trail Making Test versions A and B, The Stroop Test, Digit Span, RAVLT (Rey Auditory-Verbal Learning Test) and Verbal Fluency Test [8]. The level of intelligence in children with hydrocephalus is most frequently measured with the WISC-III (Wechsler Intelligence Scale for Children-Third Edition) [18] and WISC-IV (Wechsler Intelligence Scale for Children-Fourth Edition) [46] tests. An attempt has also been made [47] to translate some of these tests into computer versions in order to create a uniform battery of tests for the evaluation of people with hydrocephalus. Also a scale measuring the quality of life
related to health in children has been elaborated (HRQL—health-related quality of life), that is Hydrocephalus Outcome Questionnaire (HOQ) [48]. Due to the lack of definite tools specifically designed for the needs of the children with hydrocephalus, the table below presents certain tests applied for the neuropsychological evaluation of those persons along with the studies in which they were applied (Table 1).

5. Summary

The occurrence of hydrocephalus in children can be of varying etiology. Also comorbidities and individual effectiveness of treatment with pressure valves are not insignificant. The study results are not clear-cut either in respect of patients’ cognitive profiles. However hydrocephalus does undoubtedly affect the cognitive functioning of many children and frequently has consequences in adult life. The most frequently described cognitive disorders within this disease can be executive, attention, or linguistic dysfunctions, as well as behavioral problems related to memory and visual and spatial disorders. Work on improving the methods of treatment and on creating a uniform battery of tests for the cognitive evaluation of persons with hydrocephalus is being continued.

Conflict of interest

None declared.

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Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

REFERENCES


