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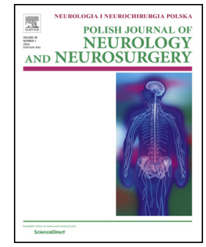

Review article

Cognitive disorders in children's hydrocephalus

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ABSTRACT

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 the increase of 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.

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

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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 1960 s 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