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ORIGINAL PAPER

Very long-term follow-up of cognitive function in adults treated in infancy for hydrocephalus

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Abstract
Purpose The aim was to investigate the very long-term cognitive outcome in adults who had been shunt treated for hydrocephalus during their first year of life.
Methods In a population-based series of 72 children born in 1967–1978 and shunt treated for infantile hydrocephalus, 43 were found to have a normal cognitive function when assessed at 6–17 years of age. Twenty-five of them agreed to participate in a follow-up study of cognition at a mean age of 35 years (range, 30–41 years). The Wechsler Adult Intelligence Scale (WAIS-III) was used.
Results The median full-scale IQ was 101 (range, 83–120), median verbal IQ was 104 (81–115) and performance IQ was 99 (80–127). The corresponding IQs in childhood in the 16 subjects who had been tested previously with the WISC were 101 (84–124), 108 (86–135), and 101 (73–124). Specific cognitive deficits were found for working memory and processing speed. Shunt complications did not affect IQ.
Conclusion This very long-term follow-up study of normally gifted children with hydrocephalus revealed that, as adults, they still had preserved cognitive functions despite recurrent shunt dysfunction. The results are encouraging and represent a tribute to neurosurgical intervention. Continued follow-up studies are needed since the etiological panorama and treatment procedures of children with hydrocephalus are changing over time.

Keywords Infantile hydrocephalus · Cognition · Prognosis · Long-term follow-up

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Hydrocephalus implies that the cerebrospinal fluid (CSF) exerts increased pressure on the brain parenchyma, especially against the immediate periventricular white matter. There are several underlying etiologies behind the disturbance in CSF circulation and different kinds of malformation, together with post-infectious and post-hemorrhagic conditions, are the most common. The general outcome in children with hydrocephalus is largely determined by the origin of hydrocephalus, but, regardless of etiology, the increase in intracranial pressure-causing hydrocephalus leaves the children at risk of developing cognitive malfunction [2, 6, 9, 11].

Since the introduction of modern shunt treatment in the 1960s, the prognosis has improved considerably, especially in children with uncomplicated hydrocephalus, i.e. without concomitant parenchymal involvement. So, for about 40 years, children with infantile hydrocephalus have been treated surgically, but there are very few reports on the long-term outcome regarding cognitive functions.

About one third of children with hydrocephalus have been found to have a normal IQ, another third to have a low-average IQ (IQ, 70–90) and one third to have learning disabilities (IQ<70) [9]. The cognitive profile in children with infantile hydrocephalus commonly displays a characteristic profile, with a verbal IQ that is significantly higher than the performance IQ [2, 3, 6, 9]. The specific cognitive

impairments reported in these children include memory problems and difficulties associated with planning and organization [5, 10, 13, 16]. In 1999, Hommet et al. [7] studied whether the nonverbal learning disabilities often described in children with shunted hydrocephalus were also present in adults. However, they found no discrepancies between performance IQ and verbal IQ in eight shunt-treated individuals, nor was there a discrepancy between verbal and visuo-spatial memory. Iddon et al. [8] studied the neuropsychological profile in young adults with spina bifida, with and without hydrocephalus with an IQ of at least 90. They found that tests measuring the executive functions in particular produced significantly poorer results in those subjects with hydrocephalus. The majority of the hydrocephalus patients performed in the low-average range or below in tests of verbal learning ability, delayed verbal recall, spatial working memory, attentional set shifting, and psychomotor speed on complex tasks involving sequencing. These studies were, however, neither population based nor longitudinal.

The primary aim of the present study was to investigate the cognitive abilities and profiles in a population-based series of adults who had been treated for hydrocephalus during their first year of life and who had been assessed at 6–17 years of age and found to have a normal IQ. A secondary aim was to analyze the impact of episodes of shunt dysfunctions that had occurred during their lifetime.

Material and methods

From a population-based series of children with infantile hydrocephalus in the south-western Swedish health care region, covering the birth years 1967–1982, those born in 1967–1978 participated in a clinical follow-up study in the early 1980s [3, 4]. The group comprised 72 children. An overall cognitive function within the normal range was found in 43 of them, formally tested in 23 children and clinically estimated in 20 children. Of these 43 individuals, 25 (16 men and nine women) with a mean age of 35 years (range, 30–41 years), agreed to participate in this follow-up study. Of the 18 individuals who were unable to take part in the study (13 men and five women), with a mean age of 35 years (range, 30–40 years), one had died, one did not respond to the invitation, three could not be traced and 13 individuals declined participation. The non-participating individuals did not differ from the study group in terms of age, gender, or etiology. Of the 25 participating individuals, six had an unknown prenatal origin of the hydrocephalus, seven had congenital malformations, six a perinatal cause, i.e. an infection or a cerebral hemorrhage, while the remaining six had pre- or perinatal risk factors but no obvious cause. Four participants had epilepsy and four had milder forms of cerebral palsy. Seventeen individuals (68%) in the study group had been assessed as children with the Wechsler Intelligence Scale for Children (WISC) [14].

In the present follow-up study, the Wechsler Adult Intelligence Scale (WAIS-III) [15] was used to assess intelligence in all 25 participants. The latest revised version comprises 14 subtests and provides scores for verbal IQ, performance IQ, and full-scale IQ, along with four secondary indices, namely Verbal Comprehension Index, Working Memory Index, Perceptual Organization Index (POI), and Processing Speed Index (PSI), Fig. 1. These indices are more specified descriptions of sub-functions of verbal and performance abilities. The Verbal Comprehension Index is part of the verbal IQ and measures the ability to understand and define words, the ability to use language for reasoning and to find logical verbal relationships. It also includes a test of general information. The second sub-index of Verbal IQ, the Working Memory Index, comprises three working memory tests; Digit span, i.e. repeating as many digits as possible forwards and backwards, Arithmetic, which contains mental arithmetic tasks, and letter–number sequencing. In the latter test, the subject listens to series of mixed letters and numbers and is asked to repeat them in an organized way. Working memory is defined as a system for temporarily storing and managing the information required to carry out complex cognitive tasks such as learning, reasoning, and comprehension. Working memory is involved in the selection, initiation, and termination of information-processing functions, such as encoding, storing, and retrieving data. In daily life, working memory problems result, for example, in difficulty retrieving more than one or a few instructions, keeping instructions in mind when working with a task or learning and remembering lists.

The sub-indices of Performance IQ are the Perceptual Organization Index and the Processing Speed Index. The POI assesses the ability to perceive and interpret pictorial information, as well as visuo-spatial ability and planning. The PSI contains two time-limited paper-and-pencil tasks; Digit Symbol Coding and Symbol Search. Both are tests of attention, speed and, to some extent, fine motor function. Working memory, attention, as well as planning and processing speed is important for executive functioning. The subtests of the WAIS-III therefore provide a more precise picture of the dimensions of the verbal and performance IQ that are impaired and how they can affect the daily life of the person.

The WAIS-III defines an IQ below 70 as exceptionally low, 70–89 as low, 80–89 as lower normal, 90–109 normal zone, 110–119 upper normal, 120–129 high, and an IQ above 130 as exceptionally high.

One of the 25 participants had a visual impairment and was therefore not able to perform the visual tasks and, as a

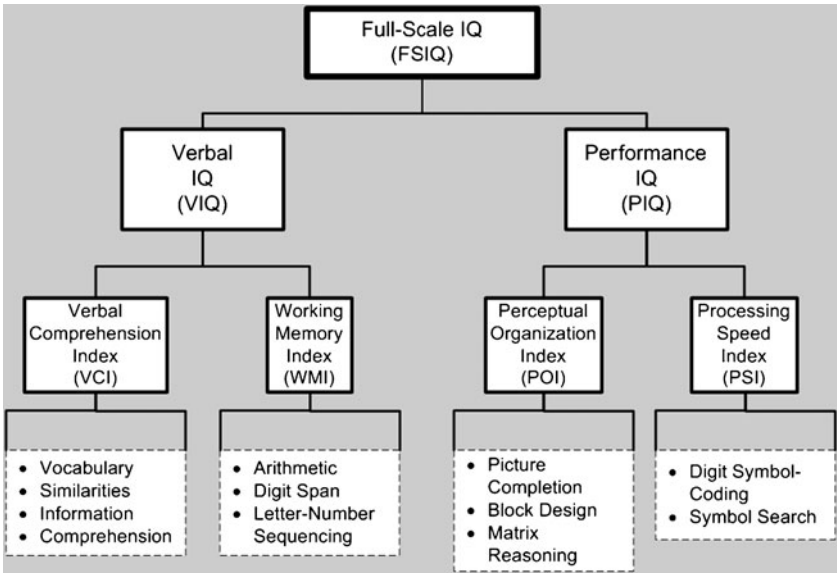

Fig. 1 Subtests and indices in the WAIS-III, the Wechsler Adult Intelligence Scale

result, 24 individuals accomplished the complete test. Episodes of shunt dysfunction were recorded. The assessments were all made by one of the authors.

All the participants were interviewed regarding health, school education, and employment; the results are reported separately.

Ethics

The study was approved by the Research Ethics Committee at Gothenburg University.

Results

Intelligence

The median IQ of the study group was 101, i.e. a result within the normal zone. The median verbal IQ was 104 (n=25; range, 81–115) and the performance IQ 99 (n=24; range, 80–127). Three of the 24 fully tested individuals had a full-scale IQ in the lower normal area (median, 86; range, 83–87), 18 (75%) in the normal (median, 101; range, 90–108), two obtained results in the upper normal zone (both 113) and one had an IQ of 120. In a reference population, 50% are in the normal zone and 16% in the lower and upper normal zone respectively. The corresponding distribution in the study group was 75%, 13%, and 13%, respectively, Fig. 2.

There was no statistically significant difference between verbal and performance IQ in the whole group. However, eight individuals had a significantly better verbal IQ and eight a significantly higher performance IQ.

The results for the four indices were more widely distributed. On the Verbal Comprehension Index, 23 (92%) obtained a result in the normal area or over and two in the lower normal zone. Nineteen (76%) had a normal result or better on the Working Memory Index; three had low normal results and three low results. On the Perceptual Organization Index, four obtained a result within the lower normal IQ zone and 20 (83%) obtained normal results or over. Six had a low result on the Processing Speed Index, eight in the lower normal zone and ten (42%) normal or over, Fig. 3.

Longitudinal follow-up

When comparing the full-scale IQ of the 17 subjects who were assessed with the WISC as children, it was revealed that the results for the 16 subjects who were fully assessed on the WAIS-III full-scale IQ were fairly similar, Fig. 4.

When assessed as children, their median IQ was 101 (range, 84–124) and, about 25 years later, their median IQ

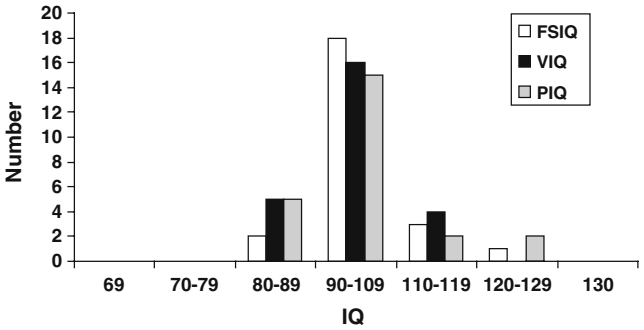

Fig. 2 Full-scale IQ (n=24), verbal IQ (n=25) and performance IQ (n=24) in adults with hydrocephalus assessed with the WAIS-III

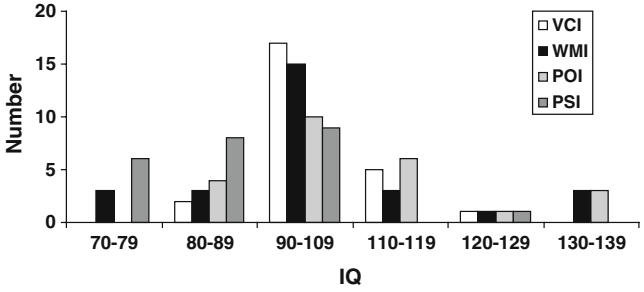

Fig. 3 IQ results Verbal Comprehension Index (n=25), Working Memory Index (n=25), Perceptual Organization Index (n=25), Processing Speed Index (n=24) in adults with hydrocephalus assessed with the WAIS-III

as adults was 101 (range, 83–120). The median verbal IQ had previously been 108 (range, 86–135) and the performance IQ 101 (range, 73–124). As adults, their corresponding IQs were 102 (range, 81–115) and 99 (range, 80–127), respectively.

Shunt complications had occurred in 22 of the 25 subjects, in 19 of them more than twice and 13 had experienced shunt complications during the last 10-year period. Fifteen of the 17 who underwent a complete IQ test both as children and as adults had had problems with the shunt, eight of them during the last 10 years. Neither those who had experienced early shunt complications nor those requiring shunt revisions during the last 10 years differed significantly in their full-scale IQ when comparing their results as children with those when they were adults, i.e. there were no evidences that the shunt complications had had any impact on their IQ.

Discussion

The 25 adults who were treated for hydrocephalus during their first year of life and assessed as normal cognitively at the age of 6–17 years were shown to have spared cognitive functions when re-assessed at the age of 30–40 years. Significant differences between verbal and performance IQs were more common among those with hydrocephalus, where two of three had such differences compared with one in five in a normal population. However, a superior performance IQ was as common as a better verbal IQ. The individuals with hydrocephalus therefore had a more uneven cognitive profile than that in the reference population. This became even more evident when analyzing the results of the four index measurements. The majority had results far below average or lower normal results on the processing speed index, i.e. they were very slow in time-limited paper-and-pencil tasks. A quarter of the participants had results below normal or in the low normal zone on the working memory index. Problems with working memory and processing speed have been described in children with hydrocephalus [1, 10]. Both working memory and processing speed are important for learning at school, independent of general cognitive ability. These cognitive abilities are also strongly linked to executive functions. Similar findings were reported by Iddon et al. [8].

When interviewed, the participants declared that they were aware of always being slow writers but now, as adults, they used computers instead of writing with a pencil. The six subjects who had working memory problems compensated for this by using the calendar function in their mobile phones, for example, or drawing up lists of things to do.

Our study group of adults seemed to cope with their specific difficulties in daily life. These specific cognitive deficits and their functional impact on daily life have to be considered early in children with hydrocephalus.

A risk that two or more shunt complications could lead to serious impairments, seizures, or affected mental function has been reported [6, 12]. However, our results did not accord with these findings. Instead, we found that shunt complications did not affect the IQ results, regardless whether they had occurred early in life or during the last 10 years.

This follow-up study is unique as it is population based, very long-term, and partly longitudinal, with subjects

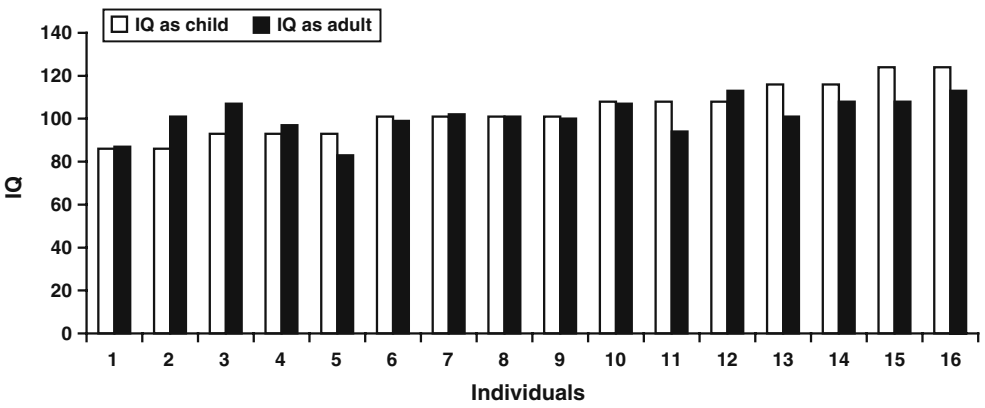

Fig. 4 Full-scale IQ of 16 individuals assessed with the WISC in childhood compared with their IQ as adults on the WAIS-III

assessed both as children or adolescents and as adults. The results partly confirm findings from the two most similar studies including adults with hydrocephalus, demonstrating that verbal IQ was not significantly superior to performance IQ [6] and that adults treated in infancy for hydrocephalus display difficulties in abilities needed for executive functions [8].

The population-based study, from which the normally gifted subjects with hydrocephalus were recruited, also included 30% children with an IQ of less than 70. They were not included in this follow-up study as their underlying brain pathology and associated neurological impairments were more decisive for the prognosis than the hydrocephalus per se, which was the focus of this study. In a more recent population-based study of children with hydrocephalus, the corresponding percentage of children with an IQ of less than 70 was 42% and the median IQ for the whole group was 88, which was in the lower normal range [9]. One explanation of the poorer outcome in the later study could be that these children represent a different etiological background than the surviving adults in the present study. In the 1970s, infants born extremely preterm did not survive. Today, this group constitutes a considerable percentage of children with hydrocephalus, caused by hemorrhages in the immature brain. In the present study, all the participants but two had been born at full term and few had additional impairments such as epilepsy or cerebral palsy.

It is notable that this first generation of children with successfully shunt-treated hydrocephalus has experienced no cognitive deterioration during an almost 40-year life span, despite the fact that the majority have had shunt problems and often required repeated shunt revisions. Despite the often occurring uneven cognitive profiles, with specific memory and processing speed deficits, most individuals had developed compensatory and functional strategies to cope with daily life.

Conclusion

This study is the first long-term, longitudinal follow-up of a population-based group of adults who had been shunt treated for hydrocephalus during infancy. The results were advantageous, but it will be necessary to continue to follow-up children with hydrocephalus who are born today, as they may represent a different etiological panorama than those born 40 years ago.

Conflicts of interest The authors declare that they have no conflict of interest.

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