

Intellectual Functioning in Children with Early Shunted Posthemorrhagic Hydrocephalus

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Key Words

Hydrocephalus, cognitive outcome • Intellectual functioning • Performance intelligence quotient • Verbal intelligence quotient • Nonsiphoning valve

Abstract

Objectives: Early intervention for cognitive impairments seen in children with hydrocephalus is key to successful developmental outcome. Thus, the examination of the cognitive and behavioral functioning of children with hydrocephalus is important, given increasing survival rates and the potential to implement successful cognitive interventions. The current literature suggests that intellectual impairment is common in hydrocephalus patients, yet the findings vary concerning the extent and specifics of the cognitive deficits seen in these children. To better understand the pattern of cognitive impairment observed in children with shunted hydrocephalus, the present study examined a cohort of children with hydrocephalus uniformly shunted with a nonsiphoning valve in the first year of life. **Methods:** Forty-one children and adolescents with a history of congenital hydrocephalus, who were shunted with nonsiphoning valves in the first year of life and are currently achieving academically within 1 year of appropriate school grade for their chronological age, were compared to 16 nonaffected age- and education-level-matched controls. The subjects completed a comprehensive neuropsychological battery that included the Wechsler Intelligence Scale for Children, Third Edition,

(WISC-III) as a measure of general intellectual functioning. Performances across the WISC-III were compared between the 2 groups. **Results:** The data were normally distributed for both groups. The children shunted for hydrocephalus scored approximately 1 standard deviation lower than the controls on the measures of general intellectual functioning (shunted group mean WISC-III full-scale intelligence quotient = 83.8 vs. control mean full-scale intelligence quotient = 102.9, $p \leq 0.001$), verbal intellectual skill development [shunted group mean WISC-III verbal IQ (VIQ) = 86.6 vs. control mean VIQ = 107.1, $p \leq 0.001$] and visuospatial and perceptual-organizational skill development [shunted group mean WISC-III performance IQ (PIQ) = 83.6 vs. control mean PIQ = 98.2; $p \leq 0.006$]. History of shunt revisions, infections, prematurity, seizures and hemorrhage was not significantly correlated with intellectual functioning. While VIQ and PIQ have been reported as discrepant in many previous studies of cognitive functioning after shunting due to hydrocephalus, the current results revealed statistically similar VIQ/PIQ performance (Spearman's ρ rank order $r_s = 0.685$, $p \leq 0.001$). **Conclusions:** Intellectual functioning in this selected group of children with hydrocephalus is normally distributed, yet significantly below that of nonaffected peers. Previously reported discrepancies between VIQ and PIQ were not evident in our study. This finding may be accounted for by the selectivity of our study population, implying a differential effect of etiology and treatment on intellectual function outcome in hydrocephalic children.

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Introduction

Research examining the intellectual functioning of children with a history of hydrocephalus has been mixed: some studies find generalized decline in intellect [i.e. full-scale intelligence quotient (FSIQ) scores] and others a discrepancy between verbal [i.e. verbal IQ (VIQ)] and visual-perceptual [i.e. performance IQ (PIQ)] intellectual function [1–5]. A clearer understanding of the exact impact hydrocephalus may have on intellectual functioning in children is imperative to the development of appropriate school-based intervention plans (i.e. individualized education plans). A recent review of the literature concerning intellectual development in children shunted for hydrocephalus [5] discussed these conflicting findings and suggested that the discrepancies observed in previous studies were due to differences in age, etiology and treatment of the sample populations.

We examined the intellectual function of children with posthemorrhagic hydrocephalus, diagnosed at an early age, who were treated with a uniform shunting strategy. History of infections, prematurity, hemorrhages and shunt revision was considered in the analysis. Additionally, we examined whether hydrocephalus-linked discrepancies between verbal intelligence (VIQ) and performance (nonverbal) intelligence (PIQ), identified in previous studies [1–5], were present in this selected study group.

Subjects and Methods

Subjects

The participants included 41 children and adolescents with hydrocephalus, between 6 and 16 years of age and 16 nonhydrocephalic controls matched for age and education (see table 1). The controls were healthy siblings and friends of the patients who volunteered for the study. All patients with hydrocephalus were shunted within the first year of life. The etiology was posthemorrhagic hydrocephalus in all cases. The children with hydrocephalus were uniformly shunted with nonsiphoning valves, which were maintained up to study inclusion. Patients were excluded if they had a history of shunt infection or revision in the year prior to study participation or if they were functioning at a level >1 year below age-expected academic grade placement, based on parental report. This latter constraint sought to control for the undue effect on group means of extremely low scores, specifically from factors associated with prematurity or other concomitant medical or developmental conditions.

After Institutional Review Board approval and informed consent, participants were recruited through the Pediatric Neurosurgery Service at the University of Chicago. Shunts were placed in the right occiput (n = 21), right frontal region (n = 8), left frontal region (n = 4), left occiput (n = 3), bilateral occiput (n = 1) and

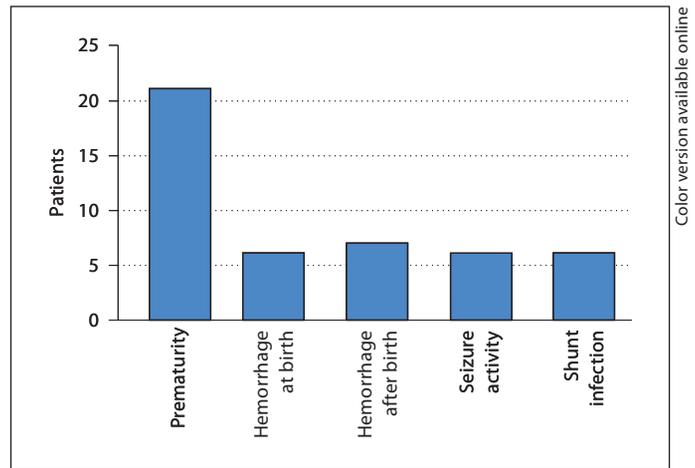


Fig. 1. Complications in hydrocephalic patients.

Table 1. Sample characteristics

	Shunted (n = 41)	Controls (n = 16)
Mean age \pm SD, years	9.81 \pm 3.22	10.36 \pm 2.93
Gender		
Males	28	11
Females	13	5
Mean education \pm SD, years	3.8 \pm 3.1	4.63 \pm 2.5
Handedness		
Right	25	15
Left	16	1

right ‘hemisphere’ (n = 4). Of the participants with hydrocephalus, 25 subjects had 1 or no revision to their shunt, 11 had 2–4 revisions and 5 subjects had ≥ 5 shunt revisions. With regard to infection history in this same group, 4 had a history of 1 infection, 3 had 2 infections, and 34 had no history of shunt infection. No child included in the patient group had undergone shunt revision for malfunction or infection within the 12 months prior to cognitive testing. Seventeen of the 35 documented children (48%) were the result of a premature birth; 4 of those (11%) had experienced a hemorrhagic episode at the time of their birth, while 6 other patients (17%) had experienced hemorrhages at a later stage of development. Finally, 6 patients had developed seizures at some point (fig. 1).

Cognitive Measures

All participants were administered the Wechsler Intelligence Scale for Children, Third Edition (WISC-III) [6], as part of a larger neuropsychological battery. Completion of the WISC-III was typically within the first hour of assessment. All tests were administered by a trained psychometrician supervised by a clinical neuropsychologist.

Fig. 2. Distribution of FSIQ for control (group 1) and shunted (group 3) subjects. The scaled scores (SS) were compressed to yield subject groupings equivalent to 0.5 standard deviation.

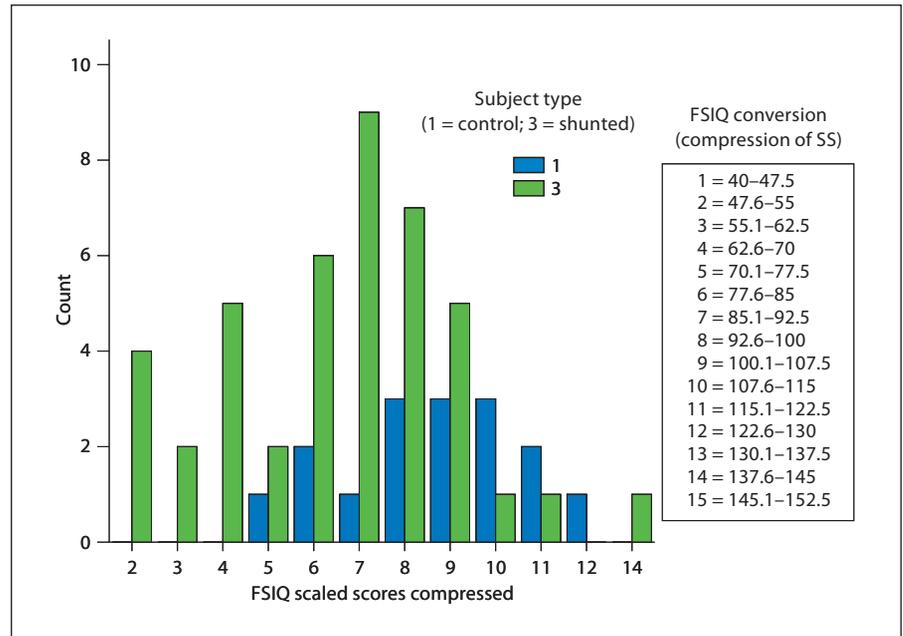
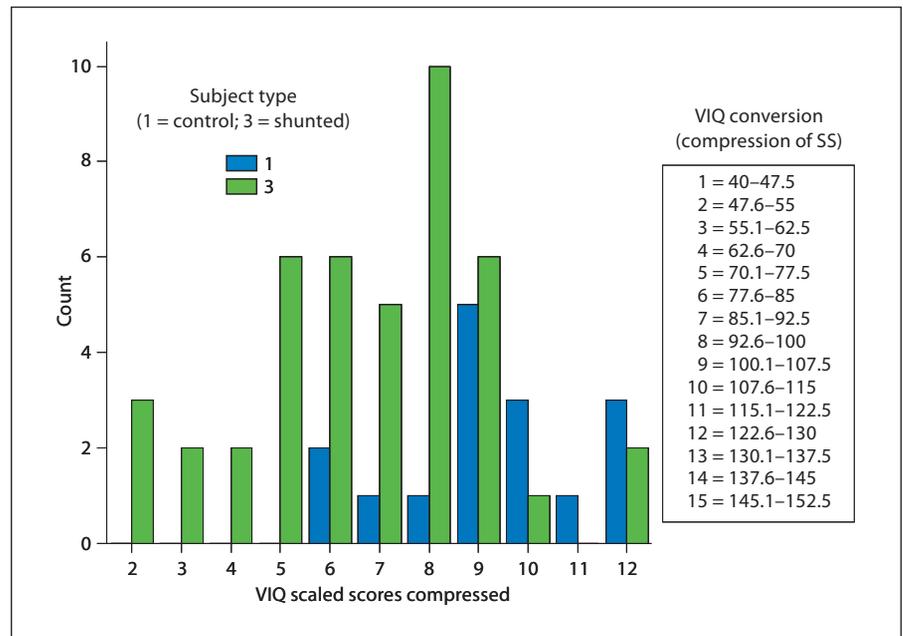


Fig. 3. Distribution of VIQ for control (group 1) and shunted (group 3) subjects. The scaled scores (SS) were compressed to yield subject groupings equivalent to 0.5 standard deviation.



The subtests comprising the WISC-III assess a fund of verbally acquired information, verbal analogical reasoning, perceptual organizational and visuospatial processing, information processing speed and auditory working memory. The WISC-III is designed for use with children between the ages of 6 and 16. It yields 3 main factor scores addressing general intellectual functioning: a VIQ, reflecting verbal problem solving skill development, a PIQ, reflecting nonverbal problem solving skill development, and a global FSIQ. The mean score on the WISC-III is 100, with a standard deviation of 15; scores falling between 85 and 115 define the broad average range of performance. The WISC-III

subtest scores are referenced by a mean of 10 and a standard deviation of 3; scores falling between 7 and 13 define the broad average range.

Statistical Methods

Statistical analyses were run utilizing SPSS 11 (SPSS, Inc., Chicago, Ill., USA), for the calculation of comparative t tests and correlational analyses. A review of data indicated that the WISC-III factor scores were normally distributed (i.e. FSIQ, VIQ, PIQ) and variances were equivalent between the groups (see fig. 2–4). All variables of intellectual functioning were found to be within nor-

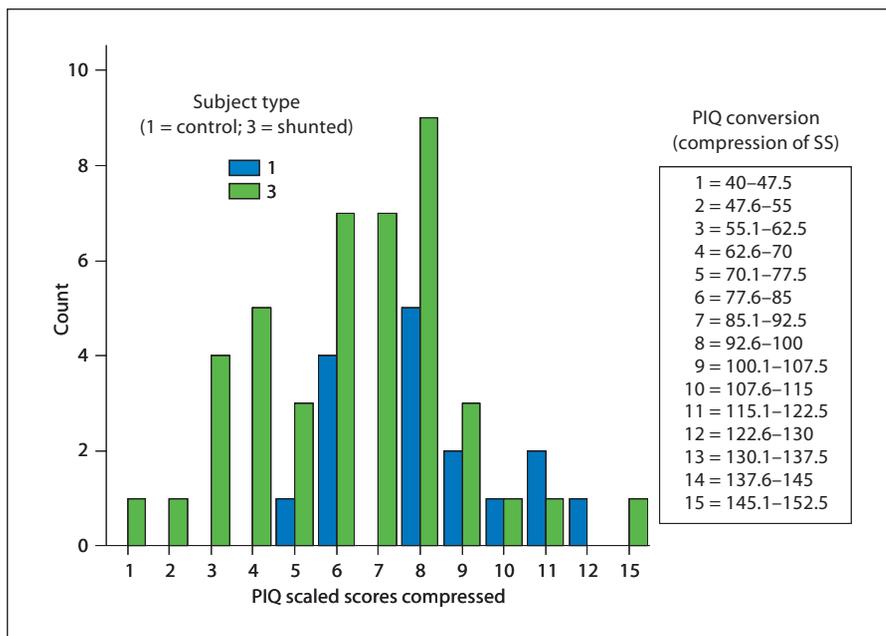


Fig. 4. Distribution of PIQ for control (group 1) and shunted (group 3) subjects. The scaled scores (SS) were compressed to yield subject groupings equivalent to 0.5 standard deviation.

mal limits (i.e. skewness and kurtosis <1 or ≥ 1). This revealed a normal distribution for both groups, with hydrocephalus patient scores centered around a mean 1 standard deviation below the mean score for the control group. For each IQ variable, a 2-tailed (independent-samples) t test was conducted comparing the mean performance of the shunted subjects to that of the nonaffected controls. Significance testing employed a p value standard of 0.01 to control for multiple comparisons.

Results

The analyses revealed significant differences across the WISC-III index scores including FSIQ [$t = 3.87$ (55), $p \leq 0.001$], VIQ [$t = 3.99$ (55), $p \leq 0.001$] and PIQ [$t = 2.86$ (55), $p \leq 0.006$], with the patient group performing significantly below the nonaffected group across all measures. The FSIQ was significantly lower (83.8 ± 17.4) in the experimental group than in the control group (102.9 ± 14.7 ; $p < 0.01$). The VIQ in the experimental group (86.6 ± 17.6) was also significantly lower than that of the controls (107.7 ± 14.4 , $p < 0.001$). Similarly, the PIQ in the experimental group (83.6 ± 17.7) was significantly lower than that of the controls (98.2 ± 17.8 , $p < 0.01$). The mean scores and ranges of the obtained scores, p values and standard deviations for each group, for both factor and subtest scores, are presented in table 2. As can be observed in table 2 and through an examination of the histograms in figures 2–4, the patient group scored 1 stan-

Table 2. Independent-samples t tests comparing intellectual functioning in patients with hydrocephalus and controls

Indices of intellectual functioning	Shunted	Controls	p values
WISC-III FSIQ	83.8 ± 17.4	102.9 ± 14.7	<0.001
WISC-III VIQ	86.6 ± 17.6	107.1 ± 14.4	<0.001
WISC-III PIQ	83.6 ± 17.7	98.2 ± 17.8	0.006

Values are means \pm standard deviation.

dard deviation below the nonaffected controls for all 3 WISC-III factor scores: FSIQ, VIQ and PIQ.

FSIQ did not correlate with history of hemorrhage, shunt revision, prematurity, shunt infection or seizure ($p = 0.95-0.20$; table 3).

Discussion

Mataro et al. [5] suggested that prior neuropsychological studies of cognitive function in children with hydrocephalus have been hindered by a number of potential selection biases: inclusion of patients with hydrocephalus stemming from different etiologies, small sample sizes, failure to document the type of hydrocephalus in the

Table 3. Shunt revisions or infection correlated with intellectual functioning

	Shunt revisions		Shunt infections	
	r(40)	p	r(40)	p
FSIQ	0.21	0.20	-0.003	0.99
VIQ	0.23	0.16	-0.18	0.91
PIQ	0.19	0.25	0.01	0.95

sample studied, the contribution of other brain abnormalities and other therapeutic variables such as the type of shunt valve implanted. In the present study, we examine the intellectual functioning of 41 children with a history of posthemorrhagic hydrocephalus, shunted with a uniform valve type in their first year of life. All patients had been free of treatment (i.e. shunt revisions or infections) for 1 year before testing. We found that these children, regardless of age, etiology or medical history (i.e. prior shunt revisions or infections), consistently performed approximately 1 standard deviation below peers on all measures of intellectual functioning. Specifically, the average child with hydrocephalus included in this study showed a mean cognitive performance (WISC-III FSIQ) at the 16th percentile, in comparison to the normative average of the 50th percentile achieved by healthy peers (i.e. WISC-III FSIQ of 103). This pattern of deficit was observed across both verbal and nonverbal measures of intellectual functioning.

These results have significant implications regarding the long-term cognitive impact of early-onset hydrocephalus and the need for early intervention. The uniform sample of children included in the current study show intellectual deficits in comparison to same-aged peers, despite reports that they function at or near academic grade level. While CSF shunting relieves intraventricular pressure and presumably decreases pressure on surrounding brain tissue, our data confirm the observation that initial damage to the developing brain results in significant developmental differences in cognitive functioning and potentially permanent impairment [2, 3, 7–10], which may very likely contribute to poorer developmental outcomes over time.

Research concerning the long-term impact of low-average intellectual functioning on developmental potential has shown that individuals with an IQ of ≤ 85 typically attain and hold less skilled positions in regard to employment and show less success at moving beyond

high school level academic training [11–14]. Below average intellectual functioning frequently is associated with poorer developmental outcomes, including reduced economic success and stability [15] and social standing [12]. The impact of early alterations to neural development, as observed with this study, both across grey and white matter regions, likely leaves these youngsters vulnerable to reduced social and academic success, and at risk for developing increased difficulties with functioning across time, ultimately limiting viability and functional standing.

The first year of life is pivotal in the development of the human brain and initial disruption of developing circuits, as occurs with hydrocephalus [9], clearly impacts ongoing development of cognitive processes. Supporting this observation, Villani et al. [16], in a long-term follow-up of 78 patients with aqueductal stenosis, found that only 45% of their sample showed normal cognitive and motor development. Futagi et al. [17] reported that earlier onset of hydrocephalus was associated with poor intellectual outcome. Aoyama et al. [18] examined morphological impairments caused by early hydrocephalus in mongrel dogs. They identified both cortical and subcortical dysfunction, characterized by swollen and deformed axons and dendrites, which remained after shunt placement. They inferred, in line with hypotheses presented by Fletcher et al. [3, 9] and Dennis et al. [2], that disruption in periventricular white matter may preferentially account for the cognitive impairment seen in hydrocephalic children.

In terms of prior studies suggesting lateralization of deficits in children with early shunted hydrocephalus (i.e. PIQ worse than VIQ), it is possible that this may hold true for individuals who experience hydrocephalus at later stages of development, when networks are already integrated; however, this pattern was not found in the present study group. Instead, in line with the findings we report, it is quite likely that when hydrocephalus occurs early in infancy and shunting takes place during the first year of life, the development of both cerebral white and gray matter is altered in some way that causes long-term consequences for global cognitive development. Rather than reflecting cortical versus subcortical development, the diversity of findings may mirror a pattern of diffuse impairment of white matter networks, in conjunction with the loss of gray matter area (i.e. as proposed in defects of neural migration [19]).

In summary, reduced intellectual abilities are documented in this relatively homogeneous group of children with posthemorrhagic hydrocephalus, treated with uni-

form shunting within the first year of life, and whose history of prematurity, infections or prior shunt revisions did not statistically impact their cognitive outcome. While we examined whether a history of hemorrhage impacted cognition, we were unable to obtain data regarding grade or type. Some studies have suggested that grade II and grade IV intraventricular hemorrhage may have different impacts on overall outcome and thus should be examined in future analyses.

When we investigated the academic performance of our older patients (age >9 vs. age <9), we found that these children began to demonstrate decreased academic scores (age ≥ 9 vs. age <9: Woodcock Johnson Tests of Achievement – 3rd Edition). Given that intellect and academic performance are correlated [20], this group of children may be academically remediable by appropriate early intervention in the early school years. Future studies assess-

ing academic performance and general cognitive development in this sample of children, particularly executive and memory skills, will be helpful in identifying the specifics of the interventions required. Given animal findings such as those by Aoyama et al. [18], further studies correlating functional imaging such as diffuse tensor imaging data with cognitive development in children with hydrocephalus will give insight into the mechanisms underlying these observed functional differences.

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