Spina bifida and neuropsychology

To The Editor: We read with great interest the recent article by Hampton et al.1 (Hampton LE, Fletcher JM, Cirino PT, et al: Hydrocephalus status in spina bifida: an evaluation of variations in neuropsychological outcomes. Clinical article. J Neurosurg Pediatr 8:289–298, September 2011), which focused on the variations in neuropsychological outcomes in 208 children with spina bifida (SB) in terms of verbal and nonverbal IQ, reading and mathematical achievement, memory, and visuospatial and executive functions associated with motor skills. They divided children with SB into groups according to their hydrocephalus status, that is, shunted hydrocephalus, arrested hydrocephalus, or no hydrocephalus. The third group included 24 cases with myelomeningocele, meningoecele, lipomyelomeningocele, lipomeningocele, and lipoma, although not all of them had hydrocephalus. In comparing the 3 groups of children with SB with normal children, they found that the SB groups had lower performance, considering the significant association with a lower socioeconomic status in these children. Except for fine motor skills, most neuropsychological profiles were similar in the groups with SB and arrested or shunt-treated hydrocephalus.

Here, we want to mention several points related to cognitive deficits in children with SB. Spina bifida is a wide spectrum of diseases ranging from a pure radiological finding in a totally normal child to severe mental and motor deficits in a child with myelomeningocele. In between we can count some disorders that are classified mostly as closed SB, which was mostly considered in the third group in the Hampton et al. study and typically lacks any structural or functional brain abnormalities. Considering cognitive or performance differences in SB as a whole is a big mistake. Spina bifida with hydrocephalus, whether treated or untreated, almost always occurs in a child with myelomeningocele. We believe that cognitive development and performance are compromised in children with myelomeningocele by the disorder itself and not by hydrocephalus. The presence of hydrocephalus and its associated management consequences may supply more neuropsychological dysfunction, but this is not comparable with the consequences of the disease itself. Considering myelomeningocele with closed SB disorders in a group to make SB without hydrocephalus confuses the findings even though the authors presented a normal group for the third group with SB.

According to our 2005 study2 in which we considered IQ evaluation using the Raven Progressive Matrices test, all children in the myelomeningocele group had an average or above-average IQ. We could not find a statistically significant correlation between the presence of a shunt or shunt-related complications, motor disability, or spinal level of the lesion and IQ. Therefore, the lower IQ and lower cognitive levels in patients with myelomeningocele result from the disease process itself and not from the associated complications.

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The authors report no conflict of interest.

References

RESPONSE: In their editorial, Nejat et al. argue that SB is not a uniform disease, but one that varies in neural and physical presentation. They also argue that the presence of hydrocephalus almost always occurs in children with SB myelomeningocele, and thus the cognitive presentation of our sample with myelomeningocele and shunted hydrocephalus was a result of the myelomeningocele and its associated neural dysmorphologies, and not hydrocephalus status. The authors refer to their study10 in which they used a measure of fluid intelligence11 and determined that children with myelomeningocele performed in the average to above-average range of IQ. In addition, their examination of the effects of shunt status, shunt complications, motor disability, and spinal lesion level did not reveal significant associations. While we appreciate the interest in our article, it is unclear how the studies are comparable or what concerns they had regarding our results.

We agree that many of the effects on neuropsychological development reflect brain dysmorphologies specific to myelomeningocele. However, it is hard to separate the effects of hydrocephalus without using more specific cognitive tasks. Neuropsychological performance is related to the status of the cerebellum, corpus callosum, and probably the severity of hydrocephalus, which cannot be assessed in posttreatment cases.2,4–6 The authors neglected to use specific neuropsychological measures, and as McLone and Zebracki9 noted, it is hard to extrapolate to the effects of hydrocephalus using only a measure of fluid intelligence.

The authors also expressed concern regarding our use of a third group of individuals with SB without myelo-
meningocele. As we stated, this group was intended to control for socioeconomic status, ethnicity, and history of reduced attention.

Furthermore, the study by the authors is alone in showing little effect of myelomeningocele on intelligence, itself a limited assessment of outcome.9 Fletcher and colleagues1 showed clear effects of lesion level because of reduced performance in patients with thoracic level lesions, which were not represented in the authors' sample. Few studies find the effects of shunt status in children, but there are associations if adults are included, and there are associations with motor disability if thoracic level lesions are included.2,8,12,13 Overall, it is hard to determine how the authors can make strong statements about the relation of neurobehavioral outcomes and myelomeningocele given the highly selected nature of the sample and the reliance on an assessment of fluid intelligence.8

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References
11. The authors performed a prospective study in 128 children with syndromic or complex craniosynostosis to determine the value of optic nerve sheath (ONS) diameter on ultrasonography in the early detection of increased intracranial pressure (ICP). They concluded that it cannot be used as a screening tool in place of conventional funduscopic in these settings.
12. As the authors of a recently published meta-analysis3 on the diagnostic accuracy of ONS diameter on ultrasonography for the detection of raised ICP, we are very interested in this emerging tool. We congratulate the authors on their interesting paper.
13. Ultrasoundography for ONS diameter has mainly been investigated in traumatic brain injuries and intracerebral hemorrhage in adults and shows good diagnostic accuracy in these acute settings, with a pooled sensitivity of 90% and a pooled specificity of 85%.3
14. In the pediatric population, there are fewer studies,1,5 and most of them have not focused on this tool to compare it with invasive monitoring devices (“gold standard”). Clearly, the best method to validate ONS diameter on ultrasoundography is to compare it directly with the gold standard, but this is not always ethically feasible. Thus, this tool needs to be validated first in settings in which invasive monitoring will be used to delineate a proper comparison and pinpoint cutoff values. Authors of several studies have tried to achieve this, so that we can be more certain of the diagnostic accuracy of ONS diameter by ultrasonography in acute settings.
15. In this study, Driessen et al. took the current recommended cutoff measure to evaluate this tool in the specific setting of pediatric craniosynostosis; indeed, such a study in other neurosurgical conditions, such as hydrocephalus, should be encouraged.
16. However, many questions persist—among them an uncertainty as regards the correlation between variations in ICP and ONS diameter: only one study4 has explored this issue to date. It is for this reason that we would like to question the authors about the 3 patients who had invasive monitoring. Did not the authors perform ultrasonography in these patients during this monitoring?
17. As stated in this interesting paper, ICP varies significantly in craniosynostosis so that the two measurements (invasive and noninvasive) should be performed at the same time. It would be very interesting to have records of ONS diameter measurements during invasive monitoring to detect any correlation between natural fluctuations of ICP and ONS diameter and to evaluate the diagnostic accuracy of ultrasonography.
18. The lack of accuracy of ONS diameter by ultrasonography in this study can be explained by two alternative hypotheses: either the ONS diameter measurement was

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