

Neuropsychological deficits in children with congenital hydrocephalus: a systematic review

María José García-Rubio^{1,2,3}, Marta Aliño^{1,2,3}, Irene Cano-López^{1,2,3}, Pilar Chisbert-Genovés^{2,4}, Luis Moreno-Oliveras^{2,4}, Aránzazu Duque^{1,3}, Encarna Rama^{1,3}, Mercedes Almela^{3,5}, Paula Martínez^{1,3}, José Piquer-Belloch^{2,4}, y Sara Puig-Pérez^{1,2,3}.

¹Faculty of Health Sciences, Valencian International University, Valencia, Spain.
²VIUNED Chair of Global Neuroscience and Social Change – Valencian International University.
³Research Group in Psychology and Quality of Life (PsiCal). Valencian International University, Valencia, Spain.
⁴NED Foundation (Valencia, Spain)
⁵Department of Cognitive Neuropsychology. Tilburg School of Social and Behavioral Sciences, Tilburg, the Netherlands.

Introduction and objectives

Pediatric hydrocephalus is a global health and social problem. The most common form of congenital hydrocephalus is due to both spina bifida, and myelomeningocele although it could also be caused by hemorrhage. The physical symptoms (disproportionate skull, headaches, nausea, irritability, etc) have been well studied; however, the neuropsychological profile associated with this neurological condition is not so clear. This review aims to explore the neuropsychological deficits associated with patients who were diagnosed during childhood with congenital hydrocephalus.

Method

This review has been based on the PRISMA 2020 Declaration. A search was carried out in the WOS, Pubmed, Scopus and ProQuest databases, using booleans and limits. The keywords were selected from Thesaurus: hydrocephalus, children, neuropsychology and others. Eligibility criteria included: empirical studies which were written in English/Spanish and published in the last 10 years whose description followed the PICOS method.

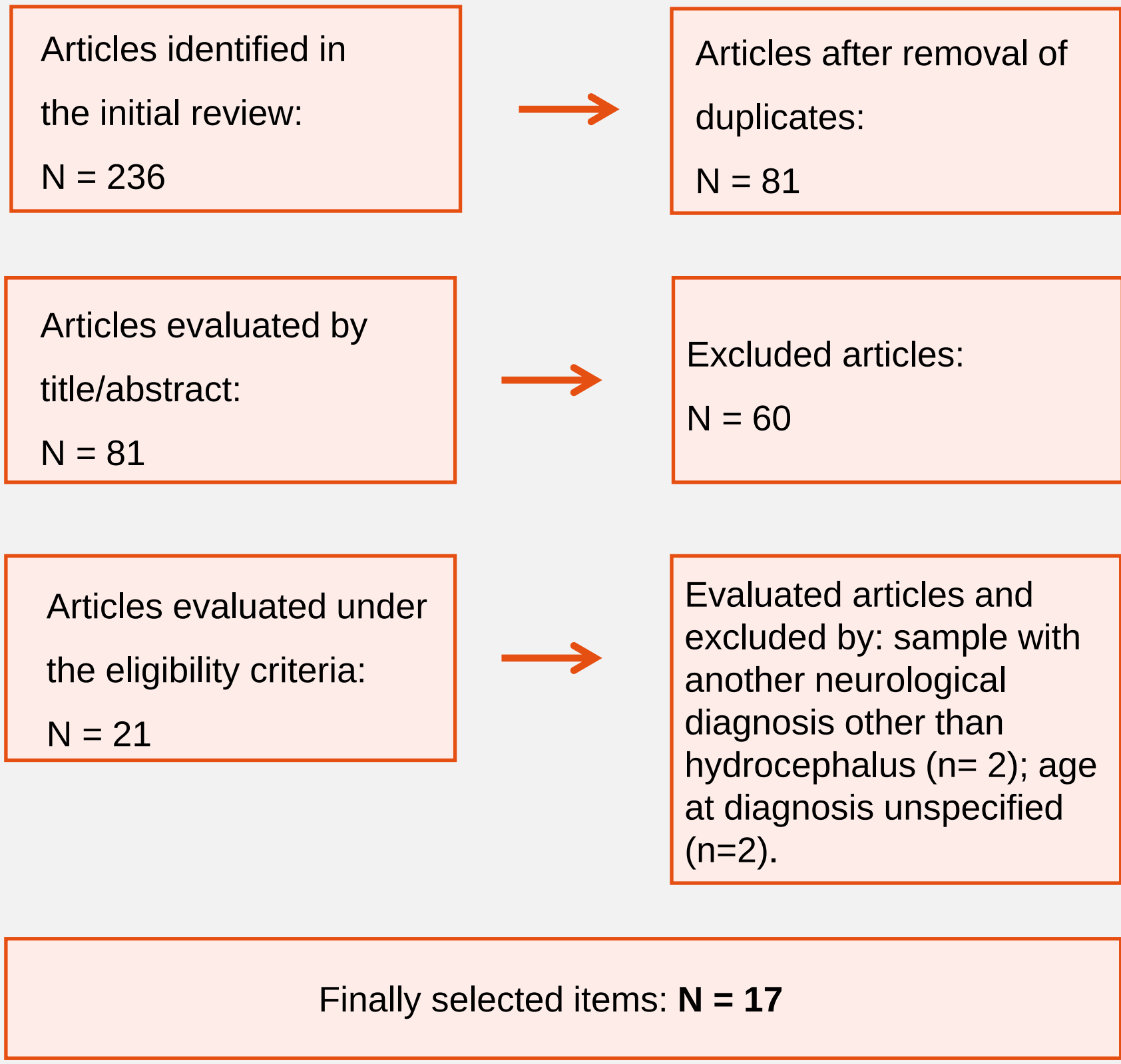


Figure 1. Flow diagram reflecting the search strategy of this review

Inclusion criteria	Exclusion criteria
1. Main diagnosis of hydrocephalus due to both spina bifida, myelomeningocele, or postnatal hemorrhage).	1. Use of general neuropsychological tests (eg, IQ measurement).
2. Age (0-6 years).	2. The work does not include the quantitative scores of the neuropsychological tests.
3. Specimen type: Humans.	3. Works in book chapter format, editorials, conference communications and systematic reviews.
4. Application of standardized neuropsychological tests with explicit quantitative scores at work.	4. Populations with a secondary rather than a primary diagnosis of hydrocephalus.
5. Experimental and quasi-experimental empirical articles published in peer review journals.	5. Studies with non-human sample.

Figure 2. Eligibility criteria

Results

The initial review yielded a total of 236 papers. After elimination of duplicate papers, 81 papers were counted and reduced to 17 when the proposed eligibility criteria were applied. Studies focused on patients with hydrocephalus due to spina bifida (H-SB) show poor cognitive performance, especially in areas such as executive function (planning and/or working memory). In addition, studies with children with hydrocephalus caused by myelomeningocele (H-MM) show a cognitive impairment in visuospatial and visuomotor skills; while studies about children with post-hemorrhagic hydrocephalus (H-PH) present a generalized neuropsychological profile which includes the impairment in memory, language, visual and motor areas. Finally, studies focused on children with a diagnosis of both spina bifida and myelomeningocele (H-SBMM) show that they have longer reaction times in the execution of tasks, and deficits in attention, response inhibition and reading comprehension, compared to a control group and also when compared to children with a diagnosis of H-PH. Further, these studies have found that factors such as age at diagnosis, amount of damaged gray matter and others may modulate cognitive performance in this specific group.

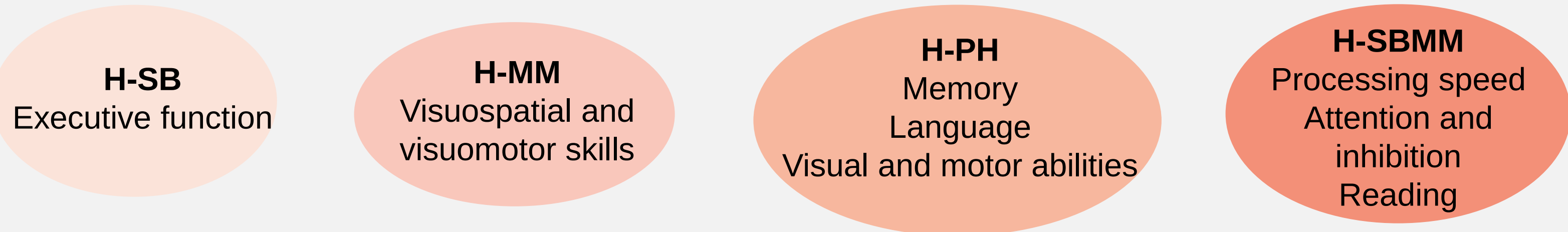


Figure 3. The cognitive deficits associated with each of the groups proposed in this review are shown. (H-SB= hydrocephalus due to spina bifida; H-MM= hydrocephalus due to myelomeningocele; H-PH post-hemorrhagic hydrocephalus; H-SBMM= hydrocephalus caused by both spina bifida and myelomeningocele)

Conclusions

Pediatric congenital hydrocephalus is associated with a heterogeneous neuropsychological profile modulated by the origin of this diagnosis. In this review significant differences in the cognitive improvement of H-SB, H-MM, H-PH, and H-SBMM groups are found; sometimes modulated by factors such as age, shunt treatment and social environment. These results implicate the adaptation of the neuropsychological evaluation protocol according to the origin of the diagnosis of congenital hydrocephalus in this child population.

References

Barnes, M. A., Faulkner, H., Wilkinson, M., & Dennis, M. (2004). Meaning construction and integration in children with hydrocephalus. *BRAIN AND LANGUAGE*, 89(1), 47-56. [https://doi.org/10.1016/S0093-934X\(03\)00295-5](https://doi.org/10.1016/S0093-934X(03)00295-5)
Barnes, M. A., Pengelly, S., Dennis, M., Wilkinson, M., Rogers, T., & Faulkner, H. (2002). Mathematics skills in good readers with hydrocephalus. *JOURNAL OF THE INTERNATIONAL NEUROPSYCHOLOGICAL SOCIETY*, 8(1), 72-82. <https://doi.org/10.1017/S1355617702811079>
Casari, E. F., & Fantino, A. G. (1998). A longitudinal study of cognitive abilities and achievement status of children with myelomeningocele and their relationship with clinical types. *EUROPEAN JOURNAL OF PEDIATRIC SURGERY*, 8, 52-54. <https://doi.org/10.1055/s-2008-1071255>
Dewan, M. C., Rattani, A., Mekary, R., Glancz, L. J., Yunusa, I., Baticulon, R. E., ... & Warf, B. C. (2018). Global hydrocephalus epidemiology and incidence: systematic review and meta-analysis. *Journal of neurosurgery*, 130(4), 1065-1079. Enslin, J. M. N., & Fieggen, A. G. (2019). Global Perspectives on the Treatment of Hydrocephalus. In *Cerebrospinal Fluid Disorders* (pp. 351-361). Springer, Cham.
Fletcher, J. M., Brookshire, B. L., Landry, S. H., Bohari, T. P., Davidson, K. C., Francis, D. J., ... Morris, R. D. (1996). Attentional skills and executive functions in children with early hydrocephalus. *DEVELOPMENTAL NEUROPSYCHOLOGY*, 12(1), 53-76. <https://doi.org/10.1080/87565649609540640>
Hampton, L. E., Fletcher, J. M., Cirino, P., Blaser, S., Kramer, L. A., & Dennis, M. (2013). Neuropsychological profiles of children with aqueductal stenosis and spina bifida myelomeningocele. *Journal of the International Neuropsychological Society*, 19(2), 127-136.
Mikkelsen, R., Rødevand, L. N., Wiig, U. S., Zahl, S. M., Børtsen, T., Skarabø, A. B., ... & Wester, K. (2017). Neurocognitive and psychosocial function in children with benign external hydrocephalus (BEH)—a long-term follow-up study. *Child's Nervous System*, 33(1), 91-99.
Yerramneni, V. K., & Kotha, V. K. (2017). Posttraumatic Hydrocephalus: Risk Factors, Treatment Modalities, and Prognosis. *Indian Journal of Neurosurgery*, 6(03), 198-202.
Zahl, S. M., Egge, A., Helseth, E., Skarabø, A. B., & Wester, K. (2019). Quality of life and physician-reported developmental, cognitive, and social problems in children with benign external hydrocephalus—long-term follow-up. *Child's Nervous System*, 35(2), 245-250.