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Abstracts

Neuropsychology and Learning

Early Cognitive Development in Spina Bifida
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Background Hydrocephalus is often associated with Spina Bifida (SB) and has been shown to lead to specific cognitive deficits in school aged children, including executive dysfunction, visuospatial deficits and poor numeracy skills. Despite these well-known cognitive deficits, only one study has previously investigated the earlier cognitive development in children with SB and hydrocephalus. Our study aimed to further investigate the early cognitive development of children with SB and shunted hydrocephalus in the Australian context.
Methods Longitudinal retrospective data of children from the Spina Bifida Service at the Children's Hospital at Westmead, Sydney, Australia (2005-current) were analysed. The proportion of children who scored below average ('at risk' or 'delayed') in cognition on routine developmental assessment prior to 4 years of age was collated. Associations between performance on early developmental assessment and later neuropsychological measures (e.g., IQ, visuospatial skills, and executive functioning) were also investigated.
Results Preliminary data revealed that 49% of children with SB and hydrocephalus were 'at risk' or 'delayed' in their cognitive skills. Of those below average on a developmental cognitive assessment, 67% also scored below average on IQ and 50% on a measure of executive functioning when tested at school age (at a mean age of 7 years).
Conclusions Our findings are consistent with previous research that suggests the cognitive deficits associated with SB and hydrocephalus can be identified earlier than school age. These findings highlight that it is important for children with SB and hydrocephalus that are at risk for delay to be identified so that they can receive the appropriate intervention and services; and their cognitive development can continue to be monitored prior to starting school. Additionally, the data demonstrates that early intervention should consider cognition alongside the physical impairments of children with SB.

Neuropsychological Performance of Preschool Children with Spina Bifida (SB)
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Background Little is known about the neurocognitive development of preschool children with SB. We examined the early cognitive development of children with SB across multiple domains of functioning.
Methods A group of 82 children with SB from two states in the USA, aged 3–6 years, participated in neuropsychological testing. We used Neuropsychological Assessment (NEPSY), Differential Abilities Scale (DAS), Wide Range Assessment of Visual Motor Abilities (WRAVMA), Peabody Picture Vocabulary Test (PPVT-4), Bracken Basic Concept Scale (BBCS-3: R) to evaluate participants on 10 subcomponents of functioning: NEPSY comprehension of instructions (n = 77), verbal fluency ($n = 69$), and memory and learning (n = 70); DAS verbal (n = 77), nonverbal (n = 81), and spatial abilities ($n = 66$); WRAVMA visuospatial (n = 62) and fine motor abilities (n = 75); PPVT-4 receptive vocabulary (n = 79); and BBCS-3:R basic concept development ($n = 76$).
Results Children with SB performed at the lower end of expected levels of the NEPSY scaled scores (M = 10, SD = 3): comprehension of instructions (M = 8.3), verbal fluency (M = 7.9), and memory and learning (M = 7.9). The other measures (M = 100, SD = 15) also indicated low average abilities on DAS verbal (M = 91.2) and nonverbal skills (M = 87.9); WRAVMA visuospatial skills (M = 90.5); PPVT-4 re-

ceptive vocabulary (M = 96.4), and BBCS-3: R basic concept development (M = 88.8). DAS spatial (M = 82.4) and fine motor (M = 78.5) abilities were below average.

Conclusions Children with SB had low average skills in language and memory functioning and below average skills in fine motor abilities. Spatial performance findings were inconsistent in that children with SB showed low average abilities on the WRAVMA and below average abilities on the DAS. Early identification of neuropsychological strengths and weaknesses among children with SB may help families to advocate for appropriate assistance and professionals to tailor services to the needs of each child.

Developmental Outcomes by Hydrocephalus and Shunting Status for MOMS Subjects Status Post Prenatal Repair

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Background Myelomeningocele-associated hydrocephalus (MMH) causes cognitive, sensory, and motor dysfunction. MMH is typically treated with shunt placement; due to high complication rates, however, infants are not always shunted despite evidence of hydrocephalus. The objective of this study was to compare developmental outcomes for children with myelomeningocele status post prenatal repair: 1) without hydrocephalus, 2) with shunted hydrocephalus, and 3) with unshunted hydrocephalus.

Methods Secondary data analysis of Management of Myelomeningocele Study (MOMS) was conducted. MOMS used standard criteria for identifying hydrocephalus. Outcomes measured were the Bayley II Mental and Psychomotor Development Indices and Peabody Gross, Fine, and Total Motor Indices (BMDI, BPDI, PGMI, PFMI, PTMI, respectively) at 30 months of life. The Kruskall-Wallis test was used to compare outcomes between groups, with p-values less than 0.017 considered statistically significant.

Results Of the 87 children who underwent prenatal repair, 25 (28.7%) did not meet hydrocephalus criteria and were not shunted, 39 subjects (44.8%) met criteria and were shunted, and 23 subjects (26.4%) met criteria but were not shunted. At 30 months, mean scores on the BMDI, BPDI, PFMI, PGMI, and PTMI were 90.0, 68.2, 94.0, 71.2, and 78.7 for subjects without hydrocephalus, 87.6, 60.6, 89.8, 65.2, and 73.0 for subjects with shunted hydrocephalus, and 92.1, 64.7, 93.1, 68.3, and 76.6 for subjects with unshunted hydrocephalus. There were no statistically significant differences for any of these outcomes across groups.

Conclusions No differences in mental or motor outcomes at 30 months were identified between those without hydrocephalus, shunted hydrocephalus, and unshunted hydrocephalus. Limitations of the study include small sample size and thus low power to detect differences between groups as well as potential confounding.

Cognition in Spina Bifida and its Consequences in Daily Life – A Life-span Perspective. A Systematic Review

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Background The number of children born with Spina Bifida in Sweden today is now 10–15 per year compared with 80–100 about thirty years ago. About half of the individuals with Spina Bifida in Sweden are 20 years old or more. It is important to maintain and spread knowledge about this disability to secure professional care, understanding and help for this decreasing group. The aim of the project was to describe actual knowledge on cognitive characteristics in children, youths and adults with Spina Bifida and its consequences for development, learning, process skills and practical everyday functioning in a life time perspective.

Methods Systematic data-base search and quality assessment of articles on cognition in Spina Bifida produced from 2000 to 2013. Sixty-seven articles were chosen relevant according to the McMaster Rating of Evidence.

Results About 70% have an intellectual function above an IQ of 70 according to tests of intelligence. Difficulties in visual perception, attention and memory function and problems with planning and initiation were described as common despite IQ-level. The difficulties still remain into adulthood. Difficulties with planning and initiation were the most hindering factors for autonomy in adult years. Cognitive dysfunction was found to be hindering in all domains of everyday-life when using the classification of ICF (International Classification of Functioning).

Conclusions Knowledge of cognitive functions is necessary to provide optimal care and treatment of chil-

dren, youths and adults with Spina Bifida. This review strongly confirms the mostly life-long need for personal support in preschool, school, at home and at work, regardless of intellectual level. This is crucial to ensure quality of life and equal participation in society.

Can the Cognitive Orientation to Daily Occupational Performance (CO-OP) Approach Enhance Executive Functions and Participation in Young Adults with Spina Bifida – A Multiple Case Study

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Background Participation occurs in the interaction between the person and the environment in activity. Performing an activity requires motor function and executive function. Research shows that persons with Spina Bifida (SB) have activity and participation restriction due both motor- and executive dysfunctions. The Cognitive Orientation to daily Occupational Performance (CO-OP) approach guides the persons to master their own activity goals by using strategies for performance and problem-solving discovered by the person with guidance from the therapist. The objective was to investigate if the CO-OP approach is feasible approach for achieving self-identified goals, executive functions and improvement in self-perceived community participation for young persons with SB.
Methods An explorative multiple case study. Five young persons (16–28 y) with SB participated in a 10-week CO-OP treatment period. Evaluations were carried out on baseline, directly after and 6 months after the treatment with outcomes on all ICF-levels. Experiences of the CO-OP approach were captured with qualitative interviews.
Results Preliminary results show considerable improvement in self-rated performance of the goals, with a median of 5 levels increase on a 10-level scale. Self-rated participation was rated higher by the majority and all participants had significant improvement in the executive ability to plan and stay focused was. In interviews feelings of higher self-efficacy were expressed as participants problem-solving ability increased.
Conclusions CO-OP is a promising approach to achieve personal goals and enhance participation and executive functions through strategy use in young persons with SB. The CO-OP approach provides the opportunity for persons to master everyday life problems by themselves.

Developing a Systematic Neurodevelopmental Monitoring Program for Infant and Toddlers with Spina Bifida: Why Is It Important?

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Background Given the changes in neurosurgical management of hydrocephalus, we are using neurodevelopmental assessments in conjunction with standard medical follow up to carefully monitor developmental progression. These assessments allow for frequent evaluation of the development of cognitive, expressive and receptive communication skills, as well as fine and gross motor skills.
Methods Infants who are followed in our Spina Bifida Center are referred for neurodevelopmental assessments (Bayley: 3, Vineland: II) at 6 months, 12 months, 18 months (with CBCL, BASC-II), and 24 months. Assessments are scheduled more frequently when there are concerns about developmental progression, regression, or neurological changes. Information from the evaluations is provided to our developmental pediatricians and neurosurgical team, community based early intervention and outpatient therapists, as well as their families to help prioritize interventions.
Results We currently have data on 66 patients, with 1 and 6 assessments each; another 25–30 assessments are expected before the conference. This includes 55 patients with a Chiari II malformation; we examined children treated for hydrocephalus with an ETV w/CPC (31) and those who were shunted (17) separately. Of the 66, 12 underwent prenatal closure and 29 were delivered prematurely. Preliminary evaluation shows that children treated with ETV have not shown a significant difference in skill development from those not treated for hydrocephalus.
Conclusions Our neurodevelopmental monitoring program has provided a vehicle for parents to focus on well child development and proactively working with our medical team to support developmental progress. We are hoping that early detection of issues (e.g., slow language acquisition) in early development can help to maintain expected skill progression when provided with rapid intervention through outpatient services.