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The Spina Bifida Multisite Neuropsychology Collaboration and future goals for neuropsychological data in the National Spina Bifida Patient Registry (NSBPR)

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Background: Neuropsychological assessment is crucial to understanding an individual with Spina Bifida's (SB) cognitive strengths and weaknesses to inform treatment, intervention, education and the development of independence. One of the research goals for the 2019-24 Children's Wisconsin site funded application for the NSBPR was to optimize neuropsychology variables in the registry. We proposed to achieve this by a collaborative pilot study "The Relationship of Select Neuropsychological Variables to Clinical Outcomes in Children with Spina Bifida."
Methods: The initial aim was to build a multisite Neuropsychology collaboration with other sites to identify, collect, and analyze neuropsychological tests typically administered to children with Spina Bifida.
Results: To date, the Spina Bifida Multisite Neuropsychology Collaboration has been created which includes clinical neuropsychologists, research personnel, and trainees from 7 sites (6 from NSBPR sites). Meeting monthly, this group has defined several initial goals for collaboration with overarching aims to a) provide systematic, optimal but efficient, and consistent neuropsychological care across sites, b) to further integrate neuropsychology into Spina Bifida Multidisciplinary Clinics by demonstrating its essential value, c) to create databases to allow for scientific inquiry regarding how to optimize cognitive, adaptive, and functional outcomes, and d) to promote the requirement of equitable care across all children with spina bifida regardless of their racial, ethnicity, and regional location backgrounds. To achieve these aims, this group has administered a survey of current practices across clinical neuropsychologists, begun discussions of how to acquire and enter intellectual and adaptive data into the registry, has defined ages for neuropsychological assessment, and clinical variables that contribute to neuropsychological functioning have been determined. Next steps include creation of a multisite neuropsychological pilot study.
Conclusions: Understanding neuropsychological functioning is critical to the care of individuals with SB; this collaboration hopes to increase awareness of the full utility of Neuropsychologists in SB clinical care teams in order to provide optimal clinical care.

Access to neuropsychology care for patients with spina bifida in the United States

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Background: Many patients with Spina bifida (SB) have congenital brain malformations and receive neurosurgical intervention that can impact cognitive, behavioral, and emotional functioning. The Spina Bifida Association's standard of care highly recommends neuropsychological evaluations throughout the lifespan.
Anecdotally, patients with SB are reported to have trouble finding and accessing adequate neuropsychological care. The goal of this study was to gain information about neuropsychology services available to patients with SB in the United States (US)

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and identify possible barriers to neuropsychology care.

Methods: Pediatric neuropsychologists from across the US were invited to complete an online survey about services they provide patients with SB.

Results: Responses from 35 neuropsychologists (representing 22 sites across the US) were collected. The survey responses indicated that pediatric neuropsychology services are available to SB patients within all regions of the US and in a variety of languages. Most referrals come from a parent/caregiver, developmental pediatrician, neurosurgeon, or a rehabilitation medicine doctor. All sites who completed the survey indicated that they offer comprehensive, outpatient evaluations. However, integration of neuropsychology services into inpatient settings and multidisciplinary clinics are reportedly limited to a few sites.

Respondents indicated that waitlist time is the most common and frequent barrier to neuropsychology care. Reported waitlist times across sites are variable, ranging from 1-3 months to over 1 year. To a lesser extent, evaluation cost/insurance/payor type, travel/transportation difficulties, limited referrals, and caregiver misunderstanding about referrals are also reportedly barriers to neuropsychology care.

Conclusions: Neuropsychologists and referring providers are encouraged to implement innovative methods for reducing waitlist times. One option is to increase provision of neuropsychology care in multidisciplinary clinics. Two models for integrating neuropsychology into multidisciplinary clinics are discussed.

The role of neuropsychology consultation prenatally and in the first year of life for children with spina bifida

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Background: Individuals with spina bifida are at risk for cognitive, language, motor, and functional impairments and deficits may be evident within the first year of life. As such, infants with spina bifida should be closely monitored to ensure early identification and prompt access to interventions as needed, consistent with the Guidelines for the Care of People with Spina Bifida. Furthermore, the Guidelines emphasize the importance of prenatal consultation with families about disability and functional outcomes across the life-course.

Methods: The Spina Bifida Baby Clinic at Kennedy Krieger Institute was recently developed to meet the needs of infants with spina bifida. Pregnant mothers are referred for a prenatal consultation visit with the Clinic, which includes rehabilitation medicine, social work, and neuropsychology. A pre-visit questionnaire helps to guide the priorities of the visit, with neuropsychology focused on providing psychoeducational information about the range of cognitive/functional outcomes and supports over the life-course. Infants receive neuropsychology consultation at multi-disciplinary clinic visits at 3, 6, and 9 months of age and a comprehensive neuropsychology outpatient evaluation at 12 months of age. Parent training is provided across time-points focused on promoting child development within the context of family culture and goals.

Results: This model has been implemented and refined over the past 2.5 years. This presentation will focus on the utility of neuropsychological services in providing these services through three illustrative case examples. The utility of telehealth to meet the unique needs of infants and their families within this context will also be discussed.

Conclusions: This presentation will highlight the development of the Spina Bifida Baby Clinic at Kennedy Krieger Institute as well as clinical utility from the neuropsychological perspective. Future directions for investigating outcomes of this model of care will also be discussed.

Implementation of systematic neuropsychological evaluations of children with spina bifida in Sweden

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Background: Individuals with spina bifida (SB) and comorbid hydrocephalus are at increased risk of a specific “cognitive phenotype”. Neuropsychological evaluations (NE) might be beneficial to identify specific cognitive strengths and weaknesses, lay the foundation for interventions, and for adaptations in

the environment. In Sweden, healthcare and education are largely covered by taxes and provided free of charge. However, not all children with SB are offered NEs at their habilitation clinics.

Methods: In 2013, a group of Nordic neuropsychologists developed guidelines NEs for children and adolescents with cerebral palsy called CP*Cog*. These guidelines were adapted for children with SB and are called MM*Cog*. At a minimum, children with SB are recommended to be offered neuropsychological testing before first grade (4-5 years of age) and before entering junior high school (12-13 years of age). To assess intelligence, the Wechsler Preschool and Primary Scale of Intelligence and the Wechsler Intelligence Scale for Children are recommended, depending on age. In addition, the Beery-Buktenica Developmental Test of Visual-Motor Integration is recommended for assessing visual-motor integration. To assess executive and adaptive functioning, it is recommended to use the Behavior Rating Inventory of Executive Function and the Vineland Adaptive Behavior Scales questionnaires, respectively. Results are entered into the national registry and multidisciplinary follow-up program MMCUP.

Results: MM*Cog* was added to MMCUP in 2017. Seventy-three children have completed at least some part/s of MM*Cog*. Of the children assessed, 48% reported a different mother tongue than Swedish.

Conclusions: Implementation is slow but an increase in number of children assessed has been noted. Based on results from a 2020-2021 survey study, a lack of neuropsychologists and children with other diagnoses being prioritized to meet with the neuropsychologists are some of the reported barriers. Furthermore, there are great differences based on geographical regions in terms of who gets assessed in MM*Cog*.

Redefining the role of the neuropsychologist in the coordinated care of adults with spina bifida

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Background: There is a significant gap in literature pertaining to best assessments, practices, monitoring, and treatment of adults with Spina Bifida, despite the significant increase in life expectancy from this congenital disorder. The recent revision of the *Guidelines for the Care of People with Spina Bifida* focuses on lifespan care and living with the disorder. These guidelines highlight the importance of patient-centered coordinated care in an interdisciplinary team approach to address all aspects of well-being and health beyond physical symptomatology. Individuals with Spina Bifida face unique health challenges that manifest differently after adulthood demands are imposed compared to those addressed in pediatric care. There is even less literature dedicated to young adults with Spina Bifida and the issues of co-morbid mood and affect disorders, self-esteem, socialization, executive dysfunction, independence, sexual education, and shared decision-making. Neuropsychology's role in this coordinated care has historically focused on separate, comprehensive cognitive assessment and resultant patterns of cognitive functioning rather than addressing and treating the impact of these likely deficits on independent living, transition to adulthood, and navigating self-management.

Methods: Creation of a brief, valid, and reliable screening tool to efficiently assess cognition, self-management, and independent living skills is needed so that techniques and strategies can be discussed by a neuropsychologist integrated into the coordinated care appointment in an Adult Spina Bifida Clinic. This presentation will highlight a model of care using this assessment given to every patient seen in an adult Spina Bifida Clinic to inform mental health, executive functioning, transition to adulthood needs, and independence. Normative data and content validity will be discussed after data are obtained and analyzed.

Results: To be obtained prior to presentation

Conclusions: With this model, therapeutic and cognitive intervention with the patient and their family can begin while they are receiving coordinated care. Barriers and opportunities will also be discussed.

Increased utilization of neuropsychology services in a pediatric spina bifida clinic

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Background: The Spina Bifida Association's Guidelines for the Care of People with Spina Bifida recommends periodic neuropsychology evaluations throughout childhood and adolescence. Neurocognitive and adaptive skills impact academic learning and performance, and also influence self-management abilities and independence in adulthood. Neuropsychological evaluations can inform need for intervention and supports; however, many children with myelomeningocele do not receive these evaluations. Our spina bifida clinic sought to increase the number of patients with completed neuropsychology evaluations to improve the long-term cognitive and adaptive functioning of our patients.

Methods: Our spina bifida clinic implemented a quality improvement (QI) initiative in 2021 designed to improve patient access to neuropsychology services, increase the number of referrals to neuropsychology, and increase completed neuropsychology evaluations. Specific changes included adding neuropsychology evaluation appointments the morning of spina bifida clinic, conducting the intake assessment and feedback session via telehealth, providing education about neuropsychology evaluations to spina bifida clinic staff, and formation of a clinical research partnership among neuropsychologists and spina bifida clinicians.

Results: We compared the volume of neuropsychology referrals and completed evaluations in 2018-2019 to 2021-2022. In 2018-2019 there were 52 referrals and 24 completed evaluations, a 46% completion rate. In 2021-2022 (YTD) the number of referrals increased 21% to 63 referrals. In 2021 there were 32 referrals and 19 completed evaluations, a 59% completion rate. We also compiled a case study highlighting the significance of periodic neuropsychology evaluations throughout childhood and adolescence to support cognitive and adaptive function growth.

Conclusions: The QI initiative implemented in spina bifida clinic increased the number of neuropsychology referrals and evaluations for patients with spina bifida. We plan to explore the demographic variables to consider possible barriers to neuropsychology evaluation for future system enhancements. Further studies are planned to determine the impact of neuropsychology testing on the cognitive and adaptive outcomes of patients with spina bifida.

Trends in neurodevelopmental outcomes in children after fetal and postnatal myelomeningocele closure

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Background: Studies on children with myelomeningocele have demonstrated decreased neurocognitive function compared to age matched cohorts without myelomeningocele, and impaired motor function. We examined changes in neurodevelopmental outcomes in this patient population.

Methods: Our institutional registry was reviewed for patients who had undergone prenatal and postnatal myelomeningocele closure between 2011 and 2021 and underwent neurodevelopmental assessments via Bayley Scales of Infant Development. Spline regression model was used to assess change in outcomes over time, adjusting for gestational age and gender.

Results: A total of 105 patients were included; 48 (45.7%) female. Average gestational age at birth was 35.5 (SD=2.38), 62 (60%) were born before 37 weeks and 8 (7.6%) before 32 weeks. Fetal surgery was performed on 89 (84.8%), and 33 (31.4%) underwent CSF diversion (4 ETV, 29 VPS). Mean assessment ages were 6.4 months (SD=0.94), 14.2 months (SD=3.1) and 31.5 months (SD=5.9). We found a significant increase in composite cognitive score from 6 to 12 months (β=0.82, **p=0.033**); when we stratified the cohort, this increase was limited to patients who did not undergo CSF diversion (**p=0.014**). Composite language and receptive communication significantly increased from 6 and 12 months (β=1.05, **p=0.029** and β=0.22, **p=0.027**,

respectively). Composite motor scores significantly decreased between 6 and 12 months (β=-0.93, **p=0.013**) and 12 and 30 months (β=-0.25, **p=0.042**). While fine motor scores increased between 6 and 12 months (β=0.22, **p=0.005**), gross motor scores declined in that period (β=-0.51, **p<0.001**). When comparing fetal and postnatal cohorts, composite language at 6 months was significantly higher in the postnatal group (**p=0.042**), but this difference disappeared by 30 months.

Conclusions: Among all patients with myelomeningocele, cognitive and language outcomes improve in the first year of life, particularly in patients not requiring CSF diversion. Motor outcomes initially decline, before stabilizing. Larger scale studies on predictors of improved neurocognitive outcomes in myelomeningocele patients are warranted.

Impact of prenatal versus postnatal closure on cognitive and adaptive outcomes in children with myelomeningocele

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Background: Prenatal closure of myelomeningocele has transformed the early management of children with spina bifida over the past decade. However, little is known about the adaptive and cognitive outcomes of children treated with prenatal versus postnatal surgery. The aim of the present investigation was to examine differences in functioning in early childhood between children whose closure was conducted in the prenatal versus postnatal period.

Methods: The present sample included 27 children with myelomeningocele (M_{age}=37.1 months, SD_{age}=12.6 months; $range_{age}$=18-58 months; 48.1% female; 92.6% white). Medical and sociodemographic data were extracted from patient medical records and coded for closure status (i.e., prenatal closure [33.3%] vs postnatal closure [66.7%]). The following measures were included in analyses: Wechsler Preschool and Primary Scale of Intelligence-Fourth Edition; Mullen Scales of Early Learning; and the Adaptive Behavior Assessment System-Third Edition. Independent-sample t-tests and chi-square tests were conducted to examine differences on neurocognitive, adaptive, and sociodemographic variables between groups.

Results: Intellectual functioning (M_{SS}=89.2, SD=14.2) and adaptive functioning (M_{SS}=86.2, SD=18.7) fell in the low average range. Financial class differed between groups, such that children with commercial insurance were significantly more likely to receive prenatal surgery than children with Medicaid or Medicaid HMO ($\chi^2[2]=7.94$, $p=.019$, $\varphi_c=0.54$). Children with prenatal closure were more likely to be born pre-term, with an average gestation of 33 weeks (t(25)=3.12, p=.005). Neither intellectual functioning (t(16)=0.01, p=.996) nor adaptive functioning (t(24)=-1.60, p=.124) differed between children with prenatal versus postnatal closure.

Conclusions: In the present sample, prenatal surgery for myelomeningocele was not associated with improved intellectual functioning nor adaptive functioning during the early childhood period. These initial results do not support neurocognitive developmental benefits of prenatal closure, despite motor functioning benefits. Future work is needed to investigate the influence of other medical variables (e.g., functional level of lesion, shunt status) in a larger and more diverse sample.

Neurodevelopmental outcome in infants with spina bifida myelomeningocele undergoing endoscopic third ventriculostomy combined with choroid plexus cauterization for hydrocephalus: A single center experience

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Background: Little is known about the neurodevelopmental (ND) outcome in spina bifida myelomeningocele (SBM) infants treated with endoscopic third ventriculostomy/choroid plexus cauterization (ETV/CPC) and the relationship between surgical and ND outcomes.

Methods: This is a retrospective study of SBM infants born between 2010 and 2019 and followed clinically at Boston Children's Hospital. Patients were grouped by surgical outcome as untreated, ETV/CPC and those that are shunted. ND outcomes were Bayley Scales of Infant and Toddler Development 3rd edition (BSID-3) and Vineland Adaptive Behavior Scales 3rd edition (Vineland-3) administered at approximately 24 months age.
Results: Fifty SBM patients included. Age at initial neurosurgery was significantly younger in the shunted group than ETV/CPC. Group results are in Table 1. The mean BSID-3 composite scores of the untreated group were lower than population norms except for higher social emotional score. No significant differences were found between the untreated and the ETV/CPC groups. The shunted group has lower scores on BSID-3 Cognitive and Motor domains than untreated and ETV/CPC groups. For Vineland-3, the shunted group had significantly lower scores than the ETV/CPC group in adaptive behavior composite, personal, and gross motor skill domains and trended lower in others. ETV/CPC patients also had significantly lower scores than the untreated group in personal and gross motor skill domains.
Conclusions: The SBM patients had scores lower than population norms in most domains, whether treated or not. SBM infants requiring a shunt for hydrocephalus had significantly lower BSID-3 scores than other groups. Data collection is still on going.

Associations between neuropsychological functioning, bowel and bladder continence, and independent self-management in adolescents with spina bifida

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Background: The ability to perform clean intermittent catheterization (CIC) and bowel programs is a key self-management goal for adolescents with spina bifida (SB). While cognitive function, neurologic level of lesion, age, and gender play a role, prior literature has shown inconsistent associations with independent self-management and continence. We examine the relationship between neuropsychological functioning and independent self-management in adolescents with SB in a tertiary clinic population.
Methods: The sample included 32 older children and adolescents with SB (M_{age}=14.1 years, SD_{age}=2.0 years; $range_{age}$=8.8-18.6 years; 40.6% females; 81.3% white, 9.4% Hispanic/Latinx, 6.3% Asian, 3.1% Black). Medical records were reviewed and data regarding continence/independence status and neuropsychological functioning in areas of working memory, processing speed, sustained attention, and adaptive functioning skills were extracted. Independent-sample t-tests were used to examine group differences between neuropsychological functioning and continence/independence.
Results: This sample demonstrated impairment in working memory (M_{ss}=6.7), processing speed (M_{ss}=4.5), and adaptive functioning (M_{SS}=77.3). Performance across indices of sustained attention fell in the average range. Independent CIC was significantly associated with certain sustained attention indices: specifically, reaction time (t(19)=2.79, p=.012) and reaction time variability (t(19)=3.14, p=.005), such that slower response speed and difficulty sustaining response speed across time were associated with non-independence with CIC. No other variable was related to any other independence or continence variable.
Conclusions: Certain aspects of sustained attention including response speed were associated with greater independence with CIC, but not independence with a bowel program or continence. Contrary to hypothesis, processing speed was not associated with independence or continence status; this may be due to the impairments in processing speed across this entire sample. The relationship with sustained attention may represent the increased variability of attentional capacity in individuals with SB possibly related to history of hydrocephalus and need for shunting. Future studies should examine the possibility of a moderated relationship.