Neuropsychology

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Big Picture

• Core deficits in timing, movement, and attention are present from birth and lead to problems with assembled processing (constructing and integrating information); associative processing (rule-based learning) preserved.

• Related more to malformations (spinal lesion, Chiari II, callosal hypogenesis, midbrain anomalies) than to hydrocephalus.
Modal Neuropsychological Profile

- Typical pattern of stronger and weaker skills
  - Observed in many individuals with spina bifida myelomeningocele and hydrocephalus
  - Apparent over the lifespan
- Variability
  - Differences between individuals is principled
  - Related to moderators in genetic, brain, medical history; poverty, parenting, and perhaps teaching
- Determined by Core Processing Deficits (Timing, Attention Orienting, Movement)
  - Assembled vs. Associative processes

(Dennis et al., 2006; Dennis & Barnes, 2010)
Associative Processing is data-driven and based on the formation of associations, enhancement, engagement, and categorization: recognizing faces, decoding words, using vocabulary, carrying out routines.

Assembled Processing, is based on dissociation, suppression, disengagement, and contingent relations. It requires the assembling of models of input across various content domains: inferring, constructing meaning, integrating knowledge, problem solving, learning routines.

(Dennis et al., 2006)
Core Processing Deficits Interfere with Assembled Processing

- Timing: Synchronizing behavior with the environment (Cerebellum)
- Attention Orientation: Automatic direction and disengagement of attention to the environment (Midbrain)
- Movement: Learning and controlling coordinated movements (Spinal lesion, Cerebellum)
- Operate across and within domains; cognitive difficulties poorly understood as differences in cognitive content

*Associative processes preserved unless secondary factors present (poverty, severe hydrocephalus)*
Modal Cognitive Profile: Strengths and Weaknesses

Associative Processing: Strengths
- PERCEPTION: categories
- LANGUAGE: vocabulary
- READING: word decoding
- MATH: numbers
- BEHAVIOUR: sociability

Assembled Processing: Weaknesses
- PERCEPTION: representations
- LANGUAGE: constructing meaning
- READING: text comprehension
- MATH: algorithms
- BEHAVIOR: adaptation
Model (Dennis et al., 2006; 2010)

Domain General

Timing
Attention
Movement

Domain Specific

Perception
Language
Literacy
Numeracy
High Variability in the Neural Presentation

A) Corpus Callosum  B) Tectum  C) Cerebellum
Overall Expected Outcomes

• Optimal development of language, academic, and other learning skills.
• Optimal performance in school, university, and vocational settings.
• Acquisition of a job.
• Maximize participation in society.
• Maximize independence according to individual capabilities.
Functional Outcomes

• Utilization of learning skills is apparent in a variety of contexts.
• Enrollment in early pre-school intervention.
• Enrollment in appropriate educational environment.
• Participation in vocational assessment and training.
Clinical Question: What do teachers, psychologists, and other professionals need to know about the development of people with Spina Bifida?

Guidelines:
1. All individuals need to understand that if spina bifida is associated with myelomeningocele, it is not simply an orthopedic disorder (“physical disability”). The brain malformations and hydrocephalus (with or without shunting) affect learning.
2. Full neuropsychological evaluations that includes assessment of early literacy and numeracy skills can help to provide a more comprehensive understanding of strength and weakness profiles, as well as the significant discrepancies often present in their profiles. These findings can be related to neuroimaging findings for a more careful determination of the basis for any difficulties experience by the child.
3. It is important to advocate for eligibility for special education services under a broad rubric or other category that emphasizes the need for ongoing care and attention to changes in brain development, not one isolated domain of concern.
Age related clinical questions and guidelines
Clinical Question: What early interventions in infancy are appropriate for supporting the development of motor, cognitive, and early literacy and numeracy skills?

Guidelines:
1. EI prior to initial discharge (OT, ST, and PT), or outpatient therapy as needed
2. Parents receive adequate information and supportive services
3. Promotion of effective parenting practices for infant stimulation and care giving
4. Appropriate placement in family-based infant development programs as early as possible through state and public school (IDEA Part C, 619) resources.
5. Facilitation of parental participation in infant development programs (Individualized Family Service Plan, IFSP)
6. Monitor and evaluate onset and progression of physical, cognitive, communicative, and social development
Infancy

Clinical Questions: How is the health and development of infants and toddlers changing with prenatal surgery (MoM trials)? How is the health and development of infants changing with the use of new surgical procedure such as Endoscopic Third Ventriculostomy/Choroid Plexus Cauterization (ETV/CPC) rather than shunting? How does monitoring for hydrocephalus and delayed shunting influence development?

Guidelines:
1. For infants who have untreated hydrocephalus and developmental regression, or slower than expected development, shunt placement/ETV should be discussed with the neurosurgeon.
2. For infants repaired prenatally, close monitoring is indicated given paucity of literature on their long-term outcomes. (Adzick, 2011)
3. For infants who have not yet been surgically treated for hydrocephalus, but are monitored due to risk, assessment using infant development scales involving language, motor, and social development is warranted. Adaptive behavior assessments that are interview-based are easy to complete and sensitive to growth trajectories in development (Taylor et al., 2011).
Infancy

- **Clinical Questions** How can teams utilize early MRI findings (e.g., malformations, dysplasia, reduced volume, and agenesis) to predict domains of risk and identify potential early interventions to support development?

- **Guidelines:**
  1. For infants who are in a high risk category, which includes those born prematurely secondary to prenatal surgery intervention and those with significant changes in brain development (above Chiari II malformation), the risk for early difficulties with coordinated upper limb motor movements and attentional focus is especially higher with tectal beaking and greater severity of the Chiari.
Clinical Question: What early interventions in toddler years are appropriate for supporting the development of motor, cognitive, and early literacy and numeracy skills?

Guidelines:
1. Monitor and evaluate onset and progression of physical, cognitive, communicative, and social development.
2. Support parents in effective interactions with child that facilitate movement and exploration, language and communication, and play.
3. Equipment that facilitates object exploration and manipulation can be helpful.
4. Early interventions for language difficulties if delayed in onset, characterized by articulation difficulties, or unusual in pattern of development. The use of sign language is recommended in conjunction with verbal speech therapy.
5. PT and OT for independent mobility, strengthening, and functional activities with parental involvement.
6. Encouragement to participate in group learning experiences for children, especially when families are unable to find available day care that attends to necessary medical needs; either through community groups or early intervention.
Toddler

• **Clinical Questions:** How is the health and development of infants and toddlers changing with prenatal surgery (MoM trials)? How is the health and development of toddlers changing with the use of new surgical procedure such as Endoscopic Third Ventriculostomy/Choroid Plexus Cauterization (ETV/CPC) rather than shunting? How does monitoring for hydrocephalus and delayed shunting influence development?

• **Guidelines:**
  1. Children of parents with higher expectations who facilitate attention, require movement, and support language development have better outcomes later in development.
  2. Monitoring of hydrocephalus must be based on developmental assessments beyond determination of milestones, which are weak indicators of developmental difficulties.
     a) Periodic assessments with age appropriate assessments of early language skills can help identify more subtle difficulties with development.
     b) Ongoing monitoring of coordinated upper limb movement and attention is essential in children with greater severity of Chiari, tectal beaking, and callosal hypogenesis.
Preschool

• **Clinical Question:** How does the relation between the nervous system and mental functions among patients with Spina Bifida affect their learning in reading, mathematics, writing, social science and science, and how does it affect them at different developmental stages?

• **Guidelines:**
  1. Children with more severe hydrocephalus, hypogenesis and/or severe hypoplasia of the corpus callosum and history of CNS infection are at greater risk for difficulties involving construction of meaning from language (Bradley et al., 2016; Huber-Okrainec et al., 2005).
  2. It is important that teams advocate for children to enter formal educational programs through the public schools in order to have access to the scaffolding that can support the development of attention, self regulation, social interaction skills, and independent functioning skills. If parents choose another school setting then recommendations for support services and supplemental resources should be provided and accessed.
School age

• **Clinical Question:** How does the relation between the nervous system and mental functions among patients with Spina Bifida affect their learning in reading, mathematics, writing, social science and science, and how does it affect them at different developmental stages?

• **Guidelines:**
  1. Children with spina bifida myelomeningocele, particularly those with hydrocephalus or other changes in brain development, would benefit from a full neuropsychological assessment.
  2. Although school assessments can be helpful to track global intellectual and academic progression, they rarely assess the development of essential skills in attention, executive functioning, coordinated upper limb, and memory domains, as well as adaptive skill acquisition.
Clinical Question: What interventions in school-age children support their cognitive development and academic achievement?

Guidelines

1. Reinforce use of learning activities and technology in home.
2. Promote child’s participation in school-related and extra-curricular activities.
3. If a child has a problem with reading or math, interventions like those used with children with learning disabilities are often effective.
4. Writing programs need to be implemented using assistive devices as early as possible.
5. Bladder and bowel continence needs to be addressed and aggressively managed as improved continence translates to greater independence and social participation.
6. Evaluation and treatment for attention and behavioral difficulties, including didactic teaching of social skills if necessary. Evaluations for ADHD should follow American Academy of Pediatrics guidelines.
Teenage

- **Clinical Question:** How does the relationship between the nervous system and mental functions among patients with Spina Bifida affect their learning in reading, mathematics, writing, social science and science, and how does it affect them at different developmental stages?

- **Guidelines:**
  1. Interventions addressing integration and assimilation of information with a specific focus on reading comprehension and mathematics problem solving. Intervention programs should be maintained because the absence of intervention is associated with plateaus in skill development in most populations with disabilities.
  2. Vocational and transitional services with a focus on independence.
  3. Promoting participation in school related and extracurricular activities.
  4. Obesity and bladder/bowel continence can be major issues affecting social adjustment and should be aggressively addressed. (Fischer, Church, Lyons, & McPherson, 2015)
Clinical Question: What interventions and programs in adolescence provide smoother transitions to post-secondary education and/or career/vocational training?

Guidelines:
1. It is essential to have an established transition plan built into patient’s IEPs in order to ensure that appropriate referrals are made to adult agencies, appropriate life and vocational skill training occurs.
2. Social skills training may be useful given the tendency of many individuals with spina bifida to become withdrawn and to experience reduced social interactions as they enter adolescence.
• Clinical Question: What do teachers, psychologists, and other professionals need to know about the development of people with Spina Bifida?

• Guidelines:
  1. In addition to the cognitive and learning problems associated with the underlying neurological disorder, adolescents with spina bifida are vulnerable to feelings of depression secondary to reduced quantity and quality of social interactions. Structured opportunities for social interaction through school, church, and afterschool opportunities may be needed to decrease a potentially higher risk for depression. (Essner & Holmbeck, 2010)
• Clinical Question: How do treatment teams help prepare all of their patients for the transition to adulthood, and the assumption of their own medical care? What indicators are helpful to team in identifying those who may require ongoing support for adequate management of their medical conditions?

• Guidelines:
  1. If patients have not been fully evaluated, it is essential that a full neuropsychological assessment be completed prior to the child’s 18th birthday. These results insure that if a diagnosis of an intellectual disability is relevant, it occurs in a timely fashion.
  2. Full assessments are also essential to help identify cognitive strengths and weaknesses for those who are assuming responsibility for their own medical care. Although efforts should begin as early as possible, it may be necessary to work with the patient for several years to learn the skills necessary to understand and take responsibility for their own medical care. The transition to adolescence may present new challenges to this learning and regression may be present.
Adults continued

• **Additional Guidelines:**
  1. Vocational services addressing job skills, additional education, and related activities.
  2. University-based ADA/504 services for people with disabilities.
  3. Monitoring of cognitive skills, especially math (Dennis et al., 2007), memory (Treble-Barna et al., 2014b), and attention, to ensure maintenance of learning skills essential for work and independence. Changes may be a sign of unidentified shunt failure or shunt dependency.
  4. Transition from parent focused interventions and communication strategies (e.g., phone calls to nurse) to methods that may be easier for the young adult to utilize consistently (e.g., online portals, email).
  5. Skill building will be essential in order for young adults to shift to the adult care model where less coordination of services is often provided.
Research Gaps

• How well do interventions used across the lifespan involving cognition, learning, and social skills work with the person with spina bifida?

• How are attention problems best treated from pharmacological and non-pharmacological perspectives?

• What is the long-term effect of sequential monitoring of hydrocephalus on development? Is it better to shunt early and control hydrocephalus or to monitor ventricular expansion over time? What are the best indicators of the need for shunt diversion?
• Guidelines also include tips on:
  • risk factors
  • recommended timing for evaluations
  • discerning ADHD from more typical attention problems
  • concerns about EF/initiation/integration issues

• For references, please see full guidelines