Spina Bifida and Hydrocephalus Across The Lifespan: Part 2 – Adult

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Outline

1. Lifespan issues
   a) Adult math and functional numeracy
   b) Reaction time
   c) Sleep and circadian rhythms

2. New adult challenges
   a) Prospective memory
   b) Depression

3. What does hydrocephalus add to adult profile?

4. Implications.
Lifespan issues
Preschool, childhood numeracy

PRESCHOOLERS
- Counting
- Object-based addition, subtraction

SCHOOL-AGED CHILDREN
- Numeration and rational numbers
- Single digit addition, subtraction accuracy, speed
- Multi-digit addition, multiplication accuracy, speed
- Single digit addition speed, calculation strategies
- Multidigit subtraction
- Division
- Estimating and problem solving.
Adult numeracy

• LONGITUDINAL
  • Children with spina bifida poor at computation, math problem-solving, grow into adults with same problems.

• CROSS-SECTIONAL
  • Young adults with spina bifida have poor computation accuracy, computation speed, math problem-solving.
  • Difficulties with functional numeracy (supermarket price comparisons, estimating quantities, prices, reading prescriptions).
Poor numeracy matters

• EVERYDAY FUNCTION
  • Difficulty using numbers in life.
  • Limits financial independence (holding bank accounts, taking medications, managing personal finances).

• Adult functional numeracy predicts independence:
  • Social
  • Personal
  • Community.
Reaction time

- Simple RT controls
- Choice RT controls
- Cognitive RT controls

Simple RT spina bifida
Choice RT spina bifida
Cognitive RT spina bifida

Age (years)

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Sleep and chronotype

• **SLEEP**
  - Children with spina bifida report more problems than controls (daytime sleepiness; fatigue; poor energy).
  - Adults with spina bifida report more problems than controls (trouble falling asleep; restless/disturbed sleep).

• **CHRONOTYPE**
  - Munich Chronotype Questionnaire measures phase relationship between timing of sleep and 24-hour day.
  - Mid-point of sleep on “free” days, i.e., days without waking for school/work, corrected for sleep duration (MSFsc).
  - MSFsc correlated with objective measures of circadian timing and sensitive to developmental changes, sleep pathologies.
• Over lifespan, individuals with spina bifida (crosses, dotted line) show typical relationship between chronotype and age: as children, go to bed and wake early; as adolescents, sleep and rise times are later; as adults, sleep and rise earlier.
• Phase relationship between sleep-wake timing and 24 hour day in controls (circles, solid line); curve in controls trends downward when it peaks in SBM.
• Aberrant synchronization of circadian rhythms to 24h day in SBM may contribute to reported sleep problems.
New adult challenges
Prospective “TO DO” Memory

Event-based tasks
- tell examiner about 5 hidden objects when beeper goes off.
- hand examiner book after specific quiz question about TV program.

Time-based tasks
- tell examiner to make phone call at specific time cued by clock.
- remind examiner not to forget keys “when there are 7 min left” on timer.
• Young adults with spina bifida score lower than age peers, for event- or time-based tasks.

• Poor prospective memory three times higher in older spina bifida group (over 32 years; 37.50%) than in younger spina bifida group (18-31 years; 12.50%).
Neural basis of prospective memory

- Involves frontal, temporal lobe.
- Spina bifida involves:
  - atypical development of frontal lobes
  - thinning of key white matter tracts like uncinate fasciculus connecting inf. frontal, ant. temporal
  - hippocampi rotated outward and laterally dislocated
  - elevation of medial parahippocampal gyri
  - abnormal sulcation in mesiotemporal cortex.
Temporal lobe

- Mesial temporal cortex abnormal in size, shape 85-93%
- Hippocampi rotated outward, laterally dislocated
- Primary hypoplasia + hydrocephalus effects on hippocampus, connecting fibres
- Parahippocampal gyri elevated medial to hippocampus, not inferior

Beak: Fimbria
Head: Hippocampus
Neck: Horizontal subiculum
Torso: Parahippocampal gyrus
Arcuate fasciculus in spina bifida

- Abnormal development in spina bifida
- LEFT Arcuate AFT segment less myelinated.

ARCUATE FASCICULUS
Segments: fronto-temporal (AFT), fronto-parietal, temporo-parietal

(Hasan, Eluvathingal, Kramer, Ewing-Cobbs, Dennis, Fletcher et al., J. Magnetic Res Imag, 2008)

Depression

% at or above cut-off for clinical depression (SBM N=229; CON N=96)
What does hydrocephalus add to adult profile?
Hydrocephalus in spina bifida

• Affects brain development, contributes to dysmorphologies.
  • The Chiari II malformation
  • Corpus callosum hypoplasia.

• Contributes to abnormalities of association fibres:
  • macrostructure (poor visualization)
  • microstructure (↓ fractional anisotropy, ↑ diffusivities)
    • impaired myelination (↑ transverse diffusivity)
    • abnormal axonal features (↑ axial diffusivity).

• ~50 studies of no association IQ and shunt revisions. But:
  • Small N
  • No separation of type of shunt revisions, spina lesion level (thoracic vs. lumbar)
  • Simple linear correlations, no statistical modeling.
Hydrocephalus and adult outcome

• In adults, number of shunt revisions negatively related to:
  • functional numeracy
  • some memory functions
  • independent living
  • employment.

• Young adults with spina bifida and nonfunctioning shunts or non-shunted ventriculomegaly show improved memory after shunt treatment.
Implications
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1. Recognize lifespan challenges that affect daily life more in adulthood than in childhood.

1. For individuals 18 years and older, encourage continuing education in life skills mathematics.

1. Encourage awareness and strategies (note taking helped spina bifida with prospective memory tasks).

2. Access to appropriate care (e.g., for depression).

3. Hydrocephalus affects function more in adulthood than in childhood, so childhood shunt management and adult shunt function continue to be important.
Groups vs. individuals

• Group research helps to:
  • Write journal articles, get tenure, adopt position.
  • Hope University will provide chair to adopt position in.

• But intervening to create a more positive medical, physical, or cognitive lifetime outcome requires individual prediction of ecologically salient function.
Grant Support

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Hydrocephalus status and outcome

- Neurocognitive function in typically developing (N=61) children, children with spina bifida and shunted hydrocephalus (N=166), arrested hydrocephalus (N=18), or no obvious hydrocephalus (N=24).
• ‘Hot’ colors indicate cortical thickness or complexity GREATER in the SBM group relative to age peers; “cold” indicate LESSER in the SBM group.

• ALTERED PATTERNS OF LOCAL AND LONG-RANGE CORTICAL CONNECTIVITY

• SOME CORTICAL REGIONS TOO FAT, SOME TOO THIN.