Neuropsychology in Spina Bifida

Dr Ellen Northcott
Clinical Neuropsychologist
Kids Rehab, CHW
Who are neuropsychologists?

Undergraduate Degree (e.g. BPsysch, BSc, BA) → Honours in Psychology

Master or Doctor of Clinical Neuropsychology
  Coursework
  Thesis
  Placements

Registrar Program
Neuropsychological Assessment

- Interview
- Observation
- Testing
- School Report
Spina Bifida

- Congenital malformation due to incomplete neural tube closure.
- Wide variety of cognitive deficits have been reported.
- Possible impacts on cognition:
  - Arnold Chiari-II malformation
  - Hydrocephalus
  - Underlying brain dysfunction
Cognitive Difficulties

• Intellectual ability
• Language skills
• Visuo-spatial skills
• Attention
• Executive functioning
• New learning and memory
• Academics
Intellectual Abilities

• Most commonly assessed using the WPPSI-III, WISC-IV or WAIS-IV
• Overall ‘IQ’ score consists of verbal abilities, nonverbal abilities, processing speed and working memory.
• In spina bifida verbal abilities are generally better than nonverbal abilities
WISC-IV

• Five index scores
  – Full scale Intellectual Quotient (FSIQ)
  – Verbal Comprehension Index (VCI)
  – Perceptual Reasoning Index (PRI)
  – Working Memory Index (WMI)
  – Processing Speed Index (PSI)
Description of results

• Very Superior
• Superior
• High Average
• Average
• Low Average
• Borderline
• Extremely Low
Case Example

- 5 year old boy with Spina Bifida and Osteogenesis Imperfecta
- Shunt infection a few months prior to assessment
- Average verbal intellectual abilities
- Low Average nonverbal intellectual abilities
- Particularly poor on visuo-constructional task
Visuo-spatial skills

• Involved in analysing and understanding space.
• Involve mental imagery and navigation.
• Used to copy drawing, or use pieces to construct objects of shapes.
• Let you judge location and speed of objects (eg. for crossing roads).
Visuo-spatial skills

• Copying geometric figures
• Drawing people
• Matching patterns
• Constructing patterns using blocks
Attention

• Selective
• Sustained
• Switching
• Divided

• In children assessed with TEA-CH
Attention

- Poor persistence in classroom activities
- Difficulty following instructions
- Difficulty staying on task
- Impulsivity and distractibility
- Inability to focus is a busy environment
- Difficulty adapting to changes in classroom
- Difficulty carrying out tasks in allotted time
- Difficulty keeping up with peers in conversation
- Difficulty contributing to classroom discussions
Memory and Learning

- Learning
- Storage
- Retrieval
Executive Functioning

- Used to describe a number of cognitive processes.
- Responsible for controlling goal-directed cognitive, behavioural and emotional processing.
- Can present in a variety of ways.
Executive Functioning

- Anticipation
- Goal Selection
- Planning
- Initiation of activity
- Self-regulation
- Mental flexibility
- Responding to feedback
- Regulating emotion
- Controlling impulses

- Organisation
- Problem Solving
- Divided attention
- Insight
- Abstract thought
- Generating ideas
Executive Functioning

- A large number of tests are available:
  - Behavioural Assessment of the Dysexecutive Syndrome in Children (BADS-C)
  - Delis Kaplan Executive Function System (D-KEFS)
  - Wisconsin Card Sorting Test
  - NEPSY-II subtests
  - Tower tests
Executive Functioning

• Limitations of tests as the assessment setting is:
  – Well structured
  – Minimal distractions
  – Prompts to start tasks
  – Clear instructions on how to approach tasks

• Importance of interview, observation and questionnaires.
Case Example - NS

- 7 years 3 months
- Sacral myelomeningocele and Arnold Chiari II malformation.
- Myelomeningocele repaired on day 2.
- Shunt inserted at 6 weeks of age.
- Support class with very impaired (non-verbal) children.
- Unable to complete WISC-IV, so WPPSI-III was administered.
Case Example - NS

- Mild intellectual impairment.
- Attention poor even in context of IQ.
- Unable to write his name or read simple words.
- Reduced expectations, so limited awareness of attention and executive functioning deficits.
Language

• Superficially often have good conversational skills
• But difficulties with:
  – Understanding instructions
  – Coping with abstract language
  – Deriving meaning from text
  – Generating and organising coherent written and verbal language
Hydrocephalus Status

• Hampton et al (2011) looked at impact of hydrocephalus status on cognitive functioning.
• They compared shunt-treated (166), arrested-hydrocephalus (18), no hydrocephalus (24) and controls (61).
Hydrocephalus Status
Arnold-Chiari II Malformation

- Differences between the groups on all domains.
- After excluding children with intellectual impairment results remained significant for:
  - Visual analysis and synthesis
  - Verbal Memory
  - Verbal Fluency
Structural Abnormalities

• Kumar et al (2011) looked at 13 patients aged 10-17 years without hydrocephalus or Chiari malformation.

• Used diffusion tensor imaging with values reported for fractional anisotropy and mean diffusivity.
Structural Abnormalities
Structural Abnormalities

• Correlations with fractional anisotropy values:
  – Genu with NCT B.
  – Middle cerebellar peduncle with picture arrangement

• Correlations between mean diffusivity values:
  – Splenium with NCT-A
Conclusions

• Wide variety of cognitive deficits.
• Deficits have been linked with hydrocephalus, Arnold-Chiari II malformation and subtle structural abnormalities.
• Therefore for any one child there are a number of factors interacting which may contribute to their cognitive profile, leading to a great deal of individual variation.
Questions?
References


