Neuropsychology in Spina Bifida

Dr Ellen Northcott
Clinical Neuropsychologist
Kids Rehab, CHW

Who are neuropsychologists?

Undergraduate Degree (eg. BPsych, 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

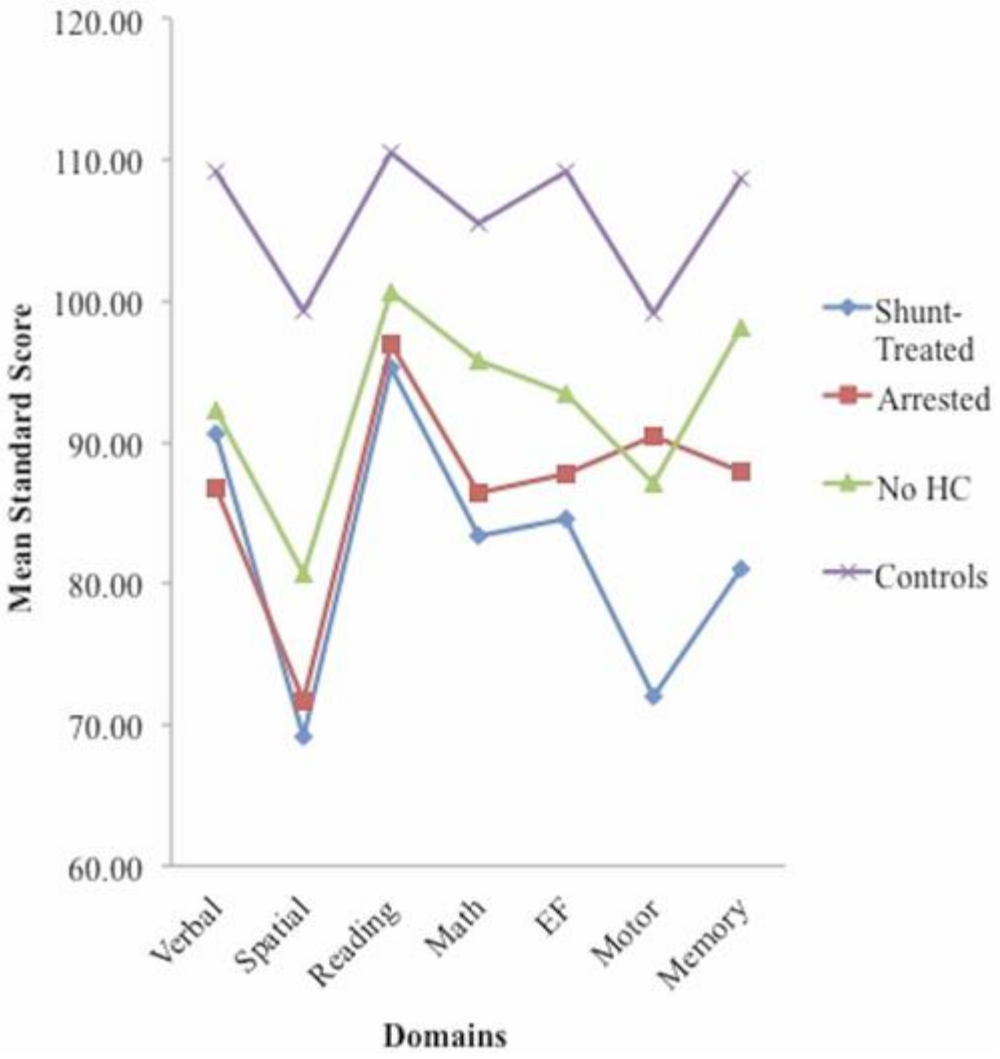
- Mild intellectual impairment.
- Attention poor even in context of IQ.
- Unable to write his name or read simple words.
- Reduced expectations, so limited awareness of attention and executive functioning deficits.

Language

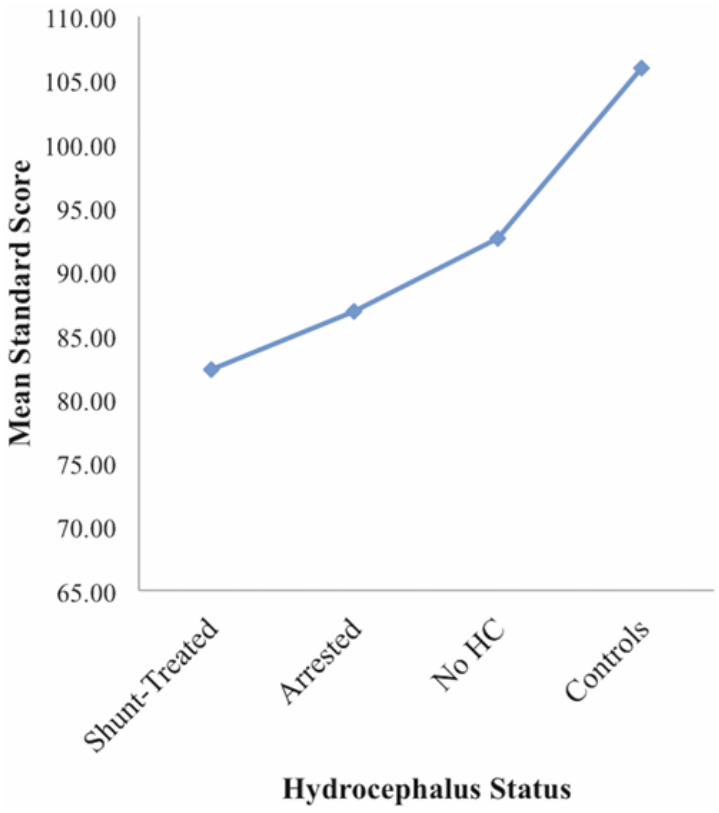
- Superficially often have good conversational skills
- But difficulties with:
  - Understanding instructions
  - Coping with abstract language
  - Deriving meaning from text
  - Generating and organising coherent written and verbal language

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).


120.00
110.00
100.00
90.00
80.00
70.00
60.00
Mean Standard Score
Verbal
Spatial
Reading
Math
EF
Motor
Memory
Domains
Shunt-Treated
Arrested
No HC
Controls

Hydrocephalus Status

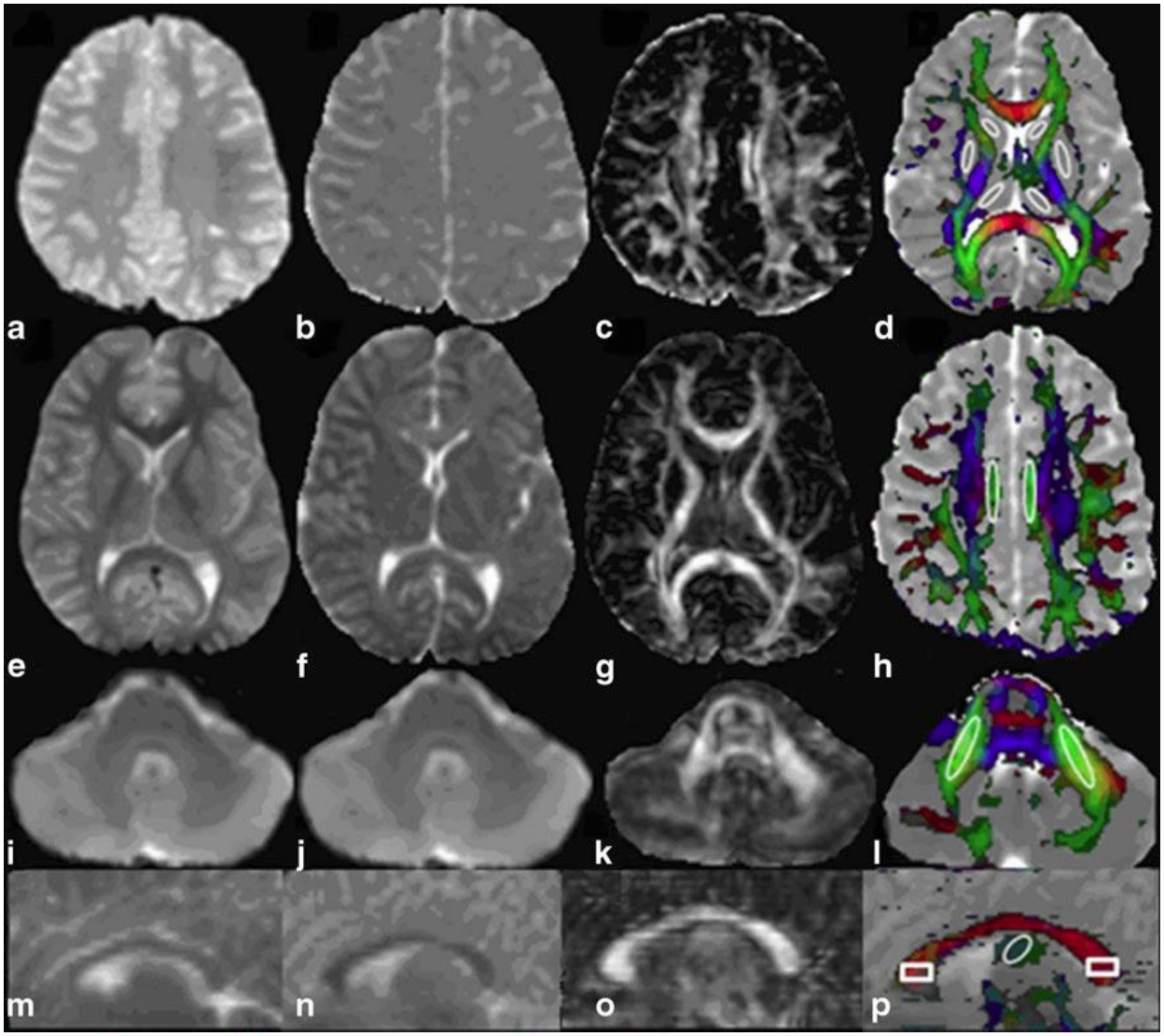


Arnold-Chiari II Malformation

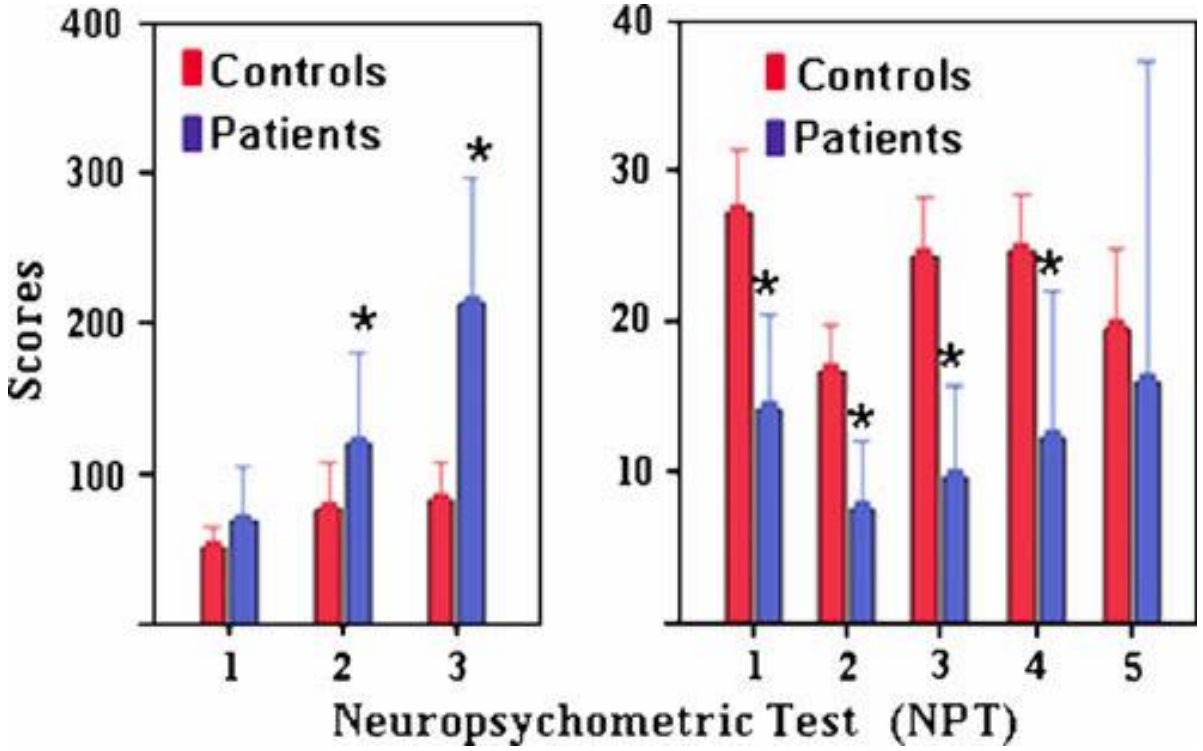
- Vinck et al (2006) compared 19 children without AC-II malformation and 27 with.
- Differences between the groups on all domains.
- After excluding children with intellectual impairment results remained significant for:
  - Visual analysis and synthesis
  - Verbal Memory
  - Verbal Fluency

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.


a
b
c
d
e
f
g
h
i
j
k
l
m
n
o
p

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.
- Therefor 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

- Hampton et al (2011) Hydrocephalus status in spina bifida: an evaluation of variations in neuropsychological outcomes. *J Neurosurg Pediatrics 8:289-298*
- Kelly et al (2012) Executive functioning and psychological adjustment in children and youth with spina bifida*. Child Neuropsychology 18 (5), 417-431*
- Dennis and Barnes (2010) The cognitive phenotype of spina bifida meningomyelocele. *Dev Disabil Res Rev 16 (1): 31-39*
- Kumar et al (2011) Cognitive functions correlate with diffusion tensor imaging metrics in patients with spina bifida cystica. *Childs Nerv Syst 27:723-728*
- Vink et al (2006) Arnold-Chiari-II malformation and cognitive functioning in spina bifida*. J Neurol Neurosurg Psychiatry 77:1083-1086*