

# 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 in 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 (BADSD-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

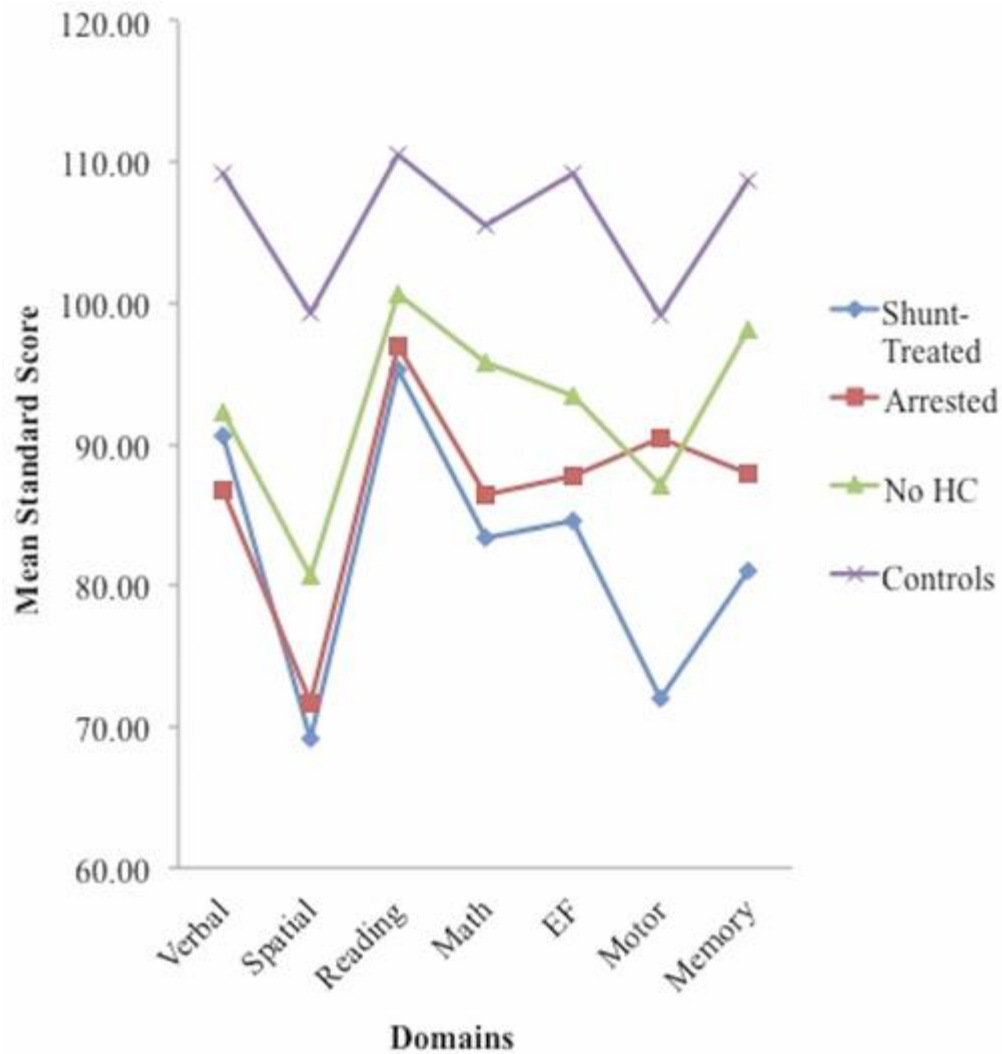
- Superficially often have good conversational skills
- But difficulties with:
  - Understanding instructions
  - Coping with abstract language
  - Deriving meaning from text
  - Generating and organising coherent written and verbal language



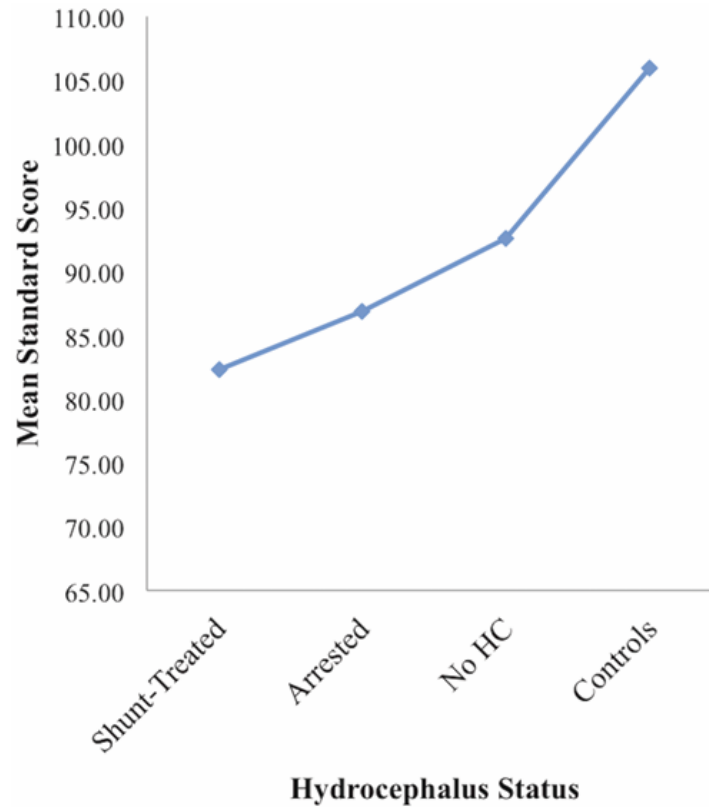
## Hydrocephalus Status

- 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

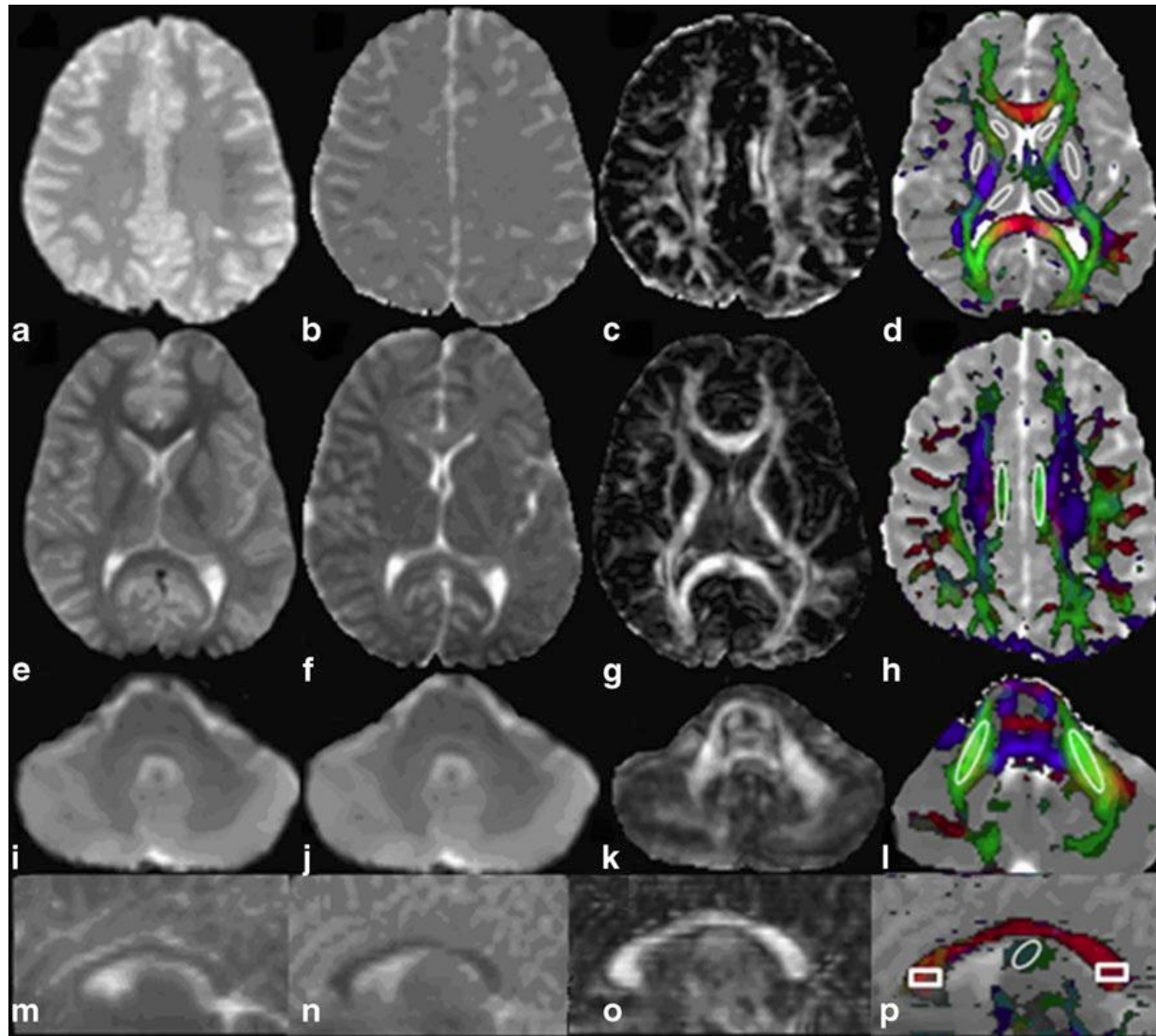
- Vinck et al (2006) compared 19 children without AC-II malformation and 27 with.
- Differences between the groups on all domains.
- After excluding children with intellectual impairment results remained significant for:
  - Visual analysis and synthesis
  - Verbal Memory
  - Verbal Fluency



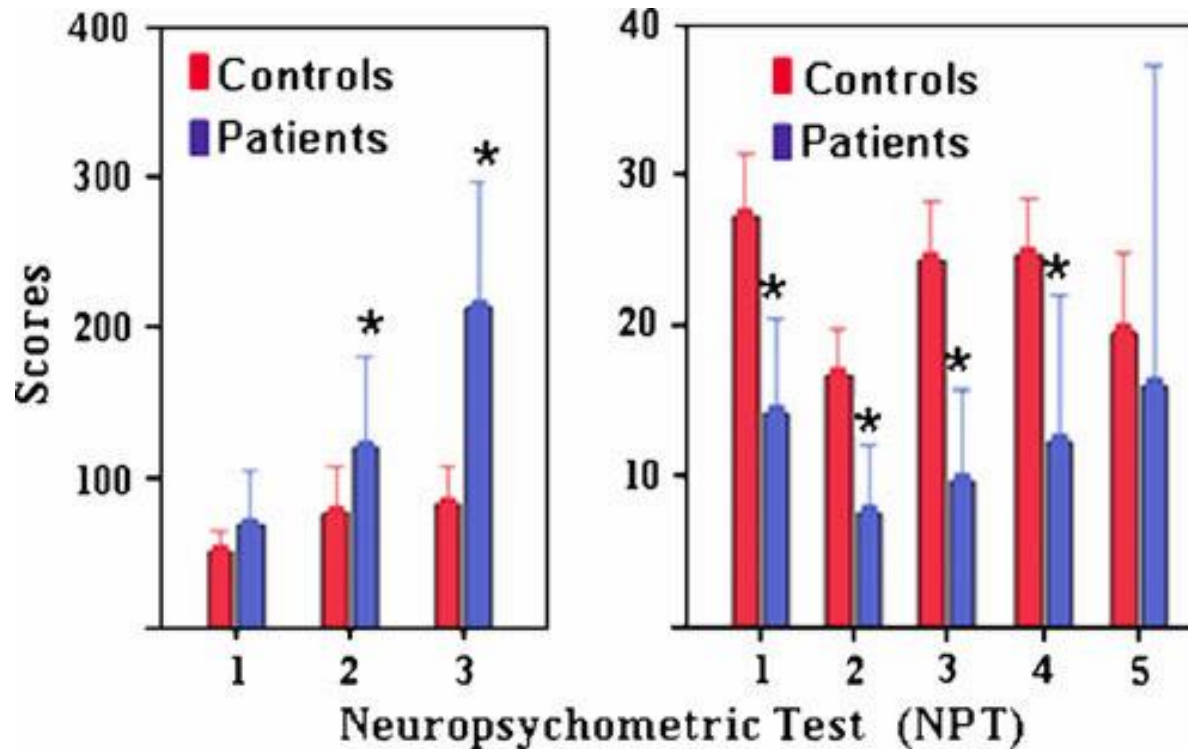
## Structural Abnormalities

- 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.





# 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

- 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

