Neuropsychological Assessment in Patients with Sickle Cell Disease

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Why Assess?

- Individuals with SCD of all ages are at higher risk of cerebrovascular complications, such as **acute ischaemic and haemorrhagic stroke** and **silent cerebral infarcts (SCI)**

- This can result in cognitive deficits that impact upon their communication with providers, medical adherence, academic and occupational achievement and overall quality of life.
Strokes in SCD

<table>
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<tr>
<th>Chances of having first stroke by</th>
<th>SS</th>
<th>SC</th>
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<tbody>
<tr>
<td>20 years of age</td>
<td>11%</td>
<td>2%</td>
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<tr>
<td>30 years</td>
<td>15%</td>
<td>4%</td>
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<tr>
<td>45 years</td>
<td>24%</td>
<td>10%</td>
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(CSSCD, Ohene-Frempong et al, 1998)

- 250 times more common in SCD than in other children
- If untreated, risk of recurrence (ischaemic stroke) = 50-92%
- Will often damage both grey and white matter

→ Leading cause of morbidity and mortality in SCD.

Commonly encountered patterns of cognitive impairment after stroke

- **Aphasias** - impairments of language
- **Apraxias** - impairments that affect limb movement and speech
- **Visuoperceptual and visuospatial disorders** - disorders of visual recognition (agnosias), visuospatial abilities and visual neglect
- **Memory impairments**
  - for events prior to the stroke (retrograde memory)
  - ability to lay down new memories (anterograde memory)
  - the inability to retain and manipulate information for a short time (working memory)
Commonly encountered patterns of cognitive impairment after stroke

- **Executive dysfunction** – impairments in conceptual reasoning, cognitive flexibility, planning, problem solving, etc.
- **Attentional impairments** and **speed of information processing**
- **General intellectual functioning** (i.e. I.Q)
- **Personality / behaviour changes**

Silent Cerebral Infarcts (SCI)

- **Silent cerebral infarcts** (“silent stroke”)
- **Most common** form of neurological injury in children with SCD
  - Prevalence increases during childhood:
    - 10% in infants
    - 28% by age 5
    - 37% by age 15
  - Prevalence continues to increase throughout adulthood
  - Typically occur within **small vessels**, generally confined to deep **white matter**, and involve non-motor areas of the brain (esp. frontal cortex)
  - Increased risk for further overt and silent strokes.
Impact of SCI in SCD – Cognitive difficulties

- Global cognitive dysfunction, particularly non-verbal IQ
  - Processing speed
  - Working memory
  - Executive function (planning, problem solving, organisation, inhibition, response monitoring, mental flexibility)
  - Attention, divided attention / switching
    (Berkelhammer et al, 2007; Mackin et al, 2014; Rawle et al, 2010; Vichinsky et al, 2010)

- In children, difficulties become more apparent in later stages of primary education, when intellectual demands increase
  - Poor school/work performance
  - Deficits in measures of executive functioning and attention/concentration
    - Difficulties with paying attention, short-term memory, organising and planning school work, initiating tasks and staying focused on them, regulating emotions, self-monitoring.

Impact of SCI in SCD – Cognitive difficulties

- Cognitive impairments tend to be more severe when patients have abnormal MRIs, but significant cognitive impairment in some patients with normal MRIs
  - MRIs not sophisticated enough to detect some brain changes; poor perfusion; effects of pain
  - Level of anaemia is more predictive (Vichinsky et al., 2010).
Standards of Care

- Sickle Cell Disease in Childhood: Standards and Guidelines (2006); Standards for Management of Sickle Cell Disease in Childhood (2008)
  - Regular neuropsychological screenings and monitoring of school attainment should be carried out on a regular basis
  - Patients should have access to a neuropsychologist within the MDT.

- Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK (draft 2017); Peer Review Standards for Sickle Cell Disease (updated draft 2017)
  - Patients should have access to neuropsychology via a defined pathway

What is a neuropsychological/cognitive assessment

- Interview
  - Medical, Educational, Employment, Family, Developmental, Language, Migration history – reasons and stressors
  - Coping, Views of problems (memory diary)
  - Mood, Pain

- Information from other sources
  - Health/Social
  - Educational records/Feedback from school
  - HCPs and family members
  - Research literature
What is a neuropsychological/cognitive assessment

- **Assessment of cognitive domains**
  - Memory, Attention, Processing Speed, Language, Executive Function, Visual-Spatial/Perception, Intellectual Functioning,
  - Word Reading, Reading Comprehension, Mathematics, Listening Comprehension, Spelling

- **Interpretation and recommendation**

- **Feedback and liaison**
  - Patient/Family/Carers
  - HCPs
  - Employers/School/College/SENCo
    - Can provide support for Education Health Care Plan (EHCP)
  - Onward referrals

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Current Service Model in Paediatric Sickle Cell & Thalassaemia Service (KCL)

**Referral to Clinical Psychology**

- **Psychology Assessment**
  - Semi-structured interview (90 mins)
  - Liaison with educational services
  - Psychometric assessment

- **Neuropsychological Assessment**
  - Cognitive Ability (2-3 hours)
  - Scholastic Achievement (1-2 hours)

- **Further assessment if required**

**Follow-up**

- F/U appointment offered to all families to discuss assessment findings
- Recommendations of appropriate educational, psychological or medical interventions
- Liaison with school/educational services (EHCP)
- Sign-posting/referral to other services where appropriate (e.g. SALT, OT)

**Discharge**
Tests used with Adults

- IQ:
  - WAIS-IV UK; WAIS-III UK; shortened versions
- Premorbid IQ
  - TOPF; WTAR
- Memory
  - WMS-IV UK; WMS-III UK; RBMT
- Executive Functioning:
  - Hayling and Brixton; Verbal & Category Fluency; BADs – key search, zoo map; Trail Making Test (TMT A&B); DKEFS Trails
- Visuospatial:
  - VOSP
- Attention:
  - WAIS subtests; Test of Everyday Attention
- Tests of Effort:
  - WAIS subtests

Tests used with Children & Adolescents

- Cognitive ability/IQ:
  - WPPSI (age range: 2:6 – 7:7)
  - WISC-V; WISC-IV (age range: 6:0 – 16:11)
  - WAIS-IV UK (age range: 17:0+)
- Scholastic Achievement
  - WIAT-II (age range: 4:0 – 16:11)
  - WIAT-III (age range: 4:0 – 25:11)
- Further assessment
  - NEPSY-II (age range: 3:0 – 16:11)
    - Attention and executive functioning; Language; Memory and Learning; Sensorimotor; Social Perception; Visuospatial Processing
  - Children’s Memory Scale (age range: 5:0 – 16:11)
  - D-KEFS (age range: 8:0 – 89:00)

Psychometric assessment:
- Connors 3rd Edition (Self-report/Parent/Teacher versions)
- Behaviour Rating of Executive Function (BRIEF) (Parent/Teacher versions)
- Strengths and difficulties questionnaire (SDQ) (Self-report/Parent/Teacher versions)
- Revised Children’s Anxiety and Depression Scale (RCADS) (Self-report/Parent)
Complexities – SCD and stroke

- Double time for interview:
  - Language, culture, education
  - How SCD affects person – pain, fatigue, expectations

- Strokes:
  - Hemiparesis/plegia, sensory, arousal, dysphasia, dysarthria, apraxia, ataxia, fatigue, sleep, epilepsy, pain, cognitive impairments

- SCD:
  - Cultural, educational background, language
  - Multiple strokes/silent strokes over time
  - Pain, medication, depression, anxiety
  - Lack of info from others as often isolated
  - Premorbid IQ? (lack of info)
  - Impact of SCD on school ach; expectations of self

Costs and time

- Neuropsychological testing is a scarce resource
  - Not widely available and time consuming
  - Therefore has not regularly been integrated into routine clinical care for patients with sickle cell disease

- The Vichinsky et al (2010) study involved a 6-hour neuropsychological battery, administered by a trained neuropsychologist

- Future? Computerised testing
  - NIH Toolbox - Cognition Battery (NIHTB-CB) (www.healthmeasures.net)
    - Need to ensure this contributes to a meaningful assessment when using it in clinical setting
  - Q-Interactive testing (http://www.helloq.co.uk/home.html) using iPads
    - Create unique, client-centric batteries at both the instrument and subtest levels
    - Improves administration accuracy and speed, provides real time scoring, and allows for flexibility in just a few simple taps.
# To screen or not to screen...

- **If stroke history and reporting concerns:**
  - Indicates comprehensive assessment (so screen not required)
  - SCD patients tend to be younger – stroke screening measures could still lack sensitivity (risk false negatives)

- **If silent strokes/no stroke history:**
  - Lack of sensitivity (risk false negatives)
  - Not in context of cognitive assessment → meaningless
    - Don’t have enough information to formulate why patient is presenting as they are → cannot make meaningful recommendations
  - Ethical dilemma
    - What happens if they have a poor score, but no service for a comprehensive assessment?
  - Self fulfilling prophecy
    - Low score → anxiety → perform worse

# Factors affecting screening scores

- e.g. why may the patient present with a **low processing speed** score?
  - Fatigue
  - Low mood
  - Anxiety
  - Trauma
  - Pain
  - Analgesia
  - Other medications
  - Effects on brain of stroke/silent stroke
  - Part of global picture of lower scores e.g. Learning disability
  - Malingering
Conclusions

- Clinicians should be aware of the risk of cognitive impairment in patients with SCD, even among those with normal MRI scans – this may impact on patient’s understanding, decision-making, and adherence to treatments.

- Neuropsychological assessments for patients with SCD are useful to highlight cognitive impairments that may otherwise be unnoticed by clinicians, and be a useful way of identifying those who require support (e.g. at school, university, work).

- Simple screening tools are not appropriate for clinical use in this population.

Questions?