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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

Chances of having first stroke by	SS	SC
20 years of age	11%	2%
30 years	15%	4%
45 years	24%	10%

(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

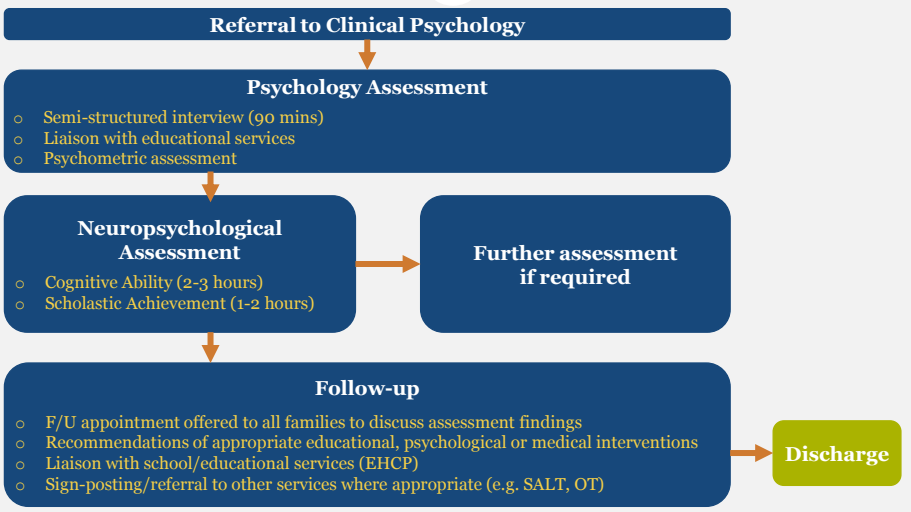
What is a neuropsychological/ cognitive assessment

- 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

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



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?