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The Psychological Needs of Patients with Sickle Cell Disease & Thalassaemia

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Introduction to Sickle Cell and Thalassaemia Study Day
King's College Hospital
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Plan for today's session

- Why is Psychology important?
- Psychological Difficulties in Children, Young People, and Adults with Sickle Cell & Thalassaemia
- Role of Psychology in Pain
- Neuropsychological Complications in Sickle Cell Disease
- Questions…

Exercise

- Turn to the person next to you, and think about what thoughts or feelings you might have as:
  - an 8 year old with sickle cell disease
  - a 16 year old with sickle cell disease
  - a 28 year old with sickle cell disease
- Share with the group…

Why is Psychology important?

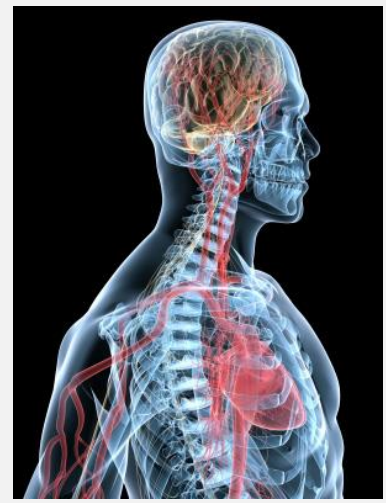
Sickle cell disease and thalassaemia is a chronic illness with psychological, social, and physical complications….

Why Psychology?

- Psychological support is particularly important for people with haemoglobin disorders because of:
  - lifelong nature of the disease
  - ongoing, frequent interactions with hospital services
  - impact on all stages of development, including cognitive development
  - psychological consequences of a lifetime living with pain.
- Needs are addition to underlying socio-economic challenges already faced by many people with haemoglobin disorders, as well as stigmatisation and discrimination.

Why is there a specialist service for people with Sickle Cell & Thalassaemia?

- Inclusion of psychology - consistent with a holistic approach to health care
  - Emphasises the interaction between psychological factors and physical health.



Background: Children & Young People

- Sickle Cell Disease in Childhood: Standards and Guidelines (2006)
  - CAMHS should provide Clinical Psychology assessment and management, and neuropsychological services for children and young people
  - Sickle Cell Centres must provide a Clinical Psychology service
  - All children with sickle cell disease and their families should have access to Clinical Psychology.
- Standards for the Clinical Care of Children and Adults with Thalassaemia in the UK (2016)
  - Psychosocial support, alongside specialist psychological care, should be provided as a standard part of thalassaemia clinical care
  - Core staffing of Specialist Haemoglobinopathy Centres should include a Clinical Psychologist with a special interest and experience in thalassaemia
  - Most recent round of peer review assessments of services found that psychology services for thalassaemia are among the most under resourced, and in many areas practically non-existent.

Background: Adults

- West Midlands Quality Review Service (2014)
  - National drive to have a dedicated Psychologist with an interest in haemoglobinopathies within MDT of services for adults with sickle cell disease
  - Drive for patients to have appropriate psychological assessments and interventions.
- Peer Review Programme (2014-16); Overview Report (2016)
  - Highlighted concerns about poor access to psychological services in 65% of those services reviewed
  - Many services lacked dedicated support from psychologists with specialist expertise in haemoglobin disorders.
  - Where psychological support was available, the amount of time allocated was usually insufficient for the number of patients and the extent of their needs.

Psychological Difficulties in Children, Young People, and Adults with Sickle Cell & Thalassaemia

Psychological Difficulties in Children, Young People, and Families – Risk Factors

- Children and adolescents with SCD are at risk for:
  - Internalising symptoms (e.g. depression, anxiety, low self-esteem)
  - Social and peer difficulties
  - Poorer quality of life
- Risk factors to psychosocial adjustment:
  - Impairments in mental abilities
  - Limited interpersonal, social, and academic opportunities due to illness-related factors (e.g. hospitalisations, crises) during critical periods of cognitive, emotional, and social development.

Psychological Difficulties in Children, Young People, and Families – Demographic Differences

- Demographic variables (such as age and gender) can influence psychosocial adjustment:
  - Children with SCD appear more concerned about family and social relationships (Schaeffer et al., 1999)
  - Adolescents seem more worried about delayed puberty and physical development, the liability of SCD on family and friends, and death and dying (Schaeffer et al., 1999)
  - Females tend to use more active coping strategies, and tend to report a better QOL than males (Casey et al., 2000; Lutz et al., 2004)
  - Males use denial as a coping strategies more than females, and utilise health care services more often than females (Royal et al., 2000; Lutz et al., 2004)
  - Girls with SCD are rated as less sociable and less well accepted by their peers, and boys are rated as less aggressive (Noll et al., 1996).

Psychological Difficulties in Children, Young People, and Families – Family Functioning

- Family distress appears to be related to family conflict and typical child rearing tasks, rather than diagnosis of SCD or its severity (Thompson et al., 2003).
- Parents and caregivers need to be:
  - Knowledgeable about SCD and its treatment
  - Skilled in recognising symptoms and implementing treatment plans at home
  - Persistent in coordinating other family responsibilities and activities.
- Coping strategies used by caregivers as they incorporate SCD into lives:
  - Active strategies – information seeking, using social support
  - Passive strategies – prayer
- Key psychological interventions for families:
  - Addressing family functioning through systemic interventions
  - Parenting skills (reduce stress/distress, improve knowledge and skills)

Psychological Difficulties in Children, Young People, and Families – Adherence

- Adherence to pharmacological treatments (e.g. analgesics, oral antibiotics) and non-pharmacological treatments (e.g. hydration) for SCD in children and adolescents varies widely.
- Family flexibility in problem solving and use of fewer passive coping strategies appear to promote adherence (Barakat et al., 2002).
- Adherence is higher for more concrete recommendations (e.g. follow-up clinic appointments), and worse for vague recommendations (e.g. hydration) (Barakat et al., 2005).
- Common issues associated with adherence:
  - Forgetting
  - Time constraints
  - Falling asleep
  - Running out of medication
  - Issues with obtaining refills

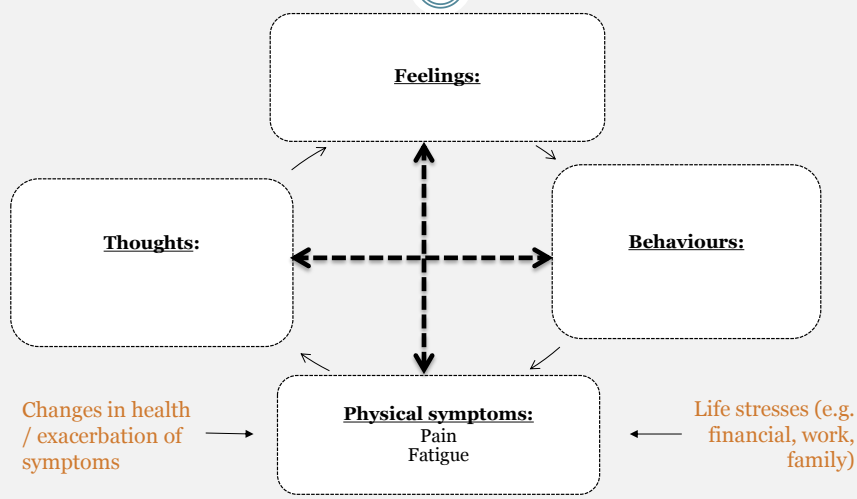
Psychological Difficulties in Adults – Quality of Life

- McClish et al. (2005)
  - Large-scale American prospective epidemiological survey
  - On average, people with SCD report severely compromised health-related quality of life in comparison to the general population (with a pattern of scores worse than people with asthma or cystic fibrosis, and most similar to patients undergoing haemodialysis).
- People with SCD report rates of mental health problems more frequently than the general population…

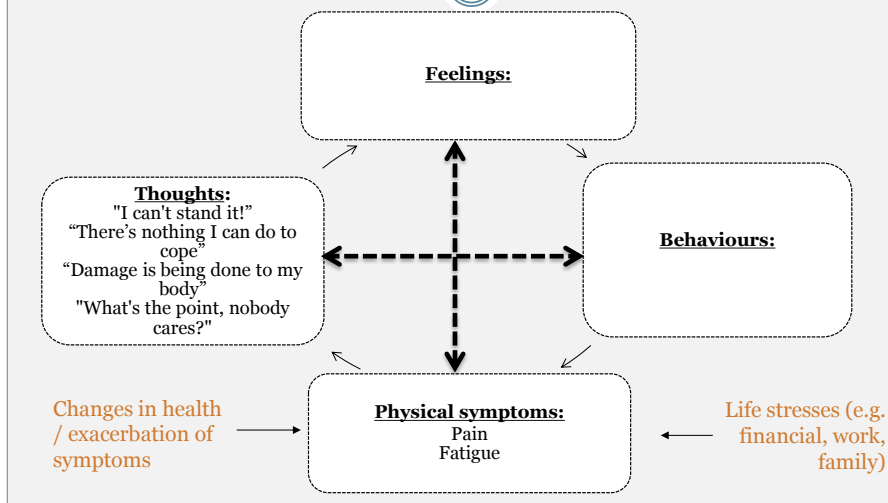
Psychological Difficulties in Adults – Depression and Anxiety

- Levenson et al. (2008)
  - American study
  - Nearly 28% of participants met criteria for depression, and another 6-7% met criteria for an anxiety disorder
  - Presence of depression or anxiety predicted increased frequency, intensity, distress and disruption due to pain
  - Depression and anxiety = larger predictors of pain factors than disease phenotype.
- Can conclude that a sizeable proportion of people with SCD will also present with psychological difficulties
  - Has a direct bearing on their experience of pain symptoms and perceived quality of life (Sogutlu et al., 2011).

Interactive cycles



Interactive cycles



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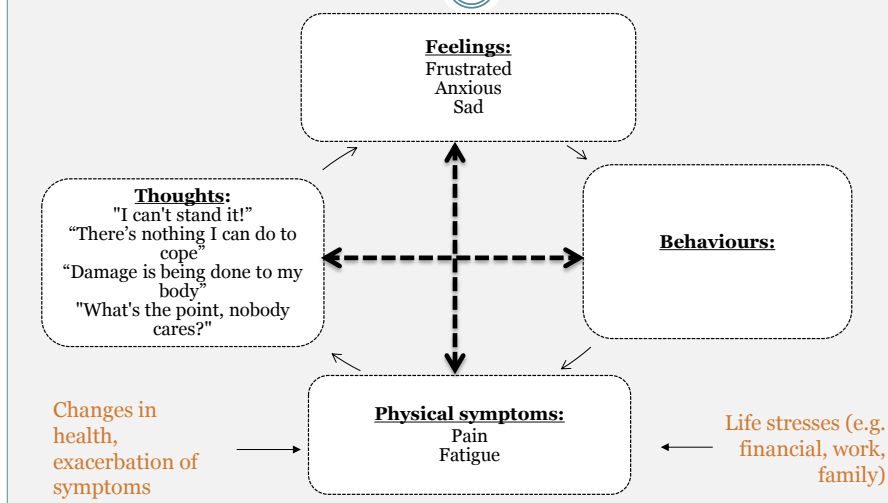

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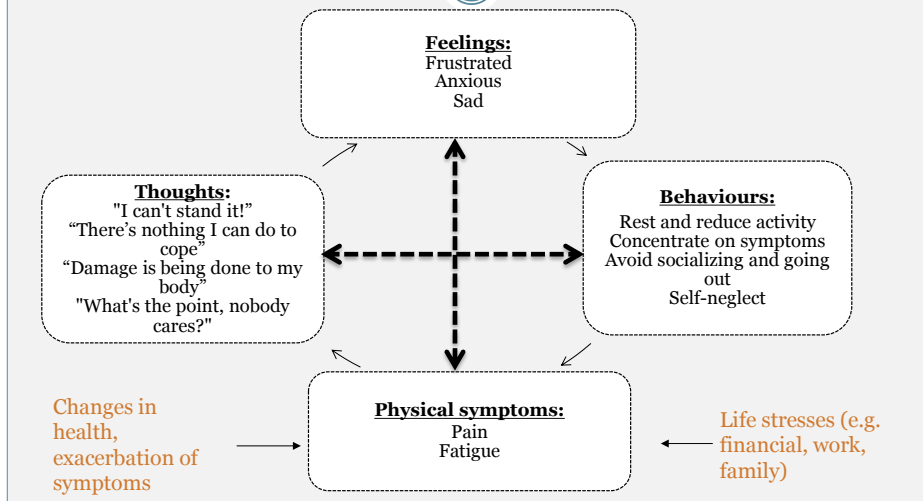

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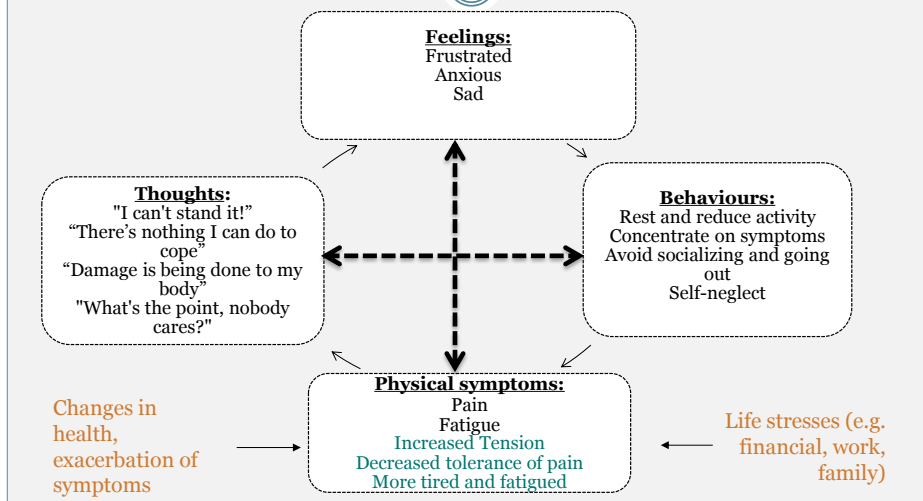


Role of Psychology in Pain

Pain

- The acute painful episode (or crisis) = characteristic presentation of SCD
- It accounts for more than 90% of hospital episodes

Pain in Sickle Cell Epidemiology Study (PiSCES) (2015)

- American single site study
  - Pain in adults with SCD is far more prevalent and severe than previous studies have portrayed, and it is mostly managed at home.
    - Therefore, pain has been vastly underestimated when measured by using only healthcare utilization!
  - >50% of adult with SCD experienced pain, crises, or healthcare utilization on 55% of the surveyed days, with 29% of patients experiencing pain on more than 95% of the days
  - Almost 33% experienced pain nearly every day, with the mean intensity in the middle range
  - In contrast, only approximately 15% rarely experienced pain.

Pain in Sickle Cell Epidemiology Study (PiSCES) (2015)

- Crises and healthcare utilization were far less common than reported pain days
  - Pain days that were not associated with a crisis occurred 10 times more often as pain days associated with healthcare utilization.
- Thus, contrary to commonly held belief, pain in adults with SCD is the rule rather than the exception
  - Since SCD adults infrequently utilize health care even in response to severe pain, there is a vast, mostly submerged, iceberg of sickle cell pain that is managed outside of medical facilities and not seen by most professionals.

- Smaller longitudinal studies measuring daily pain in children have also found that pain was most often managed at home rather than within healthcare facilities (Smith et al 2005).

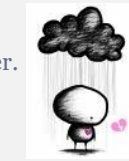
Research on pain in SCD

- Studies have shown that both depressed and anxious patients with SCD experience:
  - More frequent pain
  - Higher intensity of pain
  - Greater distress from pain
  - Larger interference from pain
- Painful episodes seem more common following negative life events (Gil et al., 1989)
- Negative thinking and passive adherence (e.g. rest, heat, massage, taking fluids) contribute to increases in:
  - Pain perception
  - Distress
  - Opioid use
  - Healthcare utilization

Coping strategies in children and young people

- Active and passive coping strategies have been identified as resiliency factors in youths with SCD
  - Active coping strategies: distraction or imagery to divert attention from pain (Casey et al., 2000)
  - Passive coping strategies: rest, sleep, warm baths (Mitchell et al., 2007)
- Active coping strategies are associated with:
  - Increases in active health management
  - Fewer health care contacts and school absences
  - More involvement in daily activities on pain days
  - Decreases in negative thinking

"Negative thinking"



- Unhelpful thinking habits
  - Composed of catastrophising and self-statements of fear and anger.
- Catastrophising:
  - Imagining and believing the worst possible thing will happen
  - Exaggerated negative orientation toward pain stimuli and pain experience
- Individuals who catastrophise may:
  - Develop beliefs with a high degree of aversion to pain-eliciting situations
  - Pay more attention to their pain sensations
  - Consume more opioids
- Catastrophising can be understood within a set of thinking errors that includes rumination *(repetitively going over a thought or a problem without completion)*, magnification *(exaggerating risk/negative)*, and helplessness to deal with pain.

Psychological Interventions

- A review of the literature assessing the efficacy of psychological interventions in SCD found that cognitive-behavioural interventions were *probably efficacious* for sickle-cell pain (Chen, et al., 2004).
- CBT techniques → aim to reduce pain by utilising cognitive strategies (e.g. calming self-statements, distraction, visual imagery) and behavioural strategies (e.g. progressive muscle relaxation, pacing).
- While there are numerous studies and reviews indicating the effectiveness of psychological therapies in addressing problems relevant to people with SCD, few of these actually focus on people with SCD.

Evidence base for CBT

- Anie and Green (2015)
  - Cochrane review
  - Efficacy of CBT with SCD was best supported for addressing pain
  - While patient education and development of coping strategies were seemingly important, there was a shortage of evidence to support this.
- Hoffman (2012)
  - Whilst the evidence base for CBT is generally considered very strong, there is an on-going need for meta-analytic review of CBT for patient groups involving ethnic minorities and low-income groups
    - **Both factors relevant to those with SCD**
- Growing evidence base for "third-wave CBT" approaches
  - Acceptance and Commitment Therapy

Role of Social Support

- Having friends and being integrated into social networks → helps to preserve health, prolong life, and to speed recovery from illness.
- Merely expressing feelings about upsetting or traumatic challenges reduces physiological arousal and helps to reduce damaging physiological effects.
- The presence of other people does not necessarily equate to feeling emotionally supported
  - Support occurs where the patient feels that someone is available who understands, listens and respects their concerns.
- Belgrave & Lewis (1994)
  - In adults with SCD, social support was associated with adherence with appointments and health promoting behaviours.

Neuropsychological complications of Sickle Cell Disease

Cerebrovascular disease

- Cerebrovascular disease (particularly stroke or ischaemic brain injury) is reported to be the most disabling complication in SCD.
- Younger children are more likely to develop silent cerebral infarcts, and older patients are more inclined to haemorrhage.
- **Neuropsychological screening and assessments** for patients with SCD
  - Useful way of identifying those who require support (e.g. at school, university, work).
- 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.

Strokes and Silent Cerebral Infarcts (SCI) in Children and Young People

- Overt strokes = 9-11%
  - Onset is generally abrupt
  - Typically occur within large vessels, commonly involve both cortex and deep white matter, and are ass. w/greater deficits.
- Silent cerebral infarcts ("silent stroke") = 10-37%
  - 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
  - Typically occur within small vessels, generally confined to deep white matter, and involve non-motor areas of the brain (esp. frontal cortex)
  - MRI studies of stroke-free children with HbSS have shown ischemic lesions which are not apparent on neurological examination
  - Increased risk for further overt and silent strokes.

Cognitive and Academic Functioning: Children and Young People

- Individuals with SCD of all ages are at risk for developmental delays, cognitive difficulties, and/or academic deficits.
- Overt strokes in patients with SCD:
  - Deficits in measures of general intelligence, language abilities, visual motor skills, sequential memory, and attention
- Silent strokes in patients with SCD:
  - Poor school/work performance
  - Deficits in measures of executive functioning and attention/concentration
    - Difficulties with paying attention, short-term memory, organising and planning, initiating tasks and staying focused on them, regulating emotions, self-monitoring
  - Becomes more apparent in later stages of primary education, when intellectual demands increase.

Cognitive and Academic Functioning: Adults (1)

- Vichinsky et al. (2010)
  - Multi-centre trial in US
  - Adults with SCA vs. healthy controls from same communities
  - Assessed neuropsychological condition and function
  - Administered wide-ranging battery of neurocognitive measures, as well as MRI evaluation
  - Major findings :
    1. Adults with SCA showed poorer performance on neurocognitive tests
    2. Anaemia was associated with the age-related decline in cognitive performance
    3. MRI findings did not explain the performance differences in the subset of patients with neuroimaging studies.

Cognitive and Academic Functioning: Adults (2)

- Conclusions
  - MRI is not an adequate screening tool to identify patients with cognitive impairment.
  - Clinicians should be aware of the risk of cognitive impairment in adults with SCD, even among those with normal MRI scans – this may impact on patient's understanding, decision-making, and adherence.
  - Cognitive assessment may be useful to highlight cognitive impairments that may otherwise be unnoticed by clinicians.

Questions?