

The Psychology of Sickle Cell Disease

Expanding the Toolbox for Building Your Life with SCD



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Why Is There a Psychologist Here?!

- Health psychologists study how patients handle illness, why some people have trouble following medical advice and the most effective ways to control pain or change health habits. They also develop health care strategies that foster emotional and physical well-being.
- Physicians diagnose and treat illnesses through a variety of means, but what factors contribute to our health? That's where the skills of health psychologists can help.
- From working in clinical settings to conducting research and influencing health care policy, health psychologists measure the impact of behavior on health and create ways to help people make the behavior choices that induce good health and prevent illness.
- What psychological factors determine whether someone makes a good or poor food choice when given both options? What motivates them to exercise? Why are those at risk for cancer not getting regular screening? These are all questions that health psychologists are addressing.



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Sickle Cell Disease and Pain

- Sickle cell disease refers not to a single disease but rather to a collection of inherited blood disorders that may cause blood cells to morph from adaptive into crescent or so-called sickle shapes
 - Affects approximately 100,000 persons in the United States
 - SCD patients have historically been underserved
 - Patients with SCD can face both structural and interpersonal discrimination
- Health and survival of those with sickle cell disease have improved considerably with the advent of newborn screening, penicillin prophylaxis, pneumococcal immunisation, other treatment advances, and education about disease complications.



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Sickle Cell Disease and Pain

- The most prevalent complication of SCD is the vaso-occlusive crisis (VOC) "sickle cell crisis" or "sickle cell exacerbation."
 - These acute, excruciatingly painful events are the leading cause of hospital and emergency department
 - While SCD is considered a rare disease in the United States, the burden of ED care and subsequent hospitalization is high
 - Both patients and health care providers often report feeling dissatisfied with options for SCD pain management



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Sickle Cell Disease and Pain

- The most prevalent complication of SCD is the vaso-occlusive crisis (VOC) “sickle cell crisis” or “sickle cell exacerbation.”
 - Acute pain, the hallmark clinical feature of sickle cell disease
 - Stems both from vaso-occlusion and impaired oxygen supply, infarction/reperfusion injury, and other complications
 - Pain in sickle cell disease involves peripheral nociceptor activation and hyperalgesia, possibly with mast cell activation
 - Both children and adults can have pain affecting bones in the extremities, chest, and back.
 - Non-steroidal anti-inflammatory drugs and opioids provide effective relief, and guidelines describe appropriate management of sickle-related pain. Frequently, though, these are not enough.
 - Why?

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Pain and Psychological Distress

- Pain: “an unpleasant sensory and emotional experience associated with actual or potential tissue damage.”
 - Nociceptive pain: when a part of the body is damaged and sends a signal to the brain saying so (e.g., from a fractured bone or from an inflamed organ).
 - Neuropathic pain: damage to neurons, and may persist despite the absence of ongoing disease (e.g., from phantom limb pain).

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Pain and Psychological Distress

- Fear and Anxiety: Fear occurs as a result of perceived imminent threat, anxiety is a state of anticipation about perceived future threats
 - Fear can be adaptive, e.g. when an unfriendly looking dog approaches you facilitate appropriate defensive reactions that can reduce danger or injury
 - Anxiety may occur later, at home, when you begin thinking about seeing that dog again when you next walk that path

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Pain and Psychological Distress

Pain can trigger both fear and anxiety, and these can make very real pain worse.

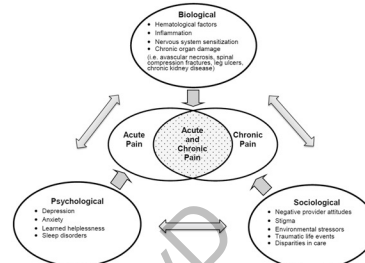
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Pain and Psychological Distress

- Hypothalamic Pituitary Adrenal (HPA) Axis in Fear and Pain
 - In processing fear and pain experiences, norepinephrine and indirect limbic inputs from the hippocampus, medial prefrontal cortex, and amygdala act on neurons to release cortisol and, eventually, adrenaline as well as epinephrine and norepinephrine
 - These hormones induce a variety of effects throughout the body to support the stress response, including inhibiting insulin and enhancing glucose availability, regulating immune system functions, and impacting electrolyte balance.
 - Systolic and diastolic blood pressure, heart rate, and muscle activity

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Pain and Psychological Distress



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Some Factors Influencing SCD Pain and Anxiety

- Things One Can Control a Little
 - Infections
 - Other medical conditions
 - Effects of prescription and non-prescription pain treatment
 - Weather
 - Environmental stressors
- Things One Can Control More
 - Self-efficacy / self-management
 - Managing medications
 - Appointment keeping
 - Tracking health issues
 - Talking with providers
 - Managing daily activities
 - Perceived support
 - Religious / spiritual beliefs
 - Attitude (without invalidating oneself)

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Some Factors Influencing SCD Pain and Anxiety



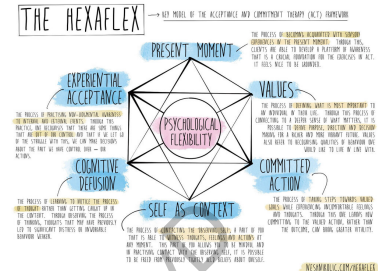
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Psychotherapy in Sickle Cell Disease: ACT

- Psychological suffering and a failure to prosper psychologically is usually caused by the interface between the evolutionarily more recent processes of human language and cognition, and more ancient sources of control of human behavior, particular those based on learning by direct experience.
- Psychological inflexibility is argued to emerge from six basic processes. Stated in their most general fashion these are emotional inflexibility, cognitive inflexibility, attentional inflexibility, failures in perspective taking, lack of chosen values, and an inability to broaden and build habits of values-based action.
- Buttressed by an extensive basic research program on a linked theory of language and cognition, Relational Frame Theory (RFT), ACT takes the view that trying to change difficult thoughts and feelings in a subtractive or eliminative way as a means of coping can be counter productive, but new, powerful alternatives are available to deal with psychological events, including acceptance, cognitive defusion, mindful attention to the now, contacting a deeper "noticing" sense of self or "self-as-context", chosen values, and committed action.
- These six flexibility processes are argued to be inter-related aspects of psychological flexibility. Each of these in turn can be extended socially. For example, acceptance of emotions can extend to compassion for others; chosen values can extend to social values; a "noticing" sense of self to healthy social attachment; and so on.

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Psychotherapy in Sickle Cell Disease: ACT



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Psychotherapy in Sickle Cell Disease: ACT

- ACT emphasizes the promotion of daily functioning and QOL while teaching a willingness to experience difficult and possibly unavoidable private events (e.g., pain, discomfort, fatigue, anxiety) without defense when doing so serves valued ends (Hayes, Luoma, Bond, Masuda, & Lillis, 2006).
- Based on its behavioral roots, ACT posits that functional disability and diminished QOL is typically characterized by the domination of inflexible attempts to modify the form, frequency, or situational sensitivities of unwanted private events (e.g., anxiety, pain).
 - For example, a patient with SCD might be taught to engage in (e.g., hydrate) or avoid (playing outside on a cold day) in order to minimize SCD symptoms. However, if these behaviors become dominant, excessive, or rigid, they might interfere with daily functioning (making a friend, socializing with peers, studying, having part-time work).
- Behaviorally, many of these negatively reinforced behaviors can be rule-governed, which are under the stimulus control of aversive private events (e.g., fatigue) and verbal rules associated with these events (e.g., "I can't do anything when I am unmotivated").
- It should be noted that these behaviors can be reinforced by others (e.g., parents, physicians). In ACT, the maladaptive behavioral pattern of attempting to alter the form, frequency, or situational sensitivity of private events (e.g., thoughts, emotions) are generally called experiential avoidance (EA), and the stimulus control of verbal antecedents that excessively or improperly regulate EA are called cognitive fusion (Hayes et al., 2006).

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Psychotherapy in Sickle Cell Disease: ACT

- What does it look like?
 - As few as eight 60-min consecutive weekly individual or group/family sessions but can go on longer as situations become more complex
 - Sessions are often structured as follows:
 - 5–10 min review of one's daily activities as well as medical and psychosocial concerns
 - ACT-specific content (e.g., acceptance, mindfulness, and values clarification) is then typically introduced in a didactic fashion and then used to address the real weeklies issues raised earlier, with this aspect of therapy spanning 30–40 min.
 - The final 10–15 min of the sessions focused on helping the individual set specific values-based goals.

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Psychotherapy in Sickle Cell Disease: ACT

- Six processes of change are theorized to be at the core of ACT (Hayes et al., 2006): acceptance of private events (experiencing those events willingly and without defense), cognitive defusion from the literal content of thoughts (not necessarily believing them or acting on them), present moment awareness of one's experience, a sense of self-as-context or perspective, clarification and induction of valued directions for life, and committed action (building patterns of overt behavior in valued directions).

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Psychotherapy in Sickle Cell Disease: ACT

- Goal 1: shift perspective from excessive and exclusive attempts to avoid and control unwanted psychological events (e.g., pain, discomfort, fatigue, anxiety) to values-focused living.
 - Discuss the inevitable, often unpredictable, and pervasive nature of SCD symptoms (e.g., pain, fatigue) despite careful symptoms management efforts.
- Discuss of the utility and costs (e.g., missed school; missed peer interactions) in trying to live a "pain-free" and "stress-free" life.
- Behaviorally, goals of this phase are to undermine the behavior regulatory function of rules associated with EA and cognitive fusion in the context of SCD symptoms and associated events so that alternative behaviors, such as value-consistent behaviors, were likely to occur.
 - Involves identifying constructive and values-consistent activities (e.g., being with family/friends, academic or professional pursuits, etc.) that are inhibited by inaction due to SCD symptoms and associated private events (e.g., fatigue, lack of motivation).
- Finally, the pursuit of value consistent and vital living despite the presence of SCD symptoms in some degree was established as the goals of therapy rather than focusing on symptom control.
 - Consider a continuum of choices from focusing energy in living a "stress-free" and "safe" life (e.g., staying home from important events when fatigued, avoiding being with friends when experiencing pain) to living a "vital" and "values-consistent" but "higher risk" life (e.g., accepting that activities despite fatigue are important in achieving the goals and consistent with ones values).
- We acknowledged that some symptom reduction behavior (e.g., hydration) can help to move in the direction of living a vital life.

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Psychotherapy in Sickle Cell Disease: ACT

- Values are emphasized in Sessions 3 and 4 in order to firmly establish ACT-congruent treatment goals.
 - Involves completing a values-clarification exercises (e.g., projecting what one would like their life to be like in 5, 10, 15, and 20 years), which are similar to activities described in ACT manuals (e.g., Hayes & Smith, 2005).
- Identify emotional and physiological obstacles that might interfere with values-consistent activities (e.g., anxiety, pain, and fatigue).
- The values-clarification exercise helps individuals and therapists agree upon concrete target behaviors and the arrangement of contingencies to promote the behaviors in the context of day-to-day living.

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Psychotherapy in Sickle Cell Disease: ACT

- The stance of acceptance and mindfulness re used throughout all sessions, but emphasized in Sessions 2 and 5.
- Several exercises re implemented to facilitate one in engaging in values-consistent behavior despite SCD symptoms.
 - Example: one may feel annoying by others prompting them to take their medication, which may leave the person feeling discouraged and disempowered. While the reminder may come from a good place of wanting the best for the person with SCD, it nonetheless impacts the situation negatively.
 - In this case, the individual and/or person giving the reminders would be encouraged to practice increasing their awareness of these thoughts and emotions—to experience these private events—without acting on or trying to change them, and to simultaneously engage in values-based behavior (building a sense of autonomy for the person, building a sense of trust for the one providing reminders).
- Behaviorally, the goals of acceptance and mindfulness exercises are to alter the EA regulatory function of their private events (e.g., thoughts, perceived SCD symptoms) so that other behaviors, including values-consistent activities, flexibly occurs in the presence of these events.
- This same approach of encouraging acceptance of difficult thoughts, feelings, and physical sensations and mindfully engaging in behavior grounded in values was used in other areas.
 - Allowing one's self to experience anxiety and reservation about applying for a job that SCD may interfere with (which may have stymied one in the past) while simultaneously applying and possibly securing that job, all of which may be consistent with his goals of obtaining work to increase his social activity, and provide financial support for other socializing.

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Psychotherapy in Sickle Cell Disease: ACT

- Committing to values-directed living is infused throughout therapy, but highlighted in Sessions 5 through 8.
 - Predominately accomplished by repeatedly identifying one's values (e.g., education, rewarding profession, independence) and linking these values to distal and proximal concrete goals (e.g., studying to improve grades, obtaining a job, spending time at church, baking).
 - The psychological and physical challenges (e.g., uncertainty, fear, pain) experienced by the person when establishing behavioral goals are continuously discussed to normalize the experience, encourage acceptance, and place the primary focus on values-consistent living.
- Toward the end the individual is encouraged to revisit their thoughts and concerns about SCD and its symptoms ("I can't stand this," "I can't pursue what I want to be because of SCD") and to allow themselves to have these thoughts without trying to alter or act on them.
 - These private events are revisited and discussed repeatedly and especially toward the end of therapy because they are likely to remain and could interfere with daily functioning. The individual is encouraged to continue to notice these thoughts but to behave and live in accord with their identified values, such as academics, professions, hobbies, family, etc.

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Psychotherapy in Sickle Cell Disease: CBT

- CBT is a psychological therapy that addresses problems in terms of the relationship between thoughts, feelings, physiological sensations, and behaviors (Currid, Nikcevic, & Spada, 2011). Individuals using cognitive strategies recognize and correct inaccurate negative thoughts, whereas behavioral strategies offer support for positive change (Townsend, 2006).
- Behavioral methods arise from the idea that inappropriate behaviors are learned and therefore can be unlearned (Anie, 2005). In CBT, individuals are encouraged to use several techniques including cognitive therapy using attention diversion, distraction, and imagery, and behavioral therapy such as relaxation, biofeedback, and goal setting (Anie & Green, 2012).
- CBT's central features consist of cognitive techniques aimed at modifying thoughts, beliefs, feelings, and behavioral strategies to promote change in coping responses (Thomas et al., 2001). In studies on the use of CBT for pain management in SCD, CBT was found to reduce pain and help patients to live more effective and fulfilling lives (Cummins & Anie, 2003; Thomas et al., 2001; Thomas et al., 1998).

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Psychotherapy in Sickle Cell Disease: CaRISMA

- The CaRISMA (Cognitive Behavioral Therapy and Real-time Pain Management Intervention for Sickle Cell via Mobile Applications) study is a multisite comparative effectiveness trial funded by the Patient-Centered Outcomes Research Institute.
 - Six clinical academic sites, in partnership with four community-based organizations.
 - Will evaluate the effectiveness of two 12-week health coach-supported digital health programs with a total of 350 participants in two groups: CBT (n=175) and Education (n=175).
 - Participants will complete a series of questionnaires at baseline and at 3, 6, and 12 months. The primary outcome will be the change in pain interference between the study arms.
 - Examines changes in pain intensity, depressive symptoms, other patient-reported outcomes, and health care use as secondary outcomes.

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Psychotherapy in Sickle Cell Disease: CaRISMA

- <https://scdstudies.com/featured/carisma/>



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THANK YOU!

Questions, Comments, Discussion

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