

A Review of Empirically Supported Psychosocial Interventions for Pain and Adherence Outcomes in Sickle Cell Disease

Edith Chen,¹ PhD, Steve W. Cole,² PhD, and Pamela M. Kato,³ EdM, PhD

¹University of British Columbia, ²University of California, Los Angeles, and

³Stanford University, HopeLab

Objective To review empirical studies of psychological interventions for pain and adherence outcomes among patients with sickle cell disease. **Method** We conducted a literature review of studies using psychological interventions targeted at pain and/or adherence behaviors related to sickle cell disease. The American Psychological Association Division 12 Task Force criteria (Chambless criteria) were used to evaluate the empirical support for three categories of interventions (cognitive-behavioral techniques, interventions aimed at behavioral change, and social support interventions). **Results** A small number of intervention studies met criteria for demonstrating empirical efficacy. As a group, cognitive-behavioral techniques fall into the category of *probably efficacious* for sickle cell pain. Other intervention types were limited by inadequate research methodologies. **Conclusions** Future studies will need to more stringently test outcomes related to acute crises (e.g., pain episodes) as well as day-to-day management of sickle cell disease to clarify the most efficacious intervention approaches. Implications and suggestions for future research directions are discussed.

Key words sickle cell; psychosocial; intervention.

Sickle cell disease (SCD) is an inherited genetic disorder that affects predominantly African Americans, occurring in approximately 1 in every 500 African American births in the United States (NHLBI, 1996). There is no cure for SCD; rather, prevention and treatment of symptoms are emphasized. The most distressing clinical problem associated with SCD is the experience of unpredictable and severe pain episodes, which is the most common reason for hospitalizations in individuals with SCD (Woods et al., 1997). Pharmacological and nonpharmacological treatments involve attempts to manage pain episodes when they occur. In addition, certain daily health practices are encouraged by many physicians in an attempt to minimize future pain episodes and SCD-related complications. For example, infection, fever, temperature extremes, dehydration, and sleep difficulties are thought to precipitate pain episodes (NHLBI, 1995). Thus recommended health practices include:

taking daily oral penicillin, in younger children; avoiding temperature extremes (as well as knowing how to take temperature); drinking at least 150 cc/kg per day of liquids; adequate rest; avoidance of stresses; and regular visits to a physician (in accordance with vaccination schedules during the first 2 years of life, then semi-annually for children) (NHLBI, 1995). Some physicians also recommend the use of heating pads or massage during pain episodes.

Medical management of pain episodes at home includes the use of over-the-counter pain relieving (aspirin, acetaminophen, and other nonsteroidal anti-inflammatory agents) and sometimes more potent prescribed analgesics such as oral narcotics (e.g., codeine) (NHLBI, 1995). Most recommendations for SCD management also include psychosocial interventions in addition to medical treatment (Collins, Kaslow, Doepke, Eckman, & Johnson, 1998; Vichinsky & Lubin, 1987).

All correspondence should be addressed to Edith Chen, University of British Columbia, Department of Psychology, 2136 West Mall, Vancouver, B.C., Canada V6T 1Z4. E-mail: echen@psych.ubc.ca.

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Table I. American Psychological Association Division 12 Task Force Criteria for Empirically Supported Treatments (also known as Chambless criteria)

Treatment Label	Criteria
<i>Well-established</i>	<ol style="list-style-type: none"> I. At least two good between-group design experiments demonstrating efficacy in one or more of the following ways: <ol style="list-style-type: none"> A. Superiority to pill or psychological placebo or alternative treatment B. Equivalence to an already established treatment in experiments with adequate statistical power (about 30 per group) II. A large series of single-case design experiments ($n \geq 9$) demonstrating efficacy. These experiments must have <ol style="list-style-type: none"> A. Used good experimental design and B. Compared the intervention with another treatment as in I(A). III. Experiments must be conducted with treatment manuals. IV. Characteristics of the client samples must be clearly specified. V. Effects must have been demonstrated by at least two different investigators or investigatory teams.
<i>Probably efficacious</i>	<ol style="list-style-type: none"> I. Two experiments showing the treatment is more effective than a wait list control group, or II. One or more experiments meeting the <i>well-established</i> treatment criteria I, III, and IV, but not V.

Note: *Well-established* interventions require I or II, plus III, IV, and V (Chambless & Hollon, 1998).

Common psychosocial interventions include relaxation and distraction techniques, cognitive coping strategies, behavioral contracts, education, social support or self-help groups, and family interventions (Collins et al., 1998; Fox & Ingram, 1999; Shapiro, 1989; Tarnowski, Brown, Dingle, & Dreelin, 1998). The overall goals of these approaches are to encourage mastery over pain experiences and maintenance of daily activities (pain-related goals), as well as to encourage daily management practices and early symptom recognition (education- and adherence-related goals) (Kaslow et al., 2000; Thompson, Gustafson, & Ware, 1998; Walco, Sterling, Conte, & Engel, 1999).

This paper reviews the empirical evidence for the efficacy of psychosocial interventions for improving SCD-related outcomes. For the purposes of this review, we have grouped treatments into one of three broad categories: (1) cognitive-behavioral techniques (this includes relaxation, biofeedback, distraction, hypnosis,

cognitive coping strategies, and cognitive-behavioral therapy [CBT]), (2) interventions aimed at behavioral change (behavioral modification strategies and education), and (3) social support interventions (support groups and family interventions). The focus of this paper is on health-related outcomes, rather than on psychological adjustment to chronic illness; thus, this review considers the extent to which the two goals described above (pain reduction and education/adherence behaviors) are met. Studies that examine only psychosocial outcomes are not included, and psychosocial outcomes contained within the studies below are not emphasized here. To locate intervention studies, Medline and PsychInfo searches were conducted from 1975 to 2002 using the key words *sickle cell*, *psychological*, *psychosocial*, *intervention*, *therapy*, *treatment*, *pain management*, and *adherence*. Reference lists from articles obtained also were scanned for additional relevant articles.

Efficacy of the three broad categories of interventions described above is evaluated according to the American Psychological Association (APA)'s Division 12 Task Force criteria (also known as the Chambless criteria) for empirically supported interventions (Chambless & Hollon, 1998). The criteria for being labeled either a *well-established* or a *probably efficacious* psychosocial treatment are summarized in Table I. Given the paucity of research conducted in children with SCD, we include in this review some intervention studies of adults with SCD that may help inform future directions of research in children. Previous reviews published have addressed levels of empirical support according to the APA/Chambless criteria for psychosocial interventions in other childhood chronic illnesses such as asthma (McQuaid & Nassau, 1999), or for chronic pain (Holden, Deichmann, & Levy, 1999; Janicke & Finney, 1999; Walco et al., 1999). Previous reviews of SCD have provided clinical guidelines or broad summaries (Collins et al., 1998; Yaster, Kost-Byerly, & Maxwell, 2000) but have not evaluated SCD intervention studies according to the APA/Chambless criteria. In addition, the efficacy of psychosocial interventions in ethnic minority groups is an understudied area, and this review provides timely and important information about empirically supported interventions relevant to African Americans. See Table II for a summary of the intervention studies included in this review. Within each section below, we describe the intervention studies that have been conducted, evaluate the evidence according to the APA/Chambless criteria, and provide specific suggestions for future research.

Cognitive-Behavioral Techniques

Interventions that have been grouped in this category utilize techniques aimed at reducing pain through the use of cognitive strategies (such as calming self-statements, hypnosis, and imagery) and/or behavioral strategies (such as biofeedback and progressive muscle relaxation). These techniques are taught in either individual or group settings, typically to the patients themselves (rather than to families as a whole).

The most rigorous set of intervention studies in this category tested the efficacy of a cognitive coping skills training program. This intervention developed out of observational studies that found that negative thinking is associated with greater pain and health care utilization, whereas active coping attempts are associated with less activity reduction and health care utilization (Gil, Abrams, Phillips, & Williams, 1992; Gil, Williams, Thompson, & Kinney, 1991). The cognitive coping intervention tested in these studies included three individual sessions to teach calming self-statements, reinterpreting pain, and strategies for relaxation and distraction (breathing relaxation, imagery, mental counting, and focusing on physical surroundings). Adults with SCD who received this intervention exhibited more active coping and less negative thinking at posttest compared with a control group that received education about SCD for an equivalent amount of time (Gil et al., 1996). In addition, during laboratory testing of pain, patients from the intervention group were less likely to report pain (to an equivalent laboratory pain stimulus) and had a greater ability to discriminate different pain stimuli at posttest compared with the control group. After a 3-month follow-up period, patients from the intervention group maintained more active coping and a lower tendency to report pain (during the laboratory pain testing) compared with patients from the control group (Gil et al., 2000). The intervention and control groups did not differ in sickle cell–related pain ratings or in health care utilization patterns (emergency department [ED] visits, hospitalizations). However, within-group analyses of only the intervention group revealed that greater coping practice on high pain days was associated with fewer health care contacts (Gil et al., 2000).

In a group of children with SCD, these investigators tested a similar intervention with fewer components (deep breathing/counting for relaxation, imagery, and calming self-statements). Children who received a single intervention session (plus one review session) reported less negative thinking and were less likely to report pain

(when exposed to a laboratory pain stimulus) compared with children in a control group that received standard medical care (Gil et al., 1997). After a 1-month follow-up, children in the intervention group reported more active coping than children in the control group, but the groups did not differ in their responses to laboratory pain stimuli at follow-up. In addition, children in the intervention and control groups did not differ on sickle cell–related pain, SCD-related health practices (e.g., resting, drinking fluids), or health care utilization at follow-up (Gil et al., 2001). Secondary analyses of daily diary records, however, revealed that for those children who were taught intervention strategies, using active coping strategies on days of high pain was associated with fewer health care contacts and less reduction in daily activity. Thus the coping intervention did not produce group differences in pain outcomes or health care utilization. Based on the within-group analyses, however, the authors concluded that coping interventions work when children are in pain and use these strategies; that is, this coping intervention improves health outcomes for those individuals who consistently practice their skills (Gil et al., 2001).

Thomas et al. tested the efficacy of another CBT approach for SCD. In a group setting, patients learned to identify pain cognitions and worked to change the meaning of their pain, as well as their perceptions of control over it. Patients were also given relaxation training and provided with health education. Differences between the two approaches include Gil and colleagues' focusing specifically on coping with pain episodes (whereas the other approach deals more generally with understanding and living with SCD), Gil and colleagues' being briefer, and Gil and colleagues' being conducted through individual sessions (as opposed to groups). In a study by Thomas, Dixon, and Milligan (1999), adolescents and young adult patients with SCD who received 2 months of weekly group CBT sessions reported using more positive coping strategies, engaging in more behavioral activities, and having greater self-efficacy in pain management compared with their counterparts who received either an attention placebo or standard medical care (control group). Patients receiving CBT also reported greater pain control and lower affective ratings of pain compared with those in the control group. No differences were found for sensory pain. In another study by this research group, adult patients with SCD in both the CBT group and the attention placebo group had shorter hospital stays than patients in the wait list control group (Thomas, Wilson-Barnett, & Goodhart, 1998).

Table II. Summary of Psychosocial Intervention Studies for Sickle Cell Disease Pain and Adherence Outcomes

Study	Intervention Type	Design	Outcomes Measured	Findings	Superior to Placebo?	Manualized?	Sample Specified?
Gil, 1996	Cognitive coping skills training	Intervention vs. education control	Lab pain Clinical pain Health care utilization	I < C Lab pain	Yes	Yes	Yes
Gil, 2000	Cognitive coping skills training	Intervention vs. education control	Lab pain Clinical pain Health care utilization	I < C Lab pain Within treatment group: high coping on pain days associated with low health care utilization	Yes	Yes	Yes
Gil, 1997	Cognitive coping skills training	Intervention vs. control (usual care)	Lab pain SCD health practices	I < C Lab pain	Yes	Yes	Yes
Gil, 2001	Cognitive coping skills training	Intervention vs. control (usual care)	Lab pain SCD health practices Clinical pain Health care utilization Activities	No group differences at 1 month FU. Within treatment group: high coping on pain days associated with low health care utilization and less reduction in activities.	No	Yes	Yes
Thomas, 1998	CBT	CBT vs. attention placebo vs. control (usual care)	Pain Health care utilization	C > CBT, AP Duration of hospitalization	Yes (vs. C) No (vs. AP)	No	Yes
Thomas, 1999	CBT	CBT vs. attention placebo vs. control (usual care)	Pain SCD health practices	GBT < C Affective pain	Yes (vs. C) No (vs. AP)	No	Yes
Thomas, 2001	CBT	CBT vs. attention placebo vs. control (usual care)	Cost of health care utilization	Lower in cost at 6 months for CBT, but not for AP or C	No	No	Yes
Thomas, 1984	CBT	Baseline vs. posttreatment	Medications Health care utilization	Fewer ED visits Fewer inpatient days after treatment	Not tested	No	Yes
Anie, 2002	CBT	Baseline vs. posttreatment	Pain Health care utilization SCD health practices	Greater SCD health practices after treatment	Not tested	Yes	Yes
Cozzi, 1987	Biofeedback + relaxation	Baseline vs. posttreatment	Pain Medications Health care utilization	Lower pain frequency and intensity after treatment	Not tested	Yes (instructions via tape)	Yes

Table II. Continued

Study	Intervention Type	Design	Outcomes Measured	Findings	Superior to Placebo?	Manualized?	Sample Specified?
Zeltzer, 1979	Hypnosis	Case study	Health care utilization	Fewer ED visits, hospitalizations, and inpatient days	Not tested	No	Yes
Agarün, 2001	Hypnosis	Case study	Pain Medications	Lower pain and medication use	Not tested	No	Yes
Dings, 1997	Hypnosis	Baseline vs. posttreatment	Pain Medication Activities	Lower pain frequency and intensity after treatment	Not tested	No	Yes
Burghardt-Fitzgerald, 1989	Behavior contract	Case study	Health care utilization	Lower in duration of hospitalizations	Not tested	No	Yes
Berkovitch, 1998	Behavior reward + education	Intervention vs. control (usual care)	Compliance w/penicillin	Higher in compliance in I group after treatment	No	No	Yes
Hazzard, 2002	Education + distraction (computerized)	Intervention vs. control (both groups receive verbal health education)	SCD knowledge	No group differences on knowledge	No	Yes (computerized program)	Yes
Day, 1992	Education	Intervention vs. historical trends	Compliance w/penicillin	Higher in compliance in I group after treatment	Not tested	No	No
Kaslow, 2000	Family intervention	Intervention vs. control (usual care)	SCD knowledge	I > C SCD knowledge	Yes	Yes	Yes
Powers, 2002	Family CBT intervention	Baseline vs. posttreatment	Pain Activities SCD health practices	Majority of patients: Higher SCD health practices after treatment. Some activity same: pain vs. no-pain days.	Not tested	Yes	Yes
Vichinsky, 1982	Social support + counseling	Baseline vs. posttreatment	Health care utilization	Lower in ED visit and hospitalization after treatment	Not tested	No	Yes
Butler & Beltran, 1993	Social support	Baseline vs. posttreatment	Pain	Higher pain self-efficacy Shorter recovery time from pain crises	Not tested	No	Yes
Nash, 1993	Social support	Correlating amount of time in group w/outcomes	Activities Physical symptoms Health care utilization	More time in group, Less interference in activities	Not tested	No	No

I = intervention; C = control; tx = treatment; SCD = sickle cell disease; AP = attention placebo; CBT = cognitive-behavioral therapy; ED = emergency department; FU = follow-up; sx = symptom. Superior to placebo? = Does it meet Chambless criteria of being superior to psychological placebo or alternative treatment, or is it a case study with at least nine single-case design experiments? Manualized? = Does it meet Chambless criteria of treatment being conducted with a treatment manual? Sample specified? = Does it meet Chambless criteria of having characteristics of the sample clearly specified? SCD health practices refers to engagement in SCD-related recommended practices, including resting, drinking fluids, and massage.

Finally, in an examination of health care costs, only those patients in the CBT group showed a decline in health care costs at 6 months postintervention (Thomas, Gruen, & Shu, 2001). No cost differences were found at the 12-month follow-up.

In an early study of cognitive-behavioral techniques that taught biofeedback, progressive relaxation, self-hypnosis, and cognitive strategies, adults with SCD in the group that received 15 sessions of this form of CBT made fewer ED visits, spent fewer days in the hospital as inpatients, and used lower amounts of analgesics at posttreatment compared with pretreatment (Thomas, Koshy, Patterson, Dorn, & Thomas, 1984). However, unlike the studies described above, no control group was included.

One study of manualized CBT from a third group of investigators documented that adults with SCD who received CBT reported increased adherence behaviors (e.g., rest, fluid intake) after treatment compared with pretreatment (Anie et al., 2002). However, there were no pre to post differences in pain or health care utilization patterns. This study is the only one to have found effects on adherence behaviors; however, no control group was used for a comparison.

Other specific techniques in the cognitive behavior intervention category include biofeedback and hypnosis. Children and adolescents (ages 10–20 years) with SCD who received 12 sessions of biofeedback involving thermal and EMG (electromyography) training showed decreases in anxiety, self-reported pain, and medication use from pre- to posttraining. No differences were found for number of hospitalizations (Cozzi, Tryon, & Sedlacek, 1987). The hypnosis studies include three case studies in patients 9–20 years old with SCD. Patients who received hypnosis reduced their analgesic use and number of hospitalizations in one study (Zeltzer, Dash, & Holland, 1979) and their pain symptoms and analgesic use in another (Agargün, Öner, & Akbayram, 2001) from pre- to posthypnotic treatment. A larger study, of self-hypnosis taught in small groups over an 18-month period, found that SCD patients ages 5–51 years who received hypnosis training reduced their number of pain days relative to baseline (under standard medical care) (Dinges et al., 1997). The more sessions patients attended in this 18-month program, the more pain-free days they reported. However, intensity of pain during pain episodes was not reduced, suggesting that perhaps hypnosis may be most effective for mild to moderate pain.

Finally, a review of empirically supported treatments for disease-related pain concluded that hypnosis and progressive muscle relaxation showed evidence of treatment efficacy that warranted further study, although the evidence did not currently meet the standard criteria for an empirically supported treatment (Walco et al., 1999). In addition, one study based on patient interviews reported that patients who successfully coped with SCD identified distraction, biofeedback, relaxation, imagery, hypnosis, reinterpreting pain, and proactive coping as some of the strategies that they found helpful for coping with pain (Fox & Ingram, 1999).

Overall, Gil and colleagues' studies on cognitive coping strategies and the Thomas et al. studies on cognitive behavioral therapy are among the most scientifically rigorous of any of the intervention studies reviewed in this paper because (1) they included a control group, (2) they involved random assignment of patients to treatment or control groups, and (3) many of the studies controlled for time spent with a therapist and for SCD-related information in their control group. The Gil et al. studies of cognitive coping strategies meet the APA/Chambless criteria in being superior to psychological placebo (education control), being manualized, and having well-described sample characteristics. To be labeled *well-established*, the APA/Chambless criteria require that at least two groups of investigators demonstrate intervention efficacy. The Thomas et al. CBT studies show significant intervention effects but do not meet the Chambless criteria in that intervention effects were not superior to psychological placebo (intervention was better than control but not different from attention placebo), and no manuals were described. Nonetheless, as a group, these cognitive-behavioral techniques meet the APA/Chambless criteria for being labeled *probably efficacious*, in having at least two studies demonstrating that treatment is more effective than wait list control.

It should be noted that the strongest effects in the above studies were found for group differences between treatment and psychological placebo on laboratory pain measures. However, beneficial effects on health care utilization patterns were noted when compared with a usual-care control group in the Thomas et al. studies and among those children in the Gil et al. intervention who practiced the intervention strategies, suggesting that cognitive-behavioral interventions may be beneficial for reducing health care contacts, as well as for reducing pain.

Those studies utilizing very specific techniques, such as hypnosis or biofeedback, were limited in not including a control group, making it difficult to ascertain the extent to which changes were due to the intervention specifically or to natural variations in pain over time. In addition, studies that did not include a psychological placebo were not able to clarify whether effects were due to the intervention technique specifically or to the attention from and time spent with a therapist. In addition, the hypnosis intervention studies did not describe the use of a treatment manual. Thus none of these studies met the APA/Chambless criteria for empirically supported studies.

Overall, across all studies that utilize cognitive-behavioral techniques, there is support for the notion that this type of intervention for treating sickle cell pain falls into the *probably efficacious* category. These studies had specific goals of reducing pain, rather than changing behaviors related to education/adherence. Nonetheless, several of the above studies included measures of coping with a subscale regarding behaviors such as resting, drinking fluids, and massage. These behaviors form part of physician-recommended daily health practices (NHLBI, 1995). Thus, we could explore whether cognitive-behavioral techniques, although not the focus of the interventions, had effects on adherence measures (listed in Table II as those studies with “SCD health practices” in the “Outcomes Measured” column). There were mixed results, with one study demonstrating increases in children’s adherence behaviors over the course of a cognitive-behavioral intervention (Anie et al., 2002), whereas two other studies that included control groups did not find differences between the intervention and control groups on adherence behaviors posttreatment (Gil et al., 1997; Thomas et al., 1999). However, it should be noted that these behaviors were assessed in response to pain episodes, rather than on a daily basis. Some researchers have conceptualized these strategies as less effective than active coping attempts for dealing with pain episodes. Although there is currently no evidence according to the APA/Chambless criteria that cognitive-behavioral techniques are efficacious for adherence behaviors, it is also the case that this issue has not been the focus of either the content of the above interventions or the outcomes measured.

Future Recommendations

Well-designed intervention studies in the category of cognitive-behavioral techniques have already been conducted. Our primary recommendation for future in-

tervention studies is to clarify the extent of effects using cognitive-behavioral techniques. Future studies are needed to determine whether variations in existing protocols will produce differential effects in terms of clinical pain and health care utilization patterns for those who receive intervention compared with those who receive a psychological placebo. This could include varying the length or timing of intervention, and/or modifying the content of intervention sessions to address long-term pain outcomes. Future studies should also explore whether cognitive behavioral techniques have effects on daily adherence behaviors. This might include broadening the scope of some intervention protocols to include strategies for living with and managing a chronic illness such as SCD on a daily basis.

Interventions Aimed at Behavioral Change

Included in this category are interventions typically aimed at modifying behavior, including behavioral contracts/rewards and education. Behavioral contracts involve setting up a system whereby children are rewarded for behaviors that are incompatible with pain (e.g., being more active). They are thought to be a means toward reducing pain behaviors and analgesic use and toward helping children to gain confidence in their ability to control pain experiences. Family education about SCD typically covers information about what SCD is and practical knowledge about how to detect early symptoms and comply with medical recommendations regarding health practices. Although the two intervention approaches can be quite different, they share a common goal (changing behavior) and are sometimes used in combination in intervention studies. Thus they are included in one category in this review. The rationale for this category of intervention is based on the notion that recognizing and acting quickly on potentially dangerous symptoms (e.g., fever, difficulty breathing) may reduce the severity of the pain episode and/or the possibility of other medical complications (e.g., rapid enlargement of spleen that could lead to the need for surgery) (Day, Brunson, & Wang, 1992; Vichinsky & Lubin, 1987). In addition, increased knowledge about the pathophysiology of SCD is thought to increase the likelihood of adherence to medical recommendations (e.g., the more parents understand about the dangers of infection in young children with SCD, the more they will ensure that their children take antibiotics daily).

Case studies have reported the utility of behavioral contracts in SCD for reducing duration of hospitalizations (Burghardt-Fitzgerald, 1989). It should be noted

that with respect to behavioral contract techniques, empirical support derives more from literature on chronic pain, as well as behavioral problems in children (e.g., aggression) (Kazdin, 1996; Zeltzer, Bush, Chen, & Rival, 1997), than from testing in the SCD population.

Behavioral rewards have also been used in young children to encourage adherence to daily antibiotic regimens for SCD. One study (Berkovitch et al., 1998) combined behavioral rewards (giving children a sticker for each day they took their antibiotic) with education about the risks of infection and benefits of antibiotics in SCD. Families that received the 8-week intervention had children who improved in antibiotic compliance pre- to postintervention. However, although the within-group analyses demonstrated that the intervention group improved over time, the between-group analyses did not find differences at posttreatment between the intervention group and a control group that received usual medical care.

Other education interventions for children have capitalized on recent technological advances. The Starbright Foundation developed an in-hospital computer network in which children ages 8–18 with SCD (or asthma) who had been hospitalized were given access to computers with Internet access for a 3-day period. These computers were set up to provide health education via the Internet as well as from Starbright programs (e.g., “The Sickle Cell Slime-O-Rama Game” [Hazzard, Celano, Collins, & Markov, 2002]). The networked computers also allowed children to interact with other hospitalized children via videoconferencing, chatrooms, and e-mail. Finally, the computers contained recreational games to allow children to engage in distraction while in the hospital. The group that received the computer intervention was compared with a control group that received the same verbal health education from a nurse or other hospital staff as the intervention group, but without access to the computer network. Children who received the intervention reported less negative coping and greater perceptions of social support compared with the control group. However, no differences were found on SCD knowledge scores, and no pain outcomes were measured (Hazzard et al., 2002). The authors reported that the children gave very positive evaluations of the Starbright computer program and spent an average of almost 4 hours (over a 3-day period) using the computer.

Others have developed booklets for parents with children who have been diagnosed with SCD. “Your Child and Sickle Cell Disease” defines SCD and gives practical information related to SCD (e.g., how to use

a thermometer). “Family Connection” explains genetics and the probabilities of inheritance of SCD in any given child. “Newborn Screening for Sickle Cell Disease” discusses the feelings parents often experience upon learning that their child has SCD. A study that incorporated these booklets, as well as follow-up phone calls and home visits from nurses over a period of 6–9 months found that parents who participated in this study had children with lower infection rates than had been reported historically in other studies. Parents in the intervention group also demonstrated high levels of knowledge about SCD (Day et al., 1992). However, no control group was included in this study.

Overall, although several of the studies demonstrated improvements within an intervention group over time, none of the behavioral or education intervention studies demonstrated superiority over either a psychological placebo or a usual-care control group in terms of SCD education/adherence outcomes. In addition, with the exception of the Starbright computer program (which we counted as manualized), none of the intervention studies reported using manualized treatment. Thus, none of the behavioral change interventions meet the APA/Chambless criteria for being a well-established empirically supported intervention.

Future Recommendations

A great deal of time and effort has already gone into developing educational materials for parents and children with SCD. In addition, behavioral contracts/rewards are appealing because they have the potential to affect not only pain behaviors, but also adherence-related outcomes (e.g., use of rewards for drinking the recommended amount of liquids daily). Our recommendations for this category of intervention include: (1) Future studies of behavioral and education interventions should always include a control group, and preferably a psychological placebo; (2) additional studies are needed that measure a full range of adherence behaviors, based on the national guidelines for daily SCD practices; and (3) future studies are needed to determine the extent to which adherence behaviors are associated with pain outcomes, and thus whether behavioral change interventions have the potential to also affect pain experiences.

Social Support Interventions

The intervention studies in this category include those that involve the use of other people to help an individual with SCD, such as support and self-help groups, as well as family intervention. The rationale

behind the use of support and self-help groups derives in part from observational studies that demonstrate that social support is a buffer for individuals against poor health or early mortality (Berkman & Syme, 1979; House, Landis, & Umberson, 1988). In addition, researchers have specifically advocated social support interventions for individuals with SCD based on the observation that ethnic minorities often rely more on respected laypersons for health information than medical professionals (Holmes, Hatch, & Robinson, 1991). Self-help groups involve training laypersons about SCD so that they can serve as informal conduits of information for groups of individuals with the disease (Holmes et al., 1991). In addition, the emphasis on close connections with immediate and extended family within the African American community suggests the importance of utilizing social connections to optimize SCD care and daily management. Similarly, family interventions for SCD build on the strength of family values typically found in the African American community (Collins et al., 1997; Kaslow et al., 1997). Both types of interventions tend to combine a focus on interpersonal relationships with education about SCD (Collins et al., 1997; Kaslow & Brown, 1995).

The most rigorous study in the social support category is a study of family therapy, conducted with an African American therapist using a manual. The protocol also included relaxation and imagery techniques. Patients who received family therapy improved in SCD knowledge posttreatment and at the 6-month follow-up compared with patients in a control group who interacted with the same staff but did not receive a structured intervention (Kaslow et al., 2000). A second study involved a pilot investigation describing a comprehensive family intervention approach that taught parents and children cognitive-behavioral techniques. This study could also be considered under the "Cognitive-Behavioral Techniques" section above; however, we include it here because of its emphasis on the family, thus being distinct from intervention with only the individual patient. Two of the 3 patients increased their adherence behaviors (e.g., rest, fluid intake) from pre- to posttreatment and also at 11 weeks posttreatment. Although children reported decreased activity on pain days compared with no-pain days overall, activity levels in certain domains remained similar across pain and no-pain days (Powers, Mitchell, Graumlich, Byars, & Kalinyak, 2002). This study, however, had only 3 participants and did not describe comparisons with

a control group. The other studies found on family interventions for SCD contained suggestions and guidelines but were not new empirical studies (Collins et al., 1997; Kaslow et al., 1995). For example, these articles discussed the necessity of training therapists to conduct culturally sensitive family interventions, the importance of making manualized treatments flexible so that they apply to each family, and the reality of other life problems that African Americans of low socioeconomic status (SES) face that could interfere with their participation in family interventions for SCD (Kaslow et al., 1997). These issues are all important to consider in future family intervention studies but did not contribute to our determination of empirical evidence for social support interventions.

With respect to such interventions, patients who participated in a social support group reported improvement in self-efficacy related to pain management, and shorter recovery time from pain crises; however, this evidence was based on qualitative data only (Butler & Beltran, 1993). A second support intervention study included a combination of social support plus counseling. After the intervention, patients had fewer ED visits and hospitalizations compared with pretreatment (Vichinsky, Johnson, & Lubin, 1982). However, this study did not include a control group. Finally, a study of sickle cell self-help support groups reported that group participation reduced feelings of depression and that the longer patients participated in the group, the fewer psychological symptoms they reported and the less interference they perceived SCD had in their lives (Nash & Kramer, 1993). However, length of time in the group was not related to physical symptoms or health care utilization patterns. This study also did not use a control group for comparison.

One study based on patient interviews reported that patients found social support groups and exposure to good role models to be helpful for SCD pain (Fox & Ingram, 1999). Social support groups encourage members not only to educate themselves about SCD but also to become more involved in identifying and reforming problem areas within the health care system in its treatment of SCD (Shelley, Kramer, & Nash, 1994). In addition, these interventions improved the psychological states of parents (reduced their anxiety) compared with a control group that had access to other SCD parents' phone numbers but were not part of an active support group (Ireys, Chernoff, DeVet, & Kim, 2001).

Among the social support interventions, the strongest study is the family intervention one because it

included a control group (usual medical care) and utilized a treatment manual. Although very well designed, the focus of this study was not on SCD pain or adherence behaviors. There is evidence that social support interventions may improve SCD knowledge (based on this one study), but there is no evidence thus far of social support interventions being efficacious according to the APA/Chambless criteria for SCD-related pain or adherence behaviors. However, as with the behavioral change interventions, it may be the case that social support interventions are efficacious but that not enough sufficiently rigorous intervention studies have been designed to demonstrate this effect.

Future Recommendations

The components of social support interventions and the methods by which these interventions should be administered already have been carefully considered by researchers. In addition, interventions that utilize family members and support networks have the potential advantage of being more cost-effective in the long term compared with other types of interventions. We recommend that future studies of social support and family interventions (1) need to include control groups and, if possible, attention placebos, (2) directly compare social support groups with control groups on a variety of pain and health care utilization outcomes, and (3) test whether improving social support or family relationships also helps patients to comply more rigorously with recommended daily practices.

Summary and Future Directions

Although there is almost universal acceptance of the idea that treatment for SCD should include psychosocial approaches, much of the research in this area suffers from methodological limitations that make it difficult to discern which psychosocial approaches are best for reducing SCD pain and increasing SCD adherence behaviors. A number of intervention studies for SCD present qualitative data or do not include control groups. The most promising interventions as a group, from a methodological standpoint, are those described in the "Cognitive-Behavioral Techniques" section above. There is evidence that cognitive-behavioral interventions fall into the category of *probably efficacious*, according to the APA/Chambless criteria. One group of investigators (Gil et al.) met the standards of the Chambless criteria (superior to psychological placebo, manualized, and well-specified sample); how-

ever, the requirements for a well-established intervention dictate that at least two separate groups of investigators publish studies that meet these standards. The second group (Thomas et al.) did not find cognitive-behavioral interventions to be superior to a psychological placebo (only to wait list control), thus making this category of intervention *probably efficacious*.

The studies utilizing behavioral change approaches targeted SCD education/adherence behaviors but did not demonstrate superiority over a control group. There was within-group evidence, however, that those who received interventions such as behavioral rewards and education showed improvements over time (Berkovitch et al., 1998; Burghardt-Fitzgerald, 1989; Day et al., 1992). In addition, evidence from some of these studies suggests that this type of intervention may be beneficial for psychological outcomes (e.g., coping, social support) (Hazzard et al., 2002). However, given that the APA/Chambless criteria do not include within-group changes over time, currently there is no evidence that behavioral change approaches are an empirically supported intervention for SCD education/adherence outcomes or pain.

Social support interventions fared similarly to behavioral change interventions. The studies for social support/self-help groups did not include a control group (Nash et al., 1993; Vichinsky et al., 1982). The data for family interventions consisted of one very solid intervention study that does meet the standards of the APA/Chambless criteria, but this study tested SCD knowledge, not pain or behavioral adherence as outcomes (Kaslow et al., 2000). Thus, currently, there is no evidence that social support groups are an empirically supported intervention for SCD-related pain or adherence behaviors.

The most obvious next step for future studies of SCD interventions is to engage in more scientifically rigorous testing of the above intervention approaches. Ideas about psychosocial interventions for SCD are prevalent, as evidenced by the collection of published intervention studies in this area. However, this review has highlighted the small proportion of these published studies that are scientifically rigorous. This indicates the need for more comprehensive testing of existing interventions, rather than a search for new intervention approaches. For example, it remains unclear whether behavioral change interventions and social support interventions would be efficacious if rigorously tested or are in fact less beneficial than cognitive-behavioral techniques for SCD pain and adherence outcomes.

Second, it is important for future intervention research to target a broad range of SCD-related outcomes. We need to determine whether psychological interventions can make a difference in the daily experiences with disease that these children with a chronic, unremitting illness have. The majority of studies that have investigated SCD outcomes have focused on pain outcomes such as severity, frequency, or ED visits and hospitalizations. These types of interventions are aimed at managing pain crises when they occur. However, national guidelines for home care of SCD also emphasize general daily practices (e.g., drinking plenty of fluids, avoiding extreme temperatures). Interventions targeted at such adherence behaviors would correspond to a prevention approach that has a focus on preventing future pain episodes and other complications by teaching and encouraging recommended daily practices. The main intervention in this domain tested parental adherence to daily antibiotic regimens for children (Berkovitch et al., 1998). However, these daily regimens usually stop after the age of 5, whereas other healthy practices are expected to be maintained throughout childhood. Future intervention studies need to test the effects of psychosocial interventions on the daily health behaviors that families with SCD engage in, and whether changes in health behaviors produce changes in SCD pain outcomes over time.

In addition, understanding the specificity of interventions provides another future direction for research. Not all interventions will be equally efficacious for all outcomes. Some may work better for psychosocial outcomes, whereas others may work for pain crises, and still others may be best for daily management practices. Thus it would be important to determine the overall effects of a specific intervention (across a number of domains), and then to determine whether certain combinations of intervention techniques might be most useful for maximizing improvement across multiple domains. To support this goal, it would be important for researchers testing different interventions to be consistent in the outcomes they measure. This would allow researchers to compare across studies in determining the specific effects of different intervention approaches.

Lastly, greater consideration needs to be given to the components included in an intervention package. Intervention components tend to be aimed at either the parent or the child, with little mention of adjusting the techniques for differences by age in children's cognitive and behavioral skills. In addition, recognizing at what ages the expectations for adherence lie with the parents

and at what ages they can be transferred to the child is important both for designing behavioral interventions and for appropriately measuring outcomes. Finally, being conscious of the larger social environment in designing intervention strategies is necessary (Kato & Mann, 1996). Given the centrality of the extended family within African American culture (Kaslow et al., 1997), interventions aimed at only the child may not be well received, regardless of how efficacious they may be scientifically. Instead, involving extended family (and/or community) members in an intervention may facilitate acceptance and implementation of the intervention by the family. Lastly, addressing the larger life issues that many of these African American families, who are often lower in SES, face is important to the success of these interventions. That is, no amount of education may change behavior until other pressing issues such as poverty and housing needs for families are addressed.

In sum, efficacious psychosocial interventions that are delivered in a manner appropriate for children with SCD can optimize their well-being. Current empirical evidence suggests that cognitive-behavioral techniques are probably efficacious for reducing children's SCD pain. Other categories of interventions such as social support or behavioral change may prove to also be beneficial but need to be more rigorously tested.

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