



Effective Health Care

Mental Health Support for Children and Adults with Sickle Cell Disease

Results of Topic Selection Process & Next Steps

The nominator, a representative of the Advancing Sickle Cell Advocacy Project (ASAP), is interested in an AHRQ evidence review to gather evidence for mental health therapy and support for children and adults with Sickle Cell Disease (SCD) to help decrease the distress symptoms that often accompany the disease. However, the topic is not feasible for a full systematic review due to the limited data available for a review at this time. No further activity on this topic will be undertaken by the Effective Health Care (EHC) Program.

The following AHRQ systematic review may be useful to the nominator:

- Meditation Programs for Psychological Stress and Well-Being
<https://www.effectivehealthcare.ahrq.gov/search-for-guides-reviews-and-reports/?pageaction=displayproduct&productID=1830> "

Topic Brief

Topic Name: Mental Health Support for Children and Adults with Sickle Cell Disease

Topic #: 0710

Nomination Date: 10/11/2016

Topic Brief Date: 12/01/2016

Authors:

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Conflicts of Interest: None of the investigators have any affiliations or financial involvement that conflicts with the material presented in this report.

Summary of Key Findings

- Appropriateness and importance: The nomination is both appropriate and important.
- Duplication: A new evidence review on this topic would not be duplicative. We identified three potentially relevant evidence reviews; however they do not address the range of outcomes and populations of interest to the nominator. A [2015 Cochrane Review](#) examining psychological therapies for sickle cell disease and pain and a [2014 Cochrane Review](#) examining psychological therapies for chronic and recurrent pain (including SCD) concluded that there was limited evidence for the efficacy of psychological therapies in sickle cell disease on pain levels. However, the nominator is interested in more outcomes than pain. A third review published in 2012 reported unclear efficacy for peer-support groups, educational/psycho-educational programs, and skills-based programs for children and young adults (10-30 years old) with chronic illness (including SCD) on their quality of life, among other outcomes.

- Impact: Because there is wide practice variation and no current standard of care, an evidence review would have high impact on comprehensive care plans for children and adults with SCD.
- Feasibility: An AHRQ evidence review on this topic is not feasible at this time because of the limited number of relevant studies identified and range of interventions studied.
 - *Size/scope of review*: From PubMed, we estimate that the number of relevant studies published between October 2011 and October 2016 may be nine across both key questions. The studies focus on a range of interventions, including cognitive behavioral therapy (CBT), social support, and positive family functioning.
 - *Clinicaltrials.gov*: The future of this topic is promising. We identified eight unique trials relevant to the key questions, including three trials which recently began to recruit, one trial in progress, and four which were recently completed. The trials cover a wide range of therapies, including music therapy, computerized CBT, and family support.

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Introduction

Sickle Cell Disease (SCD) is an inherited group of disorders, which causes red blood cells (RBC) to become crescent, or sickle, in shape.¹ The sickle-shaped cells have a much shorter period of full functionality than healthy, round RBCs, often leaving its host with a shortage of RBCs causing sickle cell anemia. These misshapen cells can also block blood flow causing random bouts of intense pain. SCD patients are more vulnerable to bone and muscle pain, organ damage, and severe infections.¹ Various forms of therapy and support can be used as complementary treatment to help patients cope with their disease and manage psychological comorbidities.

Topic nomination #0710 was received on October 11, 2016. It was nominated by the Advancing Sickle Cell Advocacy Project (ASAP). Due to the broad scope of the original nomination, the nominator was able to narrow down and define the scope of the questions. The questions for this nomination are:

Key Question 1. What are the benefits and harms of providing mental health support to children with sickle cell disease as part of a comprehensive treatment plan?

Key Question 2. What are the benefits and harms of providing mental health support to adults with sickle cell disease as part of a comprehensive treatment plan?

To define the inclusion criteria for the key questions we specify the population, interventions, comparators, and outcomes (PICO) of interest. See Table 1.

Table 1. Key Questions with PICOs

Key Question	1. What are the benefits and harms of providing mental health support to children with sickle cell disease as part of a comprehensive treatment plan?	2. What are the benefits and harms of providing mental health support to adults with sickle cell disease as part of a comprehensive treatment plan?
Population	Children (4-17 years old) with all types of Sickle Cell Disease	Adults (18-45 years old) with all types of Sickle Cell Disease
Intervention	Mental health support (eg, psychotherapy, group support, counseling)	Mental health support (eg, psychotherapy, group support, counseling)
Comparators	Other mental health therapies, passive education (eg, pamphlet), no therapy	Other mental health therapies, passive education (eg, pamphlet), no therapy
Outcomes	Decrease in distress symptoms (eg, anxiety, fear, depression), increase in coping skills, adverse events	Decrease in distress symptoms (eg, anxiety, fear, depression), increase in coping skills, adverse events

Methods

To assess topic nomination #0710 *Mental Health Support for Children and Adults with Sickle Cell Disease* for priority for a systematic review or other AHRQ EHC report, we used a modified process based on established criteria. Our assessment is hierarchical in nature, with the findings of our assessment determining the need for further evaluation. Details related to our assessment are provided in Appendix A.

1. "Determine the *appropriateness* of the nominated topic for inclusion in the EHC program.
2. "Establish the overall *importance* of a potential topic as representing a health or "healthcare issue in the United States. "
3. "Determine the *desirability of new evidence review* by examining whether a new "systematic review or other AHRQ product would be duplicative. "
4. "Assess the *potential impact* a new systematic review or other AHRQ product.

5. "Assess whether the *current state of the evidence* allows for a systematic review or other AHRQ product (feasibility).
6. "Determine the *potential value* of a new systematic review or other AHRQ product.

Appropriateness and Importance

We assessed the nomination for appropriateness and importance (see Appendix A).

Desirability of New Review/Duplication

We searched for high-quality, completed or in-process evidence reviews pertaining to the key questions of the nomination. Table 2 includes the citations for the reviews that were determined to address the key questions.

Impact of a New Evidence Review

The impact of a new evidence review was assessed by analyzing the current standard of care, the existence of potential knowledge gaps, and practice variation. We considered whether it was hypothetically possible for this review to influence the current state of practice through various dissemination pathways (practice recommendation, clinical guidelines, etc.).

Feasibility of a New Evidence Review

We conducted a literature search for randomized controlled trials in PubMed from October 2011-October 2016. Because a small number of articles were identified, we reviewed all abstracts for inclusion and classified identified studies by study design, to assess the size and scope of a potential evidence review. See *Table 2, Feasibility Column, Size/Scope of Review Section* for the citations of included studies. See Appendix C for the PubMed search strategy and links to the ClinicalTrials.gov search.

Compilation of Findings

We constructed a table outlining the selection criteria as they pertain to this nomination (see Appendix A).

Results

Appropriateness and Importance

This topic is appropriate and important. In the United States alone, between 70,000-100,000 individuals have SCD, and approximately 3 million have sickle cell trait.² Couples who both carry the sickle cell trait have a 1-in-4 chance of having a child with SCD. See Appendix A for details.

Desirability of New Review/Duplication

We identified three relevant evidence reviews, but they do not address the range of outcomes and populations of interest to the nominator. A 2015 Cochrane review examining psychological therapies for SCD-associated pain concluded that there was limited evidence for the efficacy of psychological therapies in sickle cell disease on pain levels (KQ 2).³ A 2014 Cochrane review also reported that there was limited evidence to support psychological therapies as adjuvant treatment for chronic and recurrent pain (KQ 1).⁴ Following suit, the third review, from 2012, also reported unclear efficacy for a variety of psycho-social interventions for children and young adults (10-30 years old) with SCD (KQs 1 & 2) on quality of life.⁵

Impact of a New Evidence Review

A new evidence review on including mental health support as part of a comprehensive care plan for children and adults with SCD would have a high impact. There is practice variation because of a lack of guidance about the inclusion of mental health support in care plans. At this time, mental health support is only used at the patient, the patient's family, or doctor's discretion. It is not part of most care plans. There is no current standard of care.

Feasibility of a New Evidence Review

A comprehensive AHRQ product covering all key questions may not be feasible at this time because of the small number of relevant studies identified and the range of interventions studied. We identified seven published studies⁶⁻¹² examining the effect of various mental health support techniques in children with SCD (KQ 1). Only two studies^{13,14} examining mental health support in adults with SCD were identified (KQ 2). We identified eight unique trials relevant to the key questions, including three trials which recently began to recruit, one trial in progress, and four which were recently completed. The trials cover a wide range of therapies, including music therapy, computerized cognitive behavioral therapy, and family support. These trials may provide the additional data needed for a comprehensive evidence review in the coming years.

Table 2. Key questions with the identified corresponding evidence reviews and original research

Key Question	Duplication (Completed or In-Process Evidence Reviews)	Feasibility (Published and Ongoing Research)
KQ 1: Children	Total number of completed or in-process systematic reviews: 2 <ul style="list-style-type: none"> • Cochrane: 1⁴ • Other: 1⁵ 	<u>Size/scope of review:</u> Relevant Studies Identified: 7 <ul style="list-style-type: none"> • Pre-Post: 1⁶ • Prospective Cohort: 1⁷ • Qualitative: 2^{8,9} • Validation: 2^{10,11} • Post Hoc: 1¹² <u>Clinicaltrials.gov</u> Relevant Trials Identified: 4 <ul style="list-style-type: none"> • Recruiting: 2^{15,16} • Complete: 2^{17,18}
KQ 2: Adults	Total number of completed or in-process systematic reviews: 2 <ul style="list-style-type: none"> • Cochrane: 1³ • Other: 1⁵ 	<u>Size/scope of review</u> Relevant Studies Identified: 2 <ul style="list-style-type: none"> • Pre-Post: 1¹³ • Post Hoc: 1¹⁴ <u>Clinicaltrials.gov</u> Relevant Trials Identified: 5 <ul style="list-style-type: none"> • Recruiting: 2^{15,19} • Active, not recruiting: 1²⁰ • Complete: 2^{21,22}

Summary of Findings

- Appropriateness and importance: The nomination is both appropriate and important.
- Duplication: A new evidence review on this topic would not be duplicative. We identified three potentially relevant evidence reviews but they do not address the range of outcomes and populations of interest to the nominator. A [2015 Cochrane Review](#) examining psychological therapies for sickle cell disease and pain and a [2014 Cochrane Review](#) examining psychological therapies for chronic and recurrent pain (including SCD) concluded that there was limited evidence for the efficacy of psychological therapies in sickle cell disease on pain levels. However, the nominator is interested in more outcomes than pain. A third review reported unclear efficacy for peer-support groups, educational/psycho-educational programs, and skills-based programs for children and young adults (10-30 years old) with chronic illness (including SCD) on their quality of life, among other outcomes.

- Impact: Because there is wide practice variation and no current standard of care, an evidence review would have high impact on comprehensive care plans for children and adults with SCD.
- Feasibility: An AHRQ evidence review on this topic is not feasible at this time because of the limited number of relevant studies identified and range of interventions studied.
 - *Size/scope of review*: From PubMed, we estimate that the number of relevant studies published between October 2011 and October 2016 may be nine across both key questions. The studies focus on a range of interventions, including cognitive behavioral therapy (CBT), social support, and positive family functioning.
 - *Clinicaltrials.gov*: The future of this topic is promising. We identified eight unique trials relevant to the key questions, including three trials which recently began to recruit, one trial in progress, and four which were recently completed. The trials cover a wide range of therapies, including music therapy, computerized CBT, and family support.

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Appendices

Appendix A: Selection Criteria Summary '

Appendix B: Search Strategy & Results (Feasibility)

Appendix A. Selection Criteria Summary (

Selection Criteria	Supporting Data
1. Appropriateness	
1a. Does the nomination represent a health care drug, intervention, device, technology, or health care system/setting available (or soon to be available) in the U.S.?	Yes, this topic represents a health care drug and intervention available in the U.S.
1b. Is the nomination a request for a systematic review?	Yes, this topic is a request for a systematic review.
1c. Is the focus on effectiveness or comparative effectiveness?	Yes, the focus of this review is on effectiveness.
1d. Is the nomination focus supported by a logic model or biologic plausibility? Is it consistent or coherent with what is known about the topic?	Yes, it is biologically plausible. Yes, it is consistent with what is known about the topic.
2. Importance	
2a. Represents a significant disease burden; large proportion of the population	This topic represents a moderate burden. In the united states alone, between 70,000-100,000 individuals have SCD, and approximately 3 million have sickle cell trait. ²
2b. Is of high public interest; affects health care decision making, outcomes, or costs for a large proportion of the US population or for a vulnerable population	Yes, this topic is of high public interest. SCD is an expensive disease to treat, and disproportionately affects African Americans (1 out of 365 births). ²³
2c. Represents important uncertainty for decision makers	Yes, this topic represents important uncertainty for decision makers.
2d. Incorporates issues around both clinical benefits and potential clinical harms	Yes, this nomination addresses both benefits and potential harms of mental health support for patients with SCD.
2e. Represents high costs due to common use, high unit costs, or high associated costs to consumers, to patients, to health care systems, or to payers	Yes, this topic represents high costs for insurance and patients. SCD requires a comprehensive care team, and expensive treatments.
3. Desirability of a New Evidence Review/Duplication	
3. Would not be redundant (i.e., the proposed topic is not already covered by available or soon-to-be available high-quality systematic review by AHRQ or others)	We identified three potentially relevant evidence reviews. A 2015 Cochrane review ³ examining psychological therapies for sickle cell disease and pain, and a 2014 Cochrane review ⁴ examining psychological therapies for chronic and recurrent pain (including SCD) concluded that there was limited evidence for the efficacy of psychological therapies in sickle cell disease on pain levels. However, the nominator is interested in more outcomes than just pain. A third review reported unclear efficacy for peer-support groups, educational/psycho-educational programs, and skills-based programs for children and young adults with chronic illness (including SCD) on their quality of life, among other outcomes. ⁵
4. Impact of a New Evidence Review	

4a. Is the standard of care unclear (guidelines not available or guidelines inconsistent, indicating an information gap that may be addressed by a new evidence review)?	Formal studies in mental health support in patients with SCD have only recently gained recognition as a legitimate therapeutic device in comprehensive care, and there exists no standard of care.
4b. Is there practice variation (guideline inconsistent with current practice, indicating a potential implementation gap and not best addressed by a new evidence review)?	At this time, mental health support is only used at the patient, the patient's family, or doctor's discretion. It is not part of most care plans. There is wide practice variation.
5. Primary Research	
5. Effectively utilizes existing research and knowledge by considering: - Adequacy (type and volume) of research for conducting a systematic review - Newly available evidence (particularly for updates or new technologies)	<p><i>Size/scope of the review:</i> From PubMed, we estimate that the number of relevant studies published between October 2011 and October 2016 may be nine across both key questions.⁶⁻¹⁴ The studies focus on a range of interventions, including cognitive behavioral therapy (CBT), social support, and positive family functioning.</p> <p><i>Clinicaltrials.gov:</i> We identified eight unique trials relevant to the key questions, including three trials that recently began to recruit,^{15,16,19} one trial in progress,²⁰ and four that were recently completed.^{17,18,21,22} The trials cover a wide range of therapies, including music therapy, computerized cognitive behavioral therapy, and family support.</p>

Appendix B. Search Strategy & Results (Feasibility)

Topic: Sick Cell and Mental Health Support Date: October 21, 2016 Database Searched: MEDLINE (PubMed)	
Concept	Search String
Sickle Cell	("Anemia, Sick Cell"[Mesh]) OR "sickle cell"[Title/Abstract]
AND	
Mental Health Support <ul style="list-style-type: none"> • Psychotherapy • Group support • Counseling 	(((((("Psychotherapy"[Mesh]) OR "Self-Help Groups"[Mesh]) OR "Counseling"[Mesh]) OR "psychology" [Subheading])) OR (("mental health"[Title] OR psychotherapy[Title] OR therapy[Title] OR "group support"[Title] OR counseling[Title]))
NOT	
Not Editorials, etc.	(((((("Letter"[Publication Type]) OR "News"[Publication Type]) OR "Patient Education Handout"[Publication Type]) OR "Comment"[Publication Type]) OR "Editorial"[Publication Type])) OR "Newspaper Article"[Publication Type]
Limit to last 5 years ; human ; English ;	Filters activated:
N=272	
Systematic Review N=17	PubMed subsection: Systematic [sb]
Randomized Controlled Trials N=96	Cochrane Sensitive Search Strategy for RCT's: (((((((groups[tiab])) OR (trial[tiab])) OR (randomly[tiab])) OR (drug therapy[sh])) OR (placebo[tiab])) OR (randomized[tiab])) OR (controlled clinical trial[pt])) OR (randomized controlled trial[pt])
Other N=159	

Clinicaltrials.gov was searched on October 21, 2016.

104 studies found for: sickle cell | NOT drug | Studies received on or after 01/01/2011

https://clinicaltrials.gov/ct2/results?term=&recr=&type=&rslt=&age_v=&gndr=&cond=sickle+cell&intr=NOT+drug&titles=&outc=&spons=&lead=&id=&state1=&cntry1=&state2=&cntry2=&state3=&cntry3=&locn=&rcv_s=01%2F01%2F2011&rcv_e=&lup_s=&lup_e=