Support and information needs identified in a survey of adults with sickle cell disease

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Abstract

Newborn screening and early treatment for sickle cell disease has dramatically improved outcomes for impacted children, but adults with the disease live with chronic and acute pain, organ damage, a high risk of mortality, and diminished quality of life. Adults’ access to quality care and knowledgeable providers is limited compared to other genetic diseases.

Adults living with the disease and associated with two community-based organizations were asked to complete a survey describing their health education and information needs with regard to sickle cell, as well as their challenges finding supportive and knowledgeable care. Nearly 600 responses were received.

Respondents describe their information needs, such as the best care providers nearby, insurance options, connections to support groups. Questions also address the utility of different types of support services, as ranked by respondents. Respondents also described actions they take when experiencing a vaso-occlusive/pain crisis and answered an open-ended question about their greatest needs to improve quality of life.

Findings suggest that support groups along with knowledgeable and compassionate care are important identified needs to those living with sickle cell disease. Respondents noted reliance on friends and family as well as social media for information about their disease. They reported that the emergency room was the primary source of medical care sought when an acute pain crisis happens. These survey results highlight the many unmet needs for adults living with sickle cell disease.

Background

Sickle cell disease (SCD) is the most common severe and life-threatening genetic disease impacting approximately 100,000 children and adults in the United States (Piel et al. 2010). The implementation of newborn screening for the disease in all 50 states has significantly improved life expectancy by lowering the risk of early childhood morbidity and mortality with the use of prophylactic penicillin to prevent severe infection (DeBaun et al. 2019; Paulukonis et al. 2016; Payne et al. 2020). In recent years nearly all children born with SCD in the US survive to adulthood, however SCD has become a severe, life-threatening, chronic disease for most adults, with the majority of adults dying before the age of 50 (DeBaun et al, 2019; Maitra et al. 2016). The effects of SCD in adulthood include severe chronic and acute pain, organ damage at a young age, fatigue, neurocognitive effects, a high rate of sudden mortality, and diminished quality of life for many. Further, there is clear body of research finding under-treatment and lack of support for those with this disease that affects primarily Black, Latinx, and other people of color that describes systemic and institutional racism (Power-Hays and McGann 2020; Kanter et al. 2021). Lack of access to quality care for adults has been well-documented (Lee et al. 2019; Linton et al. 2020; Kanter et al. 2020). The community of those living with SCD has also been severely impacted by the COVID-19 pandemic, with very high rates of hospitalization and mortality and a more diminished capacity than healthy populations to do daily activities in public such as work and school (Lee et al. 2021; Singh et al. 2021).
The CDC-funded Sickle Cell Data Collection Program and several small registries are working to gather important data on the health impacts of those with SCD in eleven states, including California. These efforts have supported new policies in some states, increased funding, new research, and new treatments for the disease. At the same time, there are few efforts to gather data directly from those living with the disease about how SCD impacts them, their families, their professional and educational lives, and their day-to-day activities. This information is vital for policy makers, healthcare providers, support organizations and advocates, families, and others to attend to the needs of those living with SCD. We report here on a wide-reaching survey of people living with this disease.

**Methods**

Collaborators in California, including community-based SCD support organizations Sickle Cell Anemia Awareness San Francisco (SCAASF) and the Cayenne Wellness Center (Cayenne) as well as the public health surveillance team at Tracking California, sought to ask persons living with SCD and their families in that state and throughout the US their opinions, experiences, challenges, barriers, and successes in finding care, learning more about their disease, and living a full and well-supported life. The team developed and launched an online survey during the period April-October 2021, receiving 586 responses describing areas to improve access to health information, support services, and treatment in the US SCD population.

With approval from both the California State Committee for the Protection of Human Subjects and the Public Health Institute's Institutional Review Board as part of the Sickle Cell Data Collection Program, the three teams worked together to create the survey. Members of the team looked at information gaps in the literature and used their own professional assessment, drawing on conversations and questions from support group settings, educational symposiums, and other venues in which those with SCD share their experiences, challenges, and successes. When choosing the questions and format the team took into consideration the amount of time the survey would take respondents, the amount of information people might or might not be comfortable sharing, and the potential value of the findings to inform care and support.

SCAASF distributed the survey in April 2021, first sharing it with its constituents via social media. The survey was then shared widely, with responses from across the US and internationally. Cayenne’s team included the survey as part of its SCD Educational Symposium September 16–18, 2021. This annual event, normally held in person in Southern California, was held virtually due to COVID, and so attracted a large following, eliciting responses from other states and countries as well. The surveys were developed and implemented in Microsoft Forms, with results downloaded to Excel files for analysis. Surveys did not collect names, dates of birth, or other identifiers. Emails were requested for those respondents who wanted more information about the survey and/or to be in a drawing for a $50 retail gift card.

We asked respondents their age, sex, insurance status, access to hematologic and primary care, and connection to a support group along questions about health education sources, steps taken for pain...
crises, and greatest needs and information gaps experience. The focus in presented results is on US respondents, highlighting specific results from California relevant for examining support group access – there were not enough international respondents to offer meaningful interpretation. To assess their connection to a hematologist we asked, “Do you have a hematologist or other SCD expert whom you see for SCD-related health issues?” for primary care access we asked, “Do you have a primary care physician whom you see for non-SCD related health issues?” We included open-ended/free text to draw more information from respondents on their experiences seeking health care and managing their disease. The final questions and response options are in Appendix A.

In analyses, age groups were combined because of limited responses from the youngest and oldest groups: the 70 and older age group combined with the 51–69 age group, as the former group included fewer than 10 respondents, and the 0–18 age group was combined with the 19–34 group, as the former included only 12 respondents. The results were tested for statistical significance (Kruskal Wallace Test) where noted, however most results are descriptive. Cell sizes under 11 are suppressed, in keeping with California Committee for the Protection of Human Subjects requirements.

**Results**

There was a total of 586 responses, 551 of which were from persons with SCD living in the US. Table 1 shows the age and sex of respondents as well as their insurance and healthcare access responses. Nearly 2/3 of those responding to the survey were from US states other than California, 57% were female, and over 90% were between the ages of 19 and 50 years.

Table 1 Age, Sex, and Location for Persons Living with Sickle Cell Disease Surveyed, 2021

<table>
<thead>
<tr>
<th>Age</th>
<th>Total (Col %)</th>
<th>Female (Row %)</th>
<th>Male (Row %)</th>
<th>Declined to state (Row %)</th>
<th>Location (Row %)</th>
<th>California (Row %)</th>
<th>Other US (Row %)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-34 years</td>
<td>305 (55%)</td>
<td>167 (55%)</td>
<td>137 (45%)</td>
<td>*</td>
<td>81 (27%)</td>
<td>224 (73%)</td>
<td></td>
</tr>
<tr>
<td>35-50</td>
<td>210 (38%)</td>
<td>117 (56%)</td>
<td>93 (44%)</td>
<td>*</td>
<td>56 (27%)</td>
<td>154 (73%)</td>
<td></td>
</tr>
<tr>
<td>51 years or older</td>
<td>36 (7%)</td>
<td>28 (78%)</td>
<td>*</td>
<td>19 (53%)</td>
<td>17 (47%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>551 (57%)</td>
<td>312 (57%)</td>
<td>238 (43%)</td>
<td>*</td>
<td>156 (28%)</td>
<td>395 (72%)</td>
<td></td>
</tr>
</tbody>
</table>
Almost all respondents have some form of health insurance (95%), and most have a connection with a primary care provider and/or hematologist (79% and 78% respectively). Connection with a support group was low, only 1/3 of Californians (highlighted because both community-based organizations conducting the survey were California-based), and 16% of respondents from other states, with a combined average of 21% (Table 2).

Table 2 Age, Insurance Status, and Access to Care for Persons Living with Sickle Cell Disease Surveyed, 2021

<table>
<thead>
<tr>
<th>Have Insurance</th>
<th>Have Primary Care Physician</th>
<th>Have Hematologist</th>
<th>Belong to SCD Support Group</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>California (% of Total)</td>
<td>Other US (% of Total)</td>
<td></td>
</tr>
<tr>
<td>Age</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>0-34 years</td>
<td>296 (97%)</td>
<td>241 (79%)</td>
<td>224 (74%)</td>
</tr>
<tr>
<td>35-50</td>
<td>198 (94%)</td>
<td>166 (79%)</td>
<td>171 (81%)</td>
</tr>
<tr>
<td>51 years or</td>
<td>34 (94%)</td>
<td>31 (86%)</td>
<td>33 (92%)</td>
</tr>
<tr>
<td>older</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>526 (95%)</td>
<td>438 (79%)</td>
<td>428 (78%)</td>
</tr>
</tbody>
</table>

Respondents were asked where they received health education and other sickle cell disease-specific information (with a “check all that apply” format), and responses showed that such information comes to the respondents from a variety of resources. Among the 39 respondents who selected “Other” in response to this question, most noted specifics about sources included in the survey such as social media, while six said they get information from support groups and nine mentioned healthcare providers as information sources (Figure 1). Among those choosing “social media” from the selections, the majority described Facebook, Instagram, and Twitter as their primary sources.

A follow-up question to the inquiry about information sources asked what types of information respondents would like to have; 260 (47%) listed one or more areas of information gaps they sought to fill. These open responses were coded into categories by the authors. For types of information sought: 105 responded “health education about the disease and its treatments” was the most sought type of
information, 44 responded “support with coping with the effects of the disease”, and 28 responded “information on other kinds of support.” In addition to information, 32 respondents noted “needing to find access to knowledgeable and empathetic care providers.” Information about cost and/or insurance access was included by 29 respondents; 10 asked for information on alternative therapies. A smaller number of respondents asked for more information on aging, racism as it relates to the disease, COVID, genetic counseling, mental health care, pregnancy, sexuality and sex, new clinical trials, and nutrition. Selected quotes from respondents:

- “Better care access to mental health better trained and more qualified doctors who treat with empathy.”
- “I need some encouragement.”
- “What efforts are being made to humanize treatment of adults in emergency care?”
- “Health insurance cost are rising. Are there programs for middle class ppl to help save money?”
- “Holistic medicine, and sickle cell rights and the treatment of POC with SCD”
- “I need a place to receive hydration, pain management, stress mitigation techniques, nutrition support, information regarding herbal solutions and how they react with the prescribed medication i am taking. I need a Sickle Cell Comprehensive treatment center in <respondent’s locale> where help is available after 5.”
- “I would like information about the hospital or the best doctor to treat sickle cell.”

Another question asked respondents what steps they take when they experience a vaso-occlusive pain crisis, a frequent occurrence for most adults living with sickle cell disease; respondents were again able to select more than one option. Figure 2 shows that most respondents opt to go an emergency room for care.

Finally, respondents were asked to rate the helpfulness to their own lives of access to programs such as counseling, patient transition into adult care, improved healthcare, medical community compassion and attitude toward patients, and other. Improved healthcare received the greatest number of “most helpful” and the greatest number of “least helpful” rankings; this category along with provider compassion tied for the highest mean ranking (Table 3). There were significant differences among age groups in these ratings.

Table 3 Ranking of Needs by Persons Living with Sickle Cell Disease Surveyed, 2021
Respondents were also asked about anything related to sickle cell disease in an open question at the end of the survey. Responses ranged from gratitude for the survey or the settings (i.e., the Cayenne educational symposium) to comments on respondent’s current health status. Below are quotes that describe the needs of those with sickle cell disease.

- “I would also like to see a streamlined plan to help Sickle Cell patients when they are navigating from the outpatient center to the ED or when they arrive in the ED for care. Hospitals can do so much more to facilitate quick effective treatment by having staff that are experienced and prepared to access ports, that are not afraid to administer medication and that do not have prejudices against people of color or people who require pain management with opioid medication.”
- “I really appreciate the emphasis on mental health (at the symposium). It is an aspect that has been the most difficult to access, even though it is easily available for cancer patients. I would also like to see a peer counseling aspect addressed.”
- “I marked compassion somewhere in the middle, but it’s truly important. I often get discouraged from going to the doctors because of how they treat me”
- “Having a (sickle cell disease) Center in my city has made all the difference. It’s a place that’s safe and accepting and makes all the difference.”
- “ER is still too challenging, so my daughters still wait a long time before going. It seems inhumane to schedule pain relief pills for the future! No one with sickle cell disease should be given a handful of
pills and told to stretch them out, work it out, nonetheless, get a refill as scheduled. Not acceptable. Not when i continue to see young white girls post plastic surgery with over 90 pills at a time.”

- “I want to live.”

**Discussion**

Two sickle cell disease community-based organizations, SCAASF and Cayenne, worked together with Tracking California to design and implement a brief survey of persons living with sickle cell disease, and our analyses of results focusses on 551 responses received from US residents. The input received describes the lived experience of coping with sickle cell disease shines a light on the challenges and barriers faced in just receiving needed medical care for their disease and offers specific barriers that this community faces to better health and quality of life.

Respondents were drawn from the larger community of people living with sickle cell disease connected to either social media, the Cayenne educational symposium, support groups, or some combination of these channels. Understanding this context helps us contextualize the results – this is likely a group of individuals who have better support and connection to information than others in the sickle cell disease community. For example, the high proportion of respondents with a connection to a hematologist (78%) contradicts recent evidence that adults with sickle cell in two states have low rates of connection with such specialists, finding that 56% in California and 34% in Georgia had no encounters with a hematologist over a three-year period (Horiuchi et al. 2022).

There is very limited published research on the rate of connection and the benefits of attendance of support group meetings among those living with SCD (Telfair and Gardner, 2000). We expected that the connection with support groups among these respondents would be high, given that the survey was promoted through community-based organizations with support groups as key components of their work, but found that they were low. Because both organizations promoting the survey were based in California, we reviewed connection with support groups separately for that state but found even there than only 1/3 of those responding were connected to a support group, and overall, only 1 in 5 across the US are connected. This is an important finding; researchers and those leading support groups may want to do further research to determine the reasons for this low connection rate, and funding should be made available to make such groups more accessible within the identified needs.

Given that the survey was largely distributed via social media, it may be noted that a high level of reliance on social media for disease information by respondents may be a bias. However, the high level of interest and response rate for the survey suggests that social media is wide-reaching and may be an effective means of reaching the greatest number of people for health education, advocacy, and support. Advocates, support groups, and public health professionals may draw from the information needs identified in the survey to create materials, media, and formats to fill these gaps for those living with the disease.
The findings in response to vaso-occlusive crises reveal new information about the experiences and challenges faced by those living with this disease. Recent evidence on sudden mortality among adults with sickle cell disease shows that such pain crises should be treated as an emergency event (Johnston et al. 2020). Therefore, the high rate of respondents stating that they find a way to access emergency care is a positive finding. At the same time, many respondents report that the emergency room experience is not a supportive place for them, and that staff attitudes are detrimental to their ability to cope with the disease, and these comments echo published reports (Linton et al. 2020; Kanter et al. 2020). This puts those in pain crisis in a position where getting the best care is an emotional and social burden to solving a life-threatening health problem.

When respondents were asked to rank what is needed to achieve optimal health, their answers varied widely, including across age groups. It is possible to interpret these findings as the respondents saying “all of them” to the list of choices. The basic needs expressed in the narrative, free text portions of the survey are significant and at times heart-rending.

While this survey had a high number of respondents, a limitation is a relatively narrow outreach spectrum focused on social media and one educational event. Future data collection may broaden this to more educational gatherings, paper surveys administered in the emergency room or other clinical setting, or other, different points of access.

Policy implications of the findings of this survey of needs of those with sickle cell disease in the US include supporting increased funding for access to knowledgeable, quality care, improving the emergency room experience for those with sickle cell disease, creating diverse conduits for health education, advocacy, and other types of information for this community, and developing increased opportunities for those with the disease to access support groups.

By conducting this survey, we found information essential to the provision of support and care for those with sickle cell disease. Key messages from the data show that those living with this disease have a need for more information about the disease and treatment; most respondents reported working with a primary care or hematologist; and the majority are not connected to a support group.

**Declarations**

**Author contributions**

All authors contributed to the survey conception and design. Oversight of the dissemination of the survey and data collection was performed by Carolyn Rowley, NeDina Brocks, Antwan Capla, and Kimble Torres. Niani Coker performed data analysis of the survey data, Daniel Madrigal contributed health education and policy insights to the work. The first draft of the manuscript was written by Susan Paulukonis and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.
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Compliance with Ethics Guidelines

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All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (California Committee for the Protection of Human Subjects and the Public Health Institute IRB) and with the Helsinki Declaration of 1975, as revised in 2000 (5). Informed consent was obtained from all those completing the survey.

Data Availability

The datasets (with all potential identifiers removed) generated during and/or analyzed during the current study are available from the corresponding author on reasonable request.

References


Figures

![Figure 1](image)

**Figure 1**

Health Education and Sickle Cell Disease Information Sources for Persons Living with Sickle Cell Disease Surveyed, 2021
Figure 2

Most Common Responses to a Vaso-Occlusive Pain Crisis Among Persons Living with Sickle Cell Disease Surveyed, 2021

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- AppendixA.docx