1. Introduction

Sickle cell disease (SCD) is a structural hemoglobinopathy of the β-globin gene and follows an autosomal recessive inheritance pattern. It results in abnormal red blood cell morphology with an increased tendency to occlude microvasculature and subsequently impair oxygen delivery. The cornerstones of SCD treatment include prophylactic antibiotics and immunization to prevent infections, hydroxyurea to increase fetal hemoglobin and improve oxygen delivery, and blood transfusions.

Globally, SCD is estimated to have an incidence of over 400,000 births per year, predominantly affecting regions of Africa and India. The Lancet Commission on Reframing Noncommunicable Diseases and Injuries for the Poorest Billion recommends universal newborn sickle cell screening followed by standard prophylaxis against bacterial infections and malaria. In high-income countries, over 90% of babies born with SCD survive into adulthood; conversely, in low-income countries, which lack infrastructure for screening, confirmatory diagnostics and management, up to 80% of those born with SCD are undiagnosed and less than half of them survive beyond 5 years of age.
1.1. Sickle cell disease in the Indigenous Tharu population

Sickle cell trait (SCT), consisting of a genotype of one normal and one abnormal sickle cell mutation, confers a degree of protection against malaria. Ethnic groups in Nepal that originate from malaria-endemic lowlands, such as the Indigenous Tharu population of the Terai region of Southern Nepal, have a higher prevalence of disease. In 2017, Marchand et al. reported on a 9.3% SCT prevalence amongst Tharu individuals residing in the Dang district of the western Terai region. The Terai (lowland) region of Nepal has the largest percentage of individuals (45.4%) in the poorest socioeconomic demographic. The economy of this low-resource region is largely driven by agriculture. The Tharu ethnic group consists of more than a million people who live primarily in the Terai, a narrow strip of land running along the Nepal-India border. Of the total SCD cases in Nepal, 58.3% are in the Tharu population, who make up only 6.6% of Nepal’s total population. The high prevalence of SCD among the Tharu population has led to the stigmatization of this disease as a “Tharu disease.”

Previous research has demonstrated that Tharu individuals are frustrated with their healthcare infrastructure and have difficulties navigating the local SCD screening process; misconceptions regarding the pathophysiology of SCD were also evident. Given the lack of research in this geographic area and the limited capacity of healthcare services for Tharu individuals with SCD, the purpose of this study was to conduct a needs assessment using focus groups to explore Tharu perceptions on SCD-specific care. Understanding Tharu perspectives and experiences is an important first step to improve the delivery of SCD-related care for this marginalized population.

2. Methods

2.1. Study design

This study used a qualitative approach, specifically focus group discussions (FGDs), to develop an understanding of the Tharu community’s perspectives on health-care delivery in the Dang District of Nepal. Focus group interviews were conducted with subsequent analysis based on grounded theory. Ethical approval of the study was authorized by our institution’s Clinical Research Ethics Board (H16-00187). Oral and written consent were obtained from all participants prior to enrolment in the study.

2.2. Data collection procedures

Inclusion criteria included self-reported Tharu ethnicity and awareness of SCD including its local screening process. In the Dang District where this study was conducted, community stakeholders and local organizations described requiring visits to five separate locations for a formal diagnosis. Exclusion criteria included inability to physically attend the interviews and age under seven. Snowball sampling was performed with community members recruited through randomized telephone invitations and word-of-mouth in May–June 2017. Recruitment was conducted by a local non-profit Nepalese organization, “Creating Possibilities”, until thematic saturation was achieved.

Questions related to SCD centred on general understanding of SCD and aspects of SCD care that participants believed could be improved. Examples of questions included: “Why did you decide to get screened?”, “What did you understand a positive screening test to mean?” and “Since the diagnosis, do you think you have been properly informed of the implications of this disease?”

Participants were interviewed in a community center in their language of choice: English, Hindi, Nepali, or the local Tharu dialect. FGDs were audio recorded and transcribed to Nepali, with field notes made during the process for clarification purposes. Translators from “Creating Possibilities” assisted in delivery of questions. Transcribed interviews were then translated to English by an unaffiliated translator to mitigate any potential bias.

2.3. Analysis

For the FGD data, a constant comparison analysis was used. This process was based on grounded theory and is divided into three main phases. The first phase, open coding, refers to the researcher’s initial familiarization with their data and “chunking” of data into smaller units. The second phase, axial coding, involves further categorizing these chunks of data into nodes. The final phase, selective coding, involves the development of themes from determined nodes. The themes are meant to be an overarching expression of the complete data set and are particularly useful in studies where multiple FGDs are involved. This analysis allows researchers to easily assess their approach of across-group saturation.

Inductive thematic analysis, as defined by Braun and Clarke, was also used. When studying under-researched areas using qualitative data analysis, it can be helpful to use a rich description of the complete data set compared to a more focused view of specific themes. Thematic results were thus data-driven as opposed to a theoretical approach whereby the researcher’s data coding complies to a specific area of interest.

The data was analyzed and the coding structure was developed using NVivo 11™ (QRS International). The first phase of constant comparative data-analysis was performed independently by three researchers (AI, NB, KL). The open coding results were then refined with axial coding to develop nodes. Selective coding of participant statements was then performed independently by re-analyzing all focus group interviews. Each of the independent re-analyses were then compared to determine overarching themes and sub-themes.

3. Results

In total, 133 participants were enrolled in 22 FGDs. Participants were primarily female (91.7%, n = 122) with 13.5% of participants (n = 18) between the ages of 7 and 15. Participant demographics are summarized in Table 1.

3.1. Themes identified in the SCD FGDs

The three major themes identified in the FGD interviews on SCD were: social implications of SCD, importance of SCD education, and positive perception of SCD interventions. The three themes and their sub-themes are displayed in Fig. 1.

Table 1

<table>
<thead>
<tr>
<th>Characteristics of community members participating in focus group discussions centred on sickle cell disease.</th>
<th>Sickle Cell Disease Focus Group Discussions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total participants</td>
<td>133</td>
</tr>
<tr>
<td>Total focus group discussions</td>
<td>22</td>
</tr>
<tr>
<td>Male: Female ratio</td>
<td>11:122</td>
</tr>
<tr>
<td># children (age 7–15)</td>
<td>18</td>
</tr>
<tr>
<td>Screened negative</td>
<td>59</td>
</tr>
<tr>
<td>Screened positive</td>
<td>51</td>
</tr>
<tr>
<td>· Screened positive, diagnosed homozygous sickle cell disease</td>
<td>6</td>
</tr>
<tr>
<td>· Screened positive, diagnosed sickle cell trait</td>
<td>44</td>
</tr>
<tr>
<td>· Screened positive, diagnosed false positive</td>
<td>1</td>
</tr>
<tr>
<td>Other</td>
<td>23</td>
</tr>
</tbody>
</table>

*Those who did not remember their test results, those who had family members who were diagnosed with sickle cell trait or disease, or members of the community who felt that they could contribute to the conversation.*
3.1.1. Social implications of SCD

Participants showed inconsistency in their understanding of SCD and thus variability in their perspectives of SCD’s social impact on their lives. As a result, participants were divided with respect to the amount of stigma they faced. Those who felt stigmatized often associated SCD with their Tharu identity.

“I have heard that due to eating pork meat this disease occurs and many of us Tharus have stopped eating pork. We are worried why only Tharus get this disease while other communities don’t.”

However, several participants described that there was less stigma in the community towards those living with SCD than those living with other diseases. This was due to some community members’ understanding that SCD is a non-communicable disease. Communicable diseases, such as HIV or TB, were seen as preventable and therefore were cited as conditions that faced more stigma. Keeping with this, many community members did not discuss social implications of SCD on their relationships, such as their ability to get married or employed, unlike with communicable diseases. \(^{22}\)

“So far we haven’t seen or heard about people stigmatizing those who have the disease. The positive behaviour towards them despite the disease has not been changed. Everyone treats them well.”

In addition, this participant discrepancy in SCD knowledge seemed to play a role in how men and women viewed the relevance of SCD on their community; for example, men were generally less interested in SCD testing. Several women discussed that many men in the community spend a significant amount of time out of the country for work or performing physically intensive labor. For these reasons, it was suggested that the men do not prioritize health screening. One woman highlighted this phenomenon by saying:

“...majority of [the men] work outside and are illiterate, so it’s difficult to make them understand about the advantages of screening. Most of them think that we have spent most of our life without screening, why do it now? I think they need lot of counselling about this disease.”

3.1.2. Importance of SCD education

Gaps in the understanding of SCD etiology seemed to be a main contributor to the spectrum of attitudes toward SCD as described above.
Participants frequently acknowledged their lack of understanding of SCD and their interest in learning more about it. Questions about the pathogenic mechanisms and epidemiology of SCD included:

“How does sickle cell occur and transfer? What happens when it is trait? We would like to know more.”

“We would like to know more about sickle cell. We came to know some information about it but [other] people from community still don’t know much. It would be better if [the whole community] could also be informed about it. It’s usually said it affects only Tharu people. I would like to know whether it affected people from other communities.”

Beyond SCD etiology and pathogenesis, there was also a lack of awareness regarding the screening and diagnostic process. In the Dang district, the full SCD testing pathway requires visits to five different locations. This understandably causes confusion for community members as many were unclear of the distinction between screening and diagnostic testing. One participant concluded:

“I don’t think we know the difference between positive screening and positive diagnosis. We know about the presence or absence of disease.”

To help face these SCD knowledge gaps, discussion eventually focused on how community members would like to be educated. Community awareness programs about SCD were continually brought up as a potential method for improving understanding.

“Awareness program should be conducted in the society. Most of the people of Tharu community are uneducated. [They have] to go out for work and have less access to media like TV [or] radio, so they have to provide information through awareness programs.”

Some participants that had been exposed to educational interventions in the past expressed a desire for reinforcement to consolidate their learning.

“I don’t know much about this disease. Even though I go to counselling I have forgotten most of the things.”

3.1.3. Positive perception of SCD interventions

When asked about SCD-specific interventions implemented to date, many participants in the FGDs described their positive experiences. In particular, the screening procedure was described as beneficial and those interviewed were pleased to get the chance to learn more about their health.

“If we don’t do the screening, we won’t be able to find out about our health status. So that is why [it is] important to continue the screening process.”

Furthermore, participants who were able to receive education expressed relief after being informed about available treatments and prognosis for those with SCD:

“Initially we assume that we would die once we had this disease. Then after we were informed, we knew our assumption was wrong. Now, we are not scared anymore despite [having sickle cell] trait.”

Some of those who took part in previous education interventions facilitated by local NGO workers had good retention of knowledge about SCD. A recurrent topic of conversation amongst individuals was the lack of long-term counselling services.

“They went for screening in Chainpur Health Post and nobody has provided counselling. However, in [the programs implemented] it was discussed clear about this disease and they have understood it properly.”

However, as noted previously, participants who had been exposed to interventions in the past often requested continued maintenance of intervention programs to consolidate learning.

4. Discussion

While previous studies have focused on SCD prevalence in Nepal, this study provides a unique voice to the Indigenous Tharu people, an ethnic group with a high prevalence of sickle cell trait that has been historically marginalized. Major themes identified in the FGDs were: social implications of SCD, importance of SCD education, and the positive perception of SCD interventions. Understanding the perspectives of underserved communities is critical to address their needs, and implementing tailored, patient-centred education, screening, diagnostic and therapeutic programs. Overall, our study demonstrates that while the Tharu community has identified numerous barriers to care, members have remained resilient and optimistic regarding the implications of a SCD diagnosis.

These findings should be examined within the context of Nepal. SCD was only recently identified within the country, and the first case formally reported in 2003. The Nepali government has since agreed to subsidize SCD treatment under the Disadvantaged Citizens Fund. Nonetheless, financial support for SCD treatment can only be accessed once a diagnosis is made, and having healthcare infrastructure to support this process is critical. Community stakeholders reported that the bureaucratic process to obtain a formal diagnosis of SCD from the National Ministry of Health, including visits to five locations, was a significant barrier for many in the Dang district. The route to a diagnosis in Dang includes screening at a local health post in Gangaparsup, diagnostic testing in Nepalgunj, obtaining a letter stating the positive diagnosis from the Village Development Council in Ghadawa, a certificate of SCD from the Ghorahi District Public Health office, and treatment at Ghorahi district hospital if necessary. Streamlining this diagnostic and therapeutic process could address some of the concerns community members face. For example, community outreach through mobile health units has been used in the United States to make SCD diagnosis more accessible; this strategy may be one that could be examined and applied for use in Nepal.

We found that amongst those with a better understanding of the inheritance of SCD, the role of stigma and its subsequent social implications was not as significant as expected. This is reflected by literature from Nepal that found associations of shame with communicable diseases, such as tuberculosis. Therefore, stigma predominantly occurred when community members had poor understanding of the inheritance of SCD and did not understand it was non-communicable. These findings underscore the importance of educational interventions in mitigating myths and disease-related stigma in low literacy communities. Respondents also mentioned the notion that SCD only affects Tharu individuals, consistent with prior literature. Finally, one participant postulated that illiteracy seemed to be related to screening reluctance, also demonstrated in a study from India. The finding of gender disparity in SCD screening could reflect the common practice of men working outside the village, overall lower educational attainment, or may be explained by the belief that SCD is only passed through the mother. However, the validity of this finding is uncertain, given the female predominance of participants in our FGDs.

Local community members strongly felt that there was an opportunity for empowerment by enhancing health education. The benefits of community-based participatory research when addressing education and barriers to health have been well documented. Using this approach, studies can integrate community responses into ongoing health education in the region. Suggestions for local improvement included disseminating regional health education in accordance with forms to which community members are most receptive. Examples including narrative performances and music were discussed. Such performances have been used in Tharu communities for SCD education in the past. Due to a lack of information retention, community members also requested a continued education model, with modules and lessons held periodically to solidify their knowledge and ensure a deeper understanding of the diagnostic process. Further collaboration with local...
stakeholders would be critical in ensuring continued health education.

A study published by Mathie et al. emphasized the role of Asset Based Community Development (ABCD), otherwise known as strength-based approaches, in community. Specifically, in rural communities, members have been able to identify existing local resources and mobilize these resources in innovative ways. ABCD rests on the principle that recognition of strengths and assets is more likely to inspire positive action for change than exclusive focus on needs and problems. Two notable strengths were identified in our community: their dedication to advocacy for their own health needs through promotion of better health education and SCD awareness, and their optimism regarding the implications of SCD diagnosis and prevalence within their community. A community’s resilience factors, namely their assets and strengths, should be strongly considered when incorporating health programs and analysing the outcomes.

There are several limitations to this study. For one, all participants were sampled from a small geographical area. While this allowed us to obtain the viewpoint of participants from the Indigenous Tharu population in the Dang district, the results may not be applicable to other regions of Nepal. In addition, the higher proportion of female to male participants may have resulted in a bias in thematic development. Furthermore, community members who participated may have avoided being overly critical in interviews as the researchers were affiliated with a local organization that provides social support in the region. Low literacy rates in the community were also a barrier to using data collection forms and limited our ability to gather comprehensive demographic information on participants.

4.1. Future research directions

Future research may replicate this study with other marginalized groups in areas around the world with high SCD prevalence, or with other heritable diseases found in Nepal, to further understand the perspectives of similar communities. Examining a higher-income participant population may provide information on which healthcare disparities can be attributed to low-income settings. Studies examining the viewpoint of health-care providers, instead of community members, may also offer a different perspective that would be valuable.

5. Conclusion

We found several themes highlighting the perspectives and knowledge of individuals in rural Nepal regarding SCD. The issues that arose, including low-level understanding of the disease and bureaucratic challenges of the diagnostic process, are areas that need to be addressed for the sustainability of sickle cell care and improvements in health outcomes. With the high childhood mortality in untreated SCD individuals, the importance of understanding a local community perspective of needs would aid in the streamlining of SCD screening, diagnosis, and treatment. Overall, as more work is published in this area, we hope to provide strong evidence of the need for government-initiated SCD programs for those at high-risk across Nepal.

Sources of funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Declarations of interest

None.

Acknowledgements

The authors thank Dinesh Sapkota and his colleagues at “Creating Possibilities” for their contributions, Her International for their support, the interview participants that shared their experiences, and to other members of the research team (V. Sharma, J. Yeo).

References