

Empowering patients with sickle cell anemia and their families through innovative educational methods

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Abstract

Sickle Cell Disease (SCD) is a group of inherited blood disorders caused by a mutation in the beta subunit of hemoglobin (HbS). SCD is also known as Sickle Cell Anemia (SCA). There are approximately 5000 Canadians living with SCA including children. Pediatric SCA patient education can: improve knowledge, decrease hospitalization, improve medication possession ratio, lead to better SCA related functioning, and lower pain impact. Innovative educational materials were developed to improve knowledge and self-efficacy regarding illness management of patients and parents/guardians. Patients (n=5; aged 8 – 18) with SCA and parents (n=5) of patients (aged 0 – 18) were recruited via flyers sent directly to patients and distributed through partner patient organization Sickle Cell Awareness Network of Saskatchewan. Patient and parent focus groups were held separately over Zoom to receive feedback for the video. An additional interview was held for a participant that required translation of the video. Audio recordings were transcribed using Zoom and Otter.ai. Coding of transcripts was facilitated by NVivo (QSR International Pty Ltd, 2022, release 1.6.2). Thematic analysis centred around SCA management concepts relevant to the research aims. Important themes that emerged included: ‘Age Appropriateness’, ‘Empowerment’, ‘Knowledge Gaps’, ‘Linguistic Accessibility’, ‘Medication Adherence’, ‘Strength in Community’ and ‘Transition to Adult Care’. The video was well received, and “brought peace of mind”. Patient feedback was incorporated into the final version of the educational materials.

Introduction

Sickle Cell Disease (SCD) is a group of inherited blood disorders caused by a mutation in the beta subunit of hemoglobin (HbS). SCD will hereafter be referred to as Sickle Cell Anemia (SCA) as this is the term our patients and their families prefer. There are approximately 5000 Canadians living with SCA (1). The mutated B subunit causes polymerization of hemoglobin leading to sickling of red blood cells. Sickle cells are prone to destruction and clog blood vessels resulting in ischemia (2, 3). SCA is associated with anemia and vaso-occlusive pain crises (VOCs) that can lead to complications involving the bones, lungs, abdomen, brain, eyes and reproductive system (4). Life-threatening events are also common and include acute chest syndrome (ACS), ischemic stroke and infection (4).

Pediatric SCA patient education can: improve knowledge, decrease hospitalization, improve medication possession ratio, lead to better SCA related functioning, and lower pain impact (5-10). However, there is a lack of information with features desirable to adolescents on the internet written at an appropriate reading level (11). In particular, there is a lack of information about management and prevention of emergencies like ACS (11). There is a need to develop age-appropriate educational materials for patients with SCA and their families.

Furthermore, educational materials are an important component of transition to adult care and should be provided to patients starting from a young age. Acute care visits and rehospitalizations are frequent in those

with SCA 18 -30 years old (10). Patients are vulnerable at this time and equipping youth with knowledge and self-efficacy of illness management is critical.

Methods

An innovative educational cartoon video targeted towards youth with SCA was developed to improve knowledge and self-efficacy regarding illness management of patients and parents/guardians. Following a literature review, the medical student developed a script for the video. Due to the lack of information available on the internet regarding sickle cell emergencies, the video focused on these (11). The script was then reviewed by the pediatric hematology team. Additionally, the script was reviewed by a patient and parent advisor team. The video was created using Powtoon video creator and in collaboration with the Sickle Cell Awareness Network of Saskatchewan (SCAN SK). Voice actors included a patient with SCA as representation was thought to be paramount. Ethics approval was attained through the University of Saskatchewan Behavioural Research Ethics Board (ID: 2679). All proceedings involving patients were carried out according to University research ethics board policies and procedures. Patients (n=5; aged 8 – 18) with SCA and parents (n=5) of patients (aged 0 – 18) were recruited via flyers emailed and mailed to patients and distributed through SCAN SK.

Information about age, gender, ethnicity, and race was gathered at the video interviews. All participants identified as Black, and/or of African descent. Specificities of ethnic identity will be kept confidential to protect participants' identities. Three of the patient participants were in the 16 – 18 year old age range. The remaining two were between 8 - 10 years old. All identified as male. Most of the parent participants identified as female. One parent participant identified as male.

A patient focus group and parent focus group were held separately over Zoom to receive feedback on the video. An additional interview was held over Zoom for a participant that required video translation. Student researcher Riley Plett facilitated all Zoom meetings. Subtitles were provided on the video by YouTube auto-translate for the participant requiring translation. The parent of the participant requiring translation also aided in translation of the focus group questions.

Audio recordings were transcribed using Zoom and Otter.ai. Coding of transcripts was facilitated by NVivo (QSR International Pty Ltd, 2022 release 1.6.2). Thematic analysis centred around knowledge and self-efficacy concepts relevant to the research question. Feedback from the focus groups was incorporated to refine the cartoon video and create a summary infographic requested by adolescent participants. The summary infographic was created using Canva and pulled information from the video.

Results

The video was well received by both parents and patients. It was thought to be "educational for all ages" and "brought piece of mind". Participants commented on the video's simplicity and tone. "I'm very sure my son would have enjoyed it. Maybe he felt it was going to be maybe traumatic... But there's nothing like that about the video."

Important themes that emerged from the analysis included: 'Age Appropriateness', 'Empowerment', 'Knowledge Gaps', 'Linguistic Accessibility', 'Medication Adherence', 'Strength in Community' and 'Transition to Adult Care'.

Age Appropriateness

The video's target audience was envisioned to be teens and young adults. The video was appropriate for adolescents according to parent participants. There were many requests that the video be made available to their teens. One parent (Parent participant 5) stated "It uses terms that younger people can understand... and it also gave me ideas in terms of how to communicate with younger people."

The young adult participants (16 – 18 years) had a different opinion on age-appropriateness. They thought the video would be better suited to younger audiences and that they would prefer a different media, such as an infographic.

"I believe that video is... really well done just for children. I think, grasps the children's attention. I also don't think it's too complex for the children to understand. I do think an infographic would be like, more beneficial to teenagers." - Patient Participant 1

With the teenagers, less was more. Patient Participant 3 stated that the infographic should have "all the important details summarized so there isn't too much to read".

Younger patient participants (aged 8 - 10) preferred the cartoon video format. References that adolescent participants found too juvenile were favorites for the younger ones. Patient Participant 4, our youngest participant stated "I like when they said Tik Tok is corny".

Empowerment

When asked if learning more about SCA made them feel better about managing their illness, patient participants agreed.

Parent participants were able to provide context to these responses.

"You know, [the video] lets them understand that there's something that they can do about the sickle cell to make it easier for them... I know that this information is going to help them. I can talk to them, but they need to have this information themselves. So I really appreciate that." - Parent Participant 1.

Talking about managing SCA as a youth can be difficult. Parent participant 3 unpacks this: "For young adults, a lot of times what I've noticed with my son, he's very kind of reserved when it comes to talking about sickle cell. And that's why I think one of the reasons participating in these things is a little bit tough on him. He does it but not too often." Encouragingly, the child of this participant was a leader in the patient focus group discussion. The meaningful engagement of this "reserved" patient embodied the empowerment created by connecting and learning about SCA with other youth.

Parent participant 1 spoke to the use of magma as imagery and the comfort this metaphor provided:

"The usage of the magma. It gives you an understanding better of what can happen, and the thought that you have some control over it, that it doesn't have to erupt, it doesn't have to get to that level. So that makes me feel good."

Knowledge Gaps

The video was able to provide learning for patient and parent participants. Our youngest patient participant (4) said, "I learned that sickle cells can't really bring oxygen; other blood cells can." Another young participant (5) stated they "learned about the function of the sickle cells".

Older teenage participants specifically commented on learning about why TCD ultrasound and MRI screening is important. "I did all these tests, but I never really knew why I was doing these tests. So, I felt like the video adequately explained the reasoning behind the tests." - Patient Participant 1.

For Parent Participant 2 it was helpful to hear familiar information in a new way: "You know, like today, just listening to that video... it was a different perspective, it was actually a revelation. I'm like, oh, you know, I've done so much research... but here I am, and still learning."

Although participants were happy with the educational content of the video there were suggestions for future content. One parent suggested stressing the potential for sickle cell emergencies in other areas of the body such as in the eyes and the importance of screening eye appointments. Another suggestion was to include information on what to do to receive help when experiencing a SCA emergency. Another participant inquired about whether it is safe to take hydroxyurea in pregnancy.

Linguistic Accessibility

The importance of having educational materials available in many languages arose during the focus groups. A blatant example of this is that an additional separate interview was required for a participant that required

translation for the video. Since many patients with SCA are newcomers to Canada, accessibility of the educational materials in multiple languages is important. The participant that required translation found it adequate to have the audio in English with subtitles on the video. YouTube has an automatic translation function. The parent and patient stated this worked well for understanding. They also confirmed the speed was okay for listening in English and reading subtitles in their language and that this was good practice for their child to understand SCA in both English and in their first language.

Medication Adherence

Stressing the importance of medication adherence in the video, especially for hydroxyurea came up in both the patient and parent focus groups. Parent Participant 1 stated, "We've tried almost everything. When we go to the clinic, they spoke with him. But he's still not getting it. So I don't know what else to do."

Parent participant 4 suggested that maybe adding into the video the importance of routine in medication adherence could be useful. They shared: "I always make sure after supper, I'll prepare a plate where I'll put his tablets. So, it's a routine. And because of that, it's easier to encourage him, because it's not like I'm forcing it down on him."

Patient participants also stressed the importance of encouraging youth to take their medication. This is especially important as youths take charge of their SCA management, as patient participant 3 puts it, "the older you get, the more freedom you have. But it doesn't mean you shouldn't take your medicine just because you're older". Patient Participant 2 suggested it "could be interesting to put on an infographic... how hydroxyurea, how that's useful and what your life would look like, if you were taking your medicine every day versus like, if you kind of just never took it."

Strength in Community

The importance of community was demonstrated through connections made at the focus groups themselves. Early in the parent focus group, participants began asking about connecting with other families. At this time some participants were connected to SCAN SK. Parent Participant 2 illustrated the importance of SCA families coming together:

"And especially, most of us when we come to Canada, and, you know, we come from places where family support system is very huge. And when we can hear the support system is not as huge as it was back home with our extended families, you know... So that interaction of different parents coming together different kids, you know... we need to be able to create that environment."

In the patient focus group, the importance of knowing you aren't alone was emphasized in Patient Participant 2's comment,

"Yeah, oh, maybe, um, one thing that like maybe on an infographic because they kind of always have like stats, maybe if there's like a stat of like, how many other kids also deal with sickle cell because when I was younger, I always thought that it was like, only me until I met [another patient with SCA]. And then after I met [them], like, sometimes I'd go to the doctor, and they would say that there's more people now... And I really never thought like that many people had it. So maybe like, that's like that would be kind of encouraging to older teenagers too."

Talking about SCA was found to be important, not only within SCA communities but with classmates and teachers at school. It was suggested that the cartoon video might even be useful to be shared in schools. Importantly this would also be "an opportunity to be able to put out material out there that will ease our kids." Patient participant 2 elaborates on the importance of this.

"When I was in elementary school, they gave me this book... to help me tell my classmates that I had sickle cell and there's like, certain things that they can do to help me if I'm in pain and stuff. And I found at the time I really didn't want to tell anybody but I found that it helped me a lot. So if there is like kids in elementary school who are scared to tell them, like, tell their classmates... maybe that video can help them explain what's wrong with them and stuff."

Our youngest participant stated he would like to share the video with his friends at school.

Transition to Adult Care

A common thread woven throughout the conversation was transition to adult care. Dynamics of family relationship were explored in this transition. The parent participants all stated that they were quite involved in their child's SCA management, providing that constant support at all ages. Parent Participant 3 reflected on providing support for hydroxyurea adherence, "I know, eventually he'll get it and just do it on his own. But until then I still want to be that little voice in his ear. Encouragement." Patient Participant 1 reflects on their perspective as a youth,

"I found as I was transitioning throughout my teenage years I kind of just started getting to know my body more I started to know like, what the limits were, like how hard I could push myself. And as I knew my body, people, like friends and family around me, just trusted me. So I feel like it is one of those things that does just come with experience as well."

It also became apparent that the issue of seeking emergent care was also an important one in the community. As in any emergency, quick assessment and treatment is crucial. Awareness is increasing in sickle cell emergent care, especially in pediatric departments. Participant 2 continues,

"Some doctors pretty like really know what's happening. 'We're getting your medication like all loaded up', but before it was like they had to call an on call doctor. And that would take a while just for me to get medication. So kind of slowed down the process and you're sitting in pain for a little longer, but it's gotten better now."

However, parent participants expressed that work still needs to be done in addressing sickle cell care in an adult setting, especially as youth transition to adult care. Parent Participant 3 recalls, "We had a crisis and we ended up with the adults. There was no information. They treated it, like you had a cold or you know, he was in a minor pain. And then I had to, you know, step out, got up and voiced my opinion, and eventually a doctor came and, you know, offered us the care that he deserved. They are not ready yet to deal with [them] coming in as young adults. And information needs to be there, ASAP."

For pediatric sickle cell patients, emergency cards have proved useful in the past. Patient Participant 2 recalls,

"They gave us a little card that said that, like we have sickle cell and like, like the stuff that we can do in like the first like 20 minutes that they get there to help relieve the pain. And I think that was pretty helpful. But now that I'm older, it really doesn't help. It's like for like the pediatrics."

Also, the young adults of the group highlighted the importance of recognizing their role as their own illness experts. They communicated that the rate limiting step to receiving treatment is provider awareness of emergent management for sickle cell crises, especially regarding medications.

"I also feel like I can pretty much tell a doctor, what I'm feeling like how I'm feeling and probably what medications I would need. Like, because when I go in there, I pretty much know what's happening... and like, see if they could get it approved faster, because I know not like every nurse can distribute medication like that. But stuff like that would help." - Patient Participant 2

Discussion

There is a need for age-appropriate educational resources to be available for patients with SCA and their families. There is also need for resources to ensure a smooth transition to adult care, especially since this is a medically vulnerable time for youth with SCA. Our cartoon video and teen infographic have the potential to assist in this transition and is meant to be shared with patients starting at a young age.

The major themes that arose during the focus groups highlighted important pieces of educational resource refinement and how these educational materials need to be supported by SCA health care services.

The educational materials need to be age appropriate. Breakey et al. found that there is a lack of information available on the internet for youth with SCA that is age-appropriate (11). This was echoed by our teenage participants who requested concise infographics. In the age of social media, it is important to consider the most age-appropriate media for educational material delivery. In clinic, it would be easy to show the cartoon video to younger participants as there is a growing abundance of tech available such as iPads in this setting. Teens are looking to the internet, doing independent searches for their information (11). Therefore, having infographics readily accessible on multiple social media apps and webpages would be more useful. Notably, the teens acknowledged some important information to include would be statistics about prevalence of SCA and information on how medication adherence can change their quality of life.

The materials need to be linguistically accessible. Given that many families of kids with SCA are newcomers to Canada, it is imperative that educational materials are available in a patient's own language. YouTube auto-translator was found to be acceptable to our participant but this might not accommodate every patient's needs. Involving a translator frequently in delivery of educational materials would most appropriately address accessibility.

Education requires family and community support. Medication adherence arose as a whole family concern. The developed educational materials could be used as a starting point for patient/family/provider brainstorming about how to take medication consistently. Strategies for medication adherence were incorporated into the final version of the video to prompt discussion.

Support does not stop within the family, as demonstrated in a teen's comment about needing to know that they are not the only kid with SCA. Statistics were included on the infographic about prevalence of SCA. This underlines the importance of educational materials addressing not only knowledge and management pieces but also questions youth have about SCA community and their place in it.

A major theme that arose that will require ongoing collaboration is SCA emergency department care during transition to adult care. Better SCA health outcomes are not only an issue of patient/family education but rely on health care provider education as well. Transition to adult emergent care was a particular concern. Access to timely and quality emergency care and differences between pediatric and adult populations is becoming recognized as a concern in other provinces as well. In Ontario, one study found that adults are waiting much longer than their pediatric counterparts, despite visiting the ED 2.5 times more frequently (12). This highlights the importance of collaboration for SCA emergency management. For patients and their families, future educational materials could implement components of how to navigate the health care system as an adult, especially when seeking emergent care. For providers there are many resources available for emergent SCA management including a point-of-care emergency department online tool developed by the American College of Emergency Physicians (<https://www.acep.org/sickle-cell/>).

There are limitations to this study. It should be noted that all patient participants were male and the majority of parent participants were female. There may be gender differences in how the educational materials are received and what participants value as important in educational materials. This study was also performed within Saskatchewan and was composed entirely of Saskatchewan participants. Therefore, it is not guaranteed that our findings are generalizable to other centres.

Ultimately, our novel education materials were found to be age-appropriate and empowering, addressing knowledge gaps across all ages. Reviewing the materials with participants highlighted that patient and family education does not take place in isolation. Furthermore, it was recognized for patient educational materials to be successful, they need to be complimented by ongoing improvements to collaboration between patients, their families and providers, especially in emergent contexts.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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