Initiation of Sickle Cell Education in Pediatric Hematology Annual Visits

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Background and Objectives

Background: Sickle cell disease (SCD) is an autosomal recessive blood disorder affecting approximately 100,000 Americans, making it the most common inherited blood disorder in the United States.[1] In 2010, SCD was estimated to be responsible for 113,000 hospitalizations and $488 million dollars in hospitalization costs annually.[2] Advancements have allowed nearly 95% of SCD patients to reach 18 years of age.[3] Effective education is necessary for maintaining long-term health of SCD patients, reducing hospitalizations and their associated costs, and aiding in the transition to adult care.[4] At the University of Maryland Children's Hospital, physicians, nurse practitioners, nurses, and child-life specialists, and have implemented an annual education curriculum consisting of age-appropriate information packets and activities for teaching patients and their families about SCD. Topics covered include general understanding of the disease, the importance of the patients' medications, and when to seek medical care.

Objectives: In our study, we aim to assess the efficacy of a multidisciplinary patient education curriculum implemented at the University of Maryland Children's Hospital for pediatric sickle cell patients and their families.

Methods

A total of 25 patients between the ages of 0-22 years of age with a diagnosis of SCD, including variations, were identified. Patients and their guardians provided assent or consent to participate in the study. Prior to the annual teaching session, consented and assented individuals were given brief surveys where they rated statements about their knowledge of SCD on a 5-point Likert scale. Participants were also given the opportunity to express any additional questions or thoughts they had about their overall knowledge. A post-education survey with the same questions was then administered via phone at least 1 week after the appointment. Differences in overall knowledge as well as knowledge in individual areas of understanding (e.g., medications) were assessed.

	Strongly Agree	Agree	Neutral	Disagree	Strongly Disagree
I can explain what sickle cell disease is and how it affects my body.	5	4	3	2	1
I can list what medications I take and explain why I take them.	5	4	3	2	1
I know what things trigger my pain/vaso-occlusive crises.	5	4	3	2	1
I know when to go to the doctor or seek medical help regarding my sickle cell disease.	5	4	3	2	1
I feel confident about my knowledge about my sickle cell disease.	5	4	3	2	1

Results

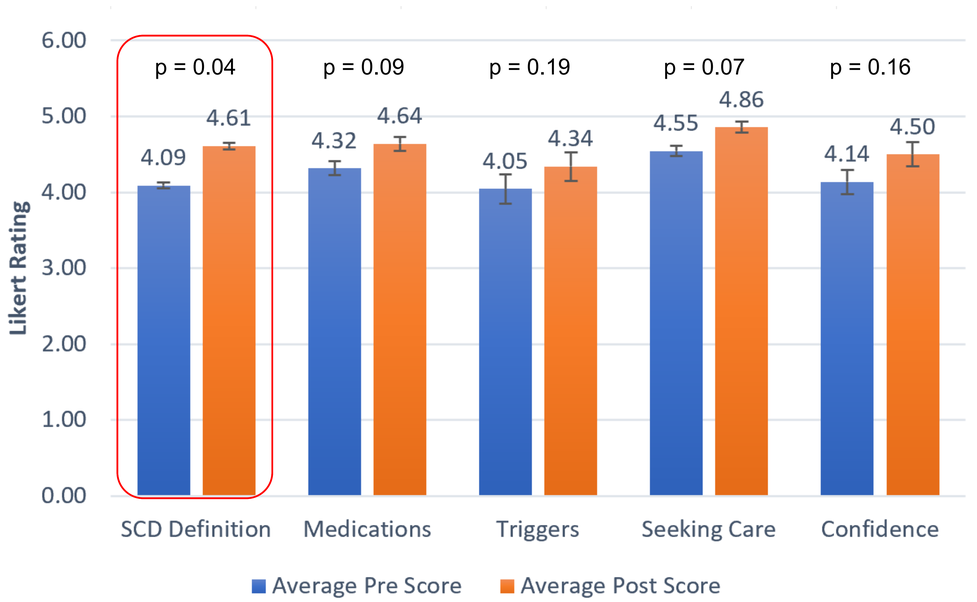

Figure 1. Average Likert rating for each survey statement on both pre- and post-education surveys.

Examples of Comments

- "The **older he gets**, the more questions I will have"
- "It was a good reminder on what I knew, what I forgot, and for my son to know certain things."
- "I want to know more about **medications** – why she takes them and what they are for."
- "Just want to know what long term **hydroxyurea** use results in."
- "I am glad to say that I learned about red blood cells and that her spleen has to be checked regularly."
- "**Adolescence/puberty** is changing his sickle cell episodes"
- "I have questions about **hydroxyurea** since I take the liquid form instead of capsules."

Discussion

While this is an ongoing project, current data suggest that study participants have a greater knowledge about sickle cell disease and how it individually impacts them after undergoing annual teaching sessions. Most notably, they have a better understanding of what sickle cell disease is ($p = 0.04$) and an overall increase in their survey scores ($p = 0.05$), indicating the education session is meeting the goal of increasing overall knowledge of SCD. Understanding of medications and individual triggers of vaso-occlusive crises, especially as patients enter adolescence and go through puberty, are frequent areas that participants have questions about. These topics may benefit from being addressed more completely or differently during the curriculum. Overall, having a multidisciplinary approach to patient education with an outlined yearly curriculum increases patient and family understanding of their disease, which in turn, improves patient outcomes and readiness to transition to adult care.

Limitations & Future Considerations

Limitations:

1. Follow-up via phone was difficult and often post-education surveys were not administered at the same time intervals, contributing to variability in time lapsed between surveys. Additionally, several participants were lost to follow-up.
2. Patients and guardians were quick to rank things at one of the extremes over the phone as compared to in-person when they were filling out the surveys themselves. This may be due to how the survey was phrased over the phone.

Future Considerations:

1. As more participants are enrolled and more data collected, aim to analyze the efficacy of the curriculum across age groups, sickle cell disease variants, as well as patients vs. guardians.
2. Many guardians of older children already have good foundational knowledge. A more in-depth survey may indicate specific areas for improvement

References

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