SciMedCentral

Annals of Pediatrics & Child Health

Research Article

Communicating Prognosis in Sickle Cell Disease: A Qualitative Study of Adolescents with Sickle Cell Disease, Their Parents and Providers

Lydia H Pecker[1]*, Michael Roth[1,2], Sari Landman[1], Leslie Cunningham[1,2], Ellen Johnson Silver[3] and Deepa Manwani[1,2]

[1]*Department of Pediatrics, Children's Hospital at Montefiore, Albert Einstein College of Medicine, USA*

[2]*Division of Pediatric Hematology and Oncology, Children's Hospital at Montefiore, Albert Einstein College of Medicine, USA*

[3]*Department of Pediatrics, Albert Einstein College of Medicine, USA*

***Corresponding author**

Lydia H Pecker, The Children's National Medical Center Fellow, Center for Cancer and Blood Disorders Washington, DC 20010, USA, Tel: 202-476-2140; Fax: 202-476-4424; Email: lpecker@childrensnational.org

Submitted: 21 November 2014

Accepted: 03 January 2015

Published: 05 January 2015

Keywords

- Sickle cell disease
- Prognosis
- Adolescents
- Communication
- Hematopoietic stem cell transplant

Abstract

Adolescents with sickle cell disease (SCD) and their parents are more optimistic about their future than their physicians. This may affect treatment preferences and therapy adherence. Disease specific recommendations for discussing disease course of children with SCD do not exist. To begin to address this gap, we held focus groups for adolescents with SCD 14-21 years old (n=6), parents of adolescents with SCD (n=4) and with pediatric (n=3) and adult (n=2) hematologists. SCD prognosis is complicated due to the uncertain disease course. Fear and worry are associated with discussing the future. Parents disagree with adolescents and hematologists about the best approach to discussing prognosis and oppose prognosticating that includes life expectancy. Guidelines to improve communication between physicians and families are needed.

ABBREVIATIONS

SCD: Sickle Cell Disease; **HU**: Hydroxyurea; **HSCT:** Hematopoietic Stem Cell Transplant.

INTRODUCTION

Despite advances in therapies for people with sickle cell disease (SCD), their life expectancy is half that of the general population [1,2]. Previous studies showed that many adolescents with SCD and their parents expect SCD to improve with time [3]. In their optimism, adolescents with SCD and their families are similar to cancer patients who overestimate their survival [4-6]. Yet this expectation contradicts the medical community's view of SCD as a chronic, progressive and morbid disease [7]. How families affected by SCD understand the risks associated with the diseaseinfluences treatment preferences, medication adherence, and participation in research [8]. Counseling families about these risks is challenging because SCD is a heterogeneous disease with variable outcomes and few clinical or biological markers to predict disease course [9].

Treatment options for patients with SCD are increasing [10]. Evidence now supports the use of hydroxyurea(HU) for many, if not all, children with HbSS and HbSβ°-thalassemia and cure with hematopoietic stem cell transplant (HSCT) is more available [11,12]. But significant barriers to treatment exist. Parents' and patients' understanding of the risks of untreated SCD, HU and HSCT and limited access to resources about these treatments compromise these therapies' use [3,13]. In addition, some physicians may have incomplete knowledgeof indicated therapies[14] and may be reluctant to discuss prognostic information [15].

The objective of this study was to understand how adolescents with SCD, their parents and providers define prognosis and to explore their preferences for discussing the future. This information informed the development of a survey for a multi-center study.

MATERIALS AND METHODS

Between December 2012 and January 2013, four confidential focus groups were conducted at the Children's Hospital at Montefiore. The groups were: adolescents with SCD ages 14 – 21, parents of adolescents with SCD, and adult and pediatric hematologist SCD specialists. Adolescents with SCD and their parents were recruited using our SCD patient database. A convenience sample of 10 pediatric and adult hematologists in academic practice received email invitations for a separate group. Verbal consent and assent was obtained for parents and adolescents as indicated by age. An expert panel (n=5) developed focus group guides through a literature review of prognosticating in chronic childhood illnesses and communication in SCD (Table 1). The panel included two pediatric hematologists, a pediatric psychologist with palliative care expertise, a developmental psychologist specializing in adolescents with chronic illness and qualitative research methods, and a pediatric resident with expertise in Africana Studies. The study was approved by the Montefiore Institutional Review Board.

Six adolescents ages 14 -21 participated in two groups. Because adolescents transition to adult care after their twenty-first birthday at our center, 18 – 21 year olds were included in the sample. Four parents (three mothers, one father) of adolescents with SCD aged 14 – 20 years old participated. One father-daughter dyad participated. Patient groups lasted 90 to 120 minutes. Five hematologists, two for adults and three in pediatric practice, participated via conference call. The hematologist group lasted 70 minutes. During each group, two researchers took notes, a data recording strategy that has been recommended for focus groups when audiotaping is not feasible [16]. Following each group, one study team member immediately transcribed the notes and clarified any inter-observer discrepancies. Qualitative content analysis was utilized to code the data into themes [17]. Discrepancies in data interpretation were resolved by research team consensus.

RESULTS

Patients, parents, and physicians associated the term prognosis with finality, the future, cure, life expectancy, and death. Adolescents, parents and hematologists agreed that prognosis defined as, "the usual course of a disease"[18] does not apply to SCD. Because all groups agreed that SCD does not have a "usual course," we explored participants' ideas about prognosis as it pertains to SCD. Four prognosis-related themes emerged: (1) Life expectancy in SCD (2) Emotional response to knowledge about the future (2) Preparing parents and children for the future (4) Learning through the experience of SCD. Table 2 provides quotes that exemplify each theme.

Life expectancy in SCD

When asked about prognosis, all groups introduced life expectancy. Hematologists thought discussing prognosis-as-life expectancy could support patients' and families' understanding of SCD's potentially grave complications, help shape families' treatment choices and might cultivate patient advocates for improved care for patients with SCD. All adolescents and parents endorsed thinking about and experiencing conversations about life expectancy. Parents related prognosis to receiving their child's diagnosis; for most, this included being told their child "wouldn't live long." Parents strongly objected to discussing life expectancy. Nevertheless they, like hematologists, had used threats of grave illness to motivate their children's medication adherence. Adolescents approved of this strategy. They thought that raising the possibility of death motivated adherence, although one teen was more concerned about his personal safety thanhis SCD. For adolescents, threats of dying were concrete, but life expectancy was more abstract. On direct questioning of the group, they evaded estimating their own life expectancy. For their neighbors they estimated life expectancy for women between 60 – 70 years old and for men, 25 – 60 years old.

Emotional responses to knowledge about the future

Fear and worry were prominent features of discussing the future. Adolescents recalled feeling isolated and scared. Parents expressed sorrow and hate for SCD on learning their child's diagnosis,and worry about medication adherence or exposing their children to conditions that could lead to acute illness. Hematologists discussed strategies for managing their patients' and parents fears and parents did report feeling reassured by pediatric specialists. Adolescents regretted missing school, prom, participating in sports, and not understanding SCD sooner, but were nevertheless determined to "do everything" their friends and siblings did, at a different pace.

Preparing parents and children for the future

To parents, their children's futures included school performance, relationships with doctors, hope for a cure, clinical

Table 1: Core Questions for Focus Group Sessions These questions guided focus group discussions with adolescents, parents and hematologists respectively.

Adolescents	1- How do you define prognosis? 2- How did you learn your diagnosis of SCD? 3- How do you discuss the future with your SCD doctor? 4- How would you like to discuss the future with your SCD doctor?
Parents	1- How do you define prognosis? 2- How did you learn your child's diagnosis of SCD? 3- How do you discuss the future with your child's SCD doctor? 4- How would you like to discuss the future with your SCD doctor?
Pediatric and adult hematologists	1- How do you define prognosis in SCD? 2- Is it important to discuss prognosis with your patients and/or their parents? Why or why not? 3- How do you discuss prognosis with your patients and/or their parents? 4- What are the barriers to discussing prognosis?

Table 2: These quotes exemplify the themes raised during focus groups.

Prognosis	
Adolescent	Prognosis is a scary word. It's a declaration of how it's going to be. But you can't go by what the doctors say. If they were right, I wouldn't be here.
Parent	This disease is like Dr. Jekyll and Mr. Hyde. He forgets he has sickle cell, then he gets sick and it's like "oh my god".
Pediatric hematologist	I tell them it's an unpredictable disease, that it's hard to predict the disease course and that each patient is an individual.
Life Expectancy	
Parent	I don't want to keep talking [with my doctor] if we're going to talk about life expectancy.
Adult Hematologist	They don't come to me for nonsense. They come to me to make them better. I don't ask them, "Do you really want to know what your disease is like?" Having that conversation is the best thing I can do for them…If I don't tell them, they can't help themselves.
Pediatric Hematologist	The problem with life expectancy is the papers are from the 1990s. I don't know mortality now. Likely life expectancy is not those numbers...It comes up at our transplant symposium because transplanters are more comfortable with those statistics [than pediatric hematologists].
Preparing parents and children for the future	
Adolescent	Ask the patient what they want to know, but do not frighten them. Tell them reality with a lollipop.
Parent	We have to give them hope. Hope for one day they can be cured. Hope of a long life. Hope to enjoy life. Hope to be like anyone else, to succeed.
Emotional responses to knowledge about the future	
Adolescent	The doctors knew from blood tests that I wasn't taking it [hydroxyurea] and told me it was serious and that I could die even if I didn't feel it. I skip days, but not often. The doctor scared me.
Adult Hematologist	I think we underestimate how afraid they are about their prognosis. Especially older patients who have been told they aren't going to live very long.
Learning through the experience of SCD	
Adolescent	I was five years old and I had pain. I asked my dad and he said I had blood shaped like the moon. But I didn't understand until I was 10 or 11 when I got the gist of it by going to a group with parents and kids.
Parent	My cousin had sickle cell and I never saw her sick until she got older. She had a regular life, went to school, had kids. She made me positive because if she's like this, he [my son] can do even better. I was scared to death [when I found out the diagnosis]. I knew what a classic sickler looked like. Long skinny arms, bony knobby kneed. I thought my son would look like that. I had a friend who died in college who had sickle cell. She just went to the ER and died.

trials and treatments for SCD, helping their children avoid and manage complications of SCD, motivating medication adherence, encouraging independence, and counseling their children on reproductive choices. Adolescents discussed sickness, school and associated absenteeism, carefully made friendships, romantic relationships and their risk of bearing a child with SCD. Hematologists said the future includedSCD progression, chronic complications, and transitioning from pediatric to adult care. Pediatric hematologists did discuss school. Adult hematologists focused on organ-specific disease complications.

All groups said that discussions with children and adolescents with SCD should be a collaboration between parents and doctors. Hematologists felt pressure to be realistic about potential SCD complications while making the future feel manageable and opportunity filled. Adolescents said children with SCD "need to understand that they're different" and should have SCD explained "as soon as they have their first attack" or in early middle school. In adolescent and hematologist groups, metaphors involving sugar were used. One hematologist explained, "I don't sugar coat anything," and a teen suggested parents and doctors tell children with SCD "reality with a lollipop." Both parents and hematologists wanted better support for communicating about SCD.

Learning through the experience of SCD

To think about the future, parents and adolescents remembered initial experiences with SCD. For parents, the future began with alarming conversations surrounding their child's birth. Most parents did not know they carried sickle cell trait and wanted this information. Adolescents and parents said that experiencing the complications of SCD was the way the disease became a concrete concern. Pediatric hematologists agreed that experience informed families' fears. Most adolescents remembered painful crises leading to conversations with doctors and parents about SCD. Adolescents identified their parents (mothers more than fathers), pastors, siblings, SCD support groups and doctors as the people who shaped their experience of SCD. Parents wanted more contact with their doctors and wished for guidance onhow to explain SCD to their children. All groups acknowledged the efforts of the others in the care of adolescents with SCD.

Adolescents remembered learning about SCD in the doctor's office and through paincrises. They described feeling blindsided by learning their diagnosis. Most understood they had SCD around age 10 when disease complications led to hospitalizations or interfered with school. Affirming these kinds of memories, parents said their children did not understand SCD until they experienced pain. Some parents felt encouraged by knowing others with SCD; others had relatives or friends who had died young from SCD. These experiences shaped their confidence and hope or caution and fear for their children's future.

As part of learning about their SCD, adolescents articulated an uneasy relationship with medications. They recalled home remediesthat did not help: bathing in bay leaves or milk and hot water, drinking bush tea and eating mushrooms. Some were skeptical of HU's benefits. Treatment adherence varied among participants. Parents and adolescents expressed ambivalence about HSCT. Adolescents described HCST as "the easy way out," "the miracle thing", and "too risky". Parents saw HSCT as a cure with limited reach.

An adolescent focus group member (JN) died of multi-organ failure shortly after transitioning to adult care and three months after participating in this research. JN experienced a decade of chronic pain and dozens of admissions for painful crises and acute chest syndrome. He had been non-adherent with hydroxyurea (HU) since his first prescription, ten years earlier. At the time of his death, neither JN, a college student and an active church member, nor his brother with similarly severe SCD, were taking HU. After JN's death, his brother resumed HU. During the focus group, JN explained his decision not to take HU and his optimistic outlook for the future (Table 3).

DISCUSSION

The NHLBI's recent practice guidelines for SCD acknowledge the importance of incorporating patient and family values into treatment recommendations, but this is an understudied area in SCD [11]. The focus groupshighlight that in order to discuss prognosis meaningfully with adolescents with SCD and their parents, it is important to address how SCD affects participation in school and associated activities, interaction with peer groups, and reproductive choices. Furthermore, acknowledging that fear and worry permeate all aspects of prognosis, while conveying hope is important to adolescents with SCD, their parents and their doctors.

Recommendations regarding how to discuss prognosis in SCD do not exist and guidelines for physician communication with families of children with chronic conditions of uncertain prognosis do not include studies of children with SCD [19]. Hematologists in this study were reluctant to make predictions about life expectancy and parents objected to discussing the subject. Our findings support evidence that parents of children with SCD think communication is an area of advancement for sickle cell care [20]. Regular conversations between parents of children with a wide array of chronic illnesses and their physicians about disease condition is associated with satisfaction with medical care, improved discussion of psychosocial concerns, and improved adherence [21-24].

Increasing availability of HSCT, a treatment with a complicated risk-benefit profile, makes discussing SCD prognosis with families of affected children necessary [25,26]. Significant barriers to bringing even eligible children with SCD to transplant exist [25-28]. These barriers are incompletely understood, but include inadequate SCD education for families [29]. As indications for HSCT in SCD expand, pediatric hematologists have a reason to discuss prognosis explicitly with families of children with SCD.

This study offers several intriguing areas for future research. First, experience with the grave consequences of SCD correlates with parents' and patients' more negative expectations for the impact of SCD on their lives,[3] and suggests that an opportunity to discuss treatment options and adherence may exist following significant medical events. Understanding what motivates treatment decisions in SCD will inform communication guidelines. Second, some adolescents spoke negatively about HSCT and struggled to imagine life without their SCD. Like all adolescents, those with SCD are engaged in the developmental task of identity formation. Resistance to HSCT may reflect their psychological work of accepting and incorporating their illness into their identity [25,30]. Specific ways to support parents and adolescents as they develop their adult identity may help foster resilience [31]. Finally, some adolescents noted a synergism between their religious and disease identities; JN felt these two identities were at odds. The role of religion and religious leaders in acceptance or rejection of treatments for SCD is incompletely explored [32].

Limitations and strengths

This current study's strengths are focused on the qualitative exploration of an under studied area of SCD. Through studies of this kind, the opinions of adolescents with SCD and their parents, may be incorporated into guidelines that inform care. The limitations of this study include its small sample size, which may lead to over or under representation of certain themes. Participants were motivated to attend focus groups despite the absence of a financial reward. It was more difficult to recruit younger adolescents, and our sample was weighted towards 18 – 21 year olds. Because participants were recruited from within a single institution, their experiences and those of their parents may not be representative of the experiences or opinions of adolescents with SCD as a whole and they also may be informed by the communication styles and strategies and prescribing practices of a small number of pediatric hematologists who treat them.

CONCLUSIONS

For all focus group participants, SCD prognosis is complicated by the disease's unpredictable course and is associated with fear and worry. Parents disagreed with adolescents and parents about whether to discuss life expectancy. Adolescents and their parents consider the future in terms of function, reproductive choices and disease management whereas hematologists discussed

Table 3: The adolescent who died shortly after transitioning to adult care explained his outlook and treatment choices.

Preparing for the future	My parents and others tell me I'll grow out of it, that I should just wait for that. Doctors tell me how important it is to take my meds. I'm very religious. I pray about it and hope that healing happens. I haven't been to the hospital in 6 months. It does seem to be improving as I get older. I've had many fewer hospitalizations than when I was younger.
Learning through experience	Hydroxyurea really does help. But the benefits don't outweigh the consequences for me. I've stopped and it doesn't seem to have bad effects. I prefer how I am without it now. I don't take it as a choice.

more disease specific concerns.Parents and hematologists want guidance on how to discuss the future with their children and patients and their families respectively. This study addresses an under represented area of SCD research; its limitations are the small number participants with parents and adolescents recruited from a single institution. This work informed a multi-center survey study of adolescents with SCD, their parents, and hematologiststhat is underway. The goal isto define each groups' experiences with and preferences for discussing the future so that decision guides for families and communication guidelines for pediatric hematologists can be developed.

ACKNOWLEDGEMENTS

The authors are grateful to the adolescents, parents and doctors who participated in these focus groups and thank Katherine Kelly for her support with the development of this manuscript. We remember J.N.

REFERENCES

1. Platt OS, Brambilla DJ, Rosse WF, Milner PF, Castro O, Steinberg MH. Mortality in sickle cell disease. Life expectancy and risk factors for early death. N Engl J Med. 1994; 330: 1639-1644.
2. Darbari DS, Kple-Faget P, Kwagyan J, Rana S, Gordeuk VR, Castro O. Circumstances of death in adult sickle cell disease patients. Am J Hematol. 2006; 81: 858-863.
3. Roth M, Krystal J, Manwani D, Driscoll C, Ricafort R. Stem Cell Transplant for Children with Sickle Cell Anemia: Parent and Patient Interest. Biol Blood Marrow Transplant. 2012; 18: 1709–1715.
4. Weeks JC, Cook EF, O'Day SJ, Peterson LM, Wenger N, Reding D, et al. Relationship between cancer patients' predictions of prognosis and their treatment preferences. JAMA. 1998; 279: 1709–1714.
5. Edwards KE, Neville BA, Cook EF Jr, Aldridge SH, Dussel V, Wolfe J. Understanding of prognosis and goals of care among couples whose child died of cancer. J Clin Oncol. 2008; 26: 1310-1315.
6. Mack JW, Wolfe J, Grier HE, Cleary PD, Weeks JC. Communication about prognosis between parents and physicians of children with cancer: parent preferences and the impact of prognostic information. J Clin Oncol. 2006; 24: 5265-5270.
7. Thompson AL, Bridley A, Twohy E, Dioguardi J, Sande J, Hsu LL, et al. An educational symposium for patients with sickle cell disease and their families: Results from surveys of knowledge and factors influencing decisions about hematopoietic stem cell transplant. Pediatr Blood Cancer. 2013; 60: 1946-1951.
8. Hankins J, Hinds P, Day S, Carroll Y, Li CS, Garvie P. Therapy preference and decision-making among patients with severe sickle cell anemia and their families. Pediatr Blood Cancer. 2007; 48: 705-710.
9. Meier ER, Wright EC, Miller JL. Reticulocytosis and anemia are associated with an increased risk of death and stroke in the newborn cohort of the Cooperative Study of Sickle Cell Disease. Am J Hematol. 2014; 89: 904-906.
10. Manwani D, Frenette PS. Vaso-occlusion in sickle cell disease: pathophysiology and novel targeted therapies. Blood. 2013; 122: 3892-3898.
11. National Heart, Lung, and Blood Institute. Sickle-cell-disease-report. 2014; 1–161.
12. King A, Shenoy S1. Evidence-based focused review of the status of hematopoietic stem cell transplantation as treatment of sickle cell disease and thalassemia. Blood. 2014; 123: 3089-3094.
13. Omondi NA, Ferguson SES, Majhail NS, Denzen EM, Buchanan GR, Haight AE, et al. Barriers to hematopoietic cell transplantation clinical trial participation of african american and black youth with sickle cell disease and their parents. J Pediatr Hematol Oncol. 2013; 35: 289–298.
14. Ware RE. How I use hydroxyurea to treat young patients with sickle cell anemia. Blood. 2010; 115: 5300-5311.
15. Christakis NA. Death Foretold. University of Chicago Press. 2001.
16. Stewart DW, Shamdasani PN. Focus Groups. SAGE Publications. 2014.
17. Hsieh HF, Shannon SE. Three approaches to qualitative content analysis. Qual Health Res. 2005; 15: 1277-1288.
18. Merriam-Webster Inc. Merriam-Webster's collegiate dictionary. 2004.
19. Levetown M, American Academy of Pediatrics Committee on Bioethics. Communicating with children and families: from everyday interactions to skill in conveying distressing information. Pediatrics. 2008; 121: e1441–1460.
20. Kenyon CC, Kavanagh PL, Fiechtner LG, Textor TE, Wang CJ. Setting the agenda for quality improvement in pediatric sickle cell disease. J Natl Med Assoc. 2012; 104: 337-341.
21. Nobile C, Drotar D. Research on the quality of parent-provider communication in pediatric care: implications and recommendations. J Dev Behav Pediatr. 2003; 24: 279-290.
22. Bryony AB, Patricia Sloper. Chronically ill adolescents' experiences of communicating with doctors: a qualitative study. J Adolesc Health. 2003; 33: 172–179.
23. Drotar D. Physician behavior in the care of pediatric chronic illness: association with health outcomes and treatment adherence. J Dev Behav Pediatr. 2009; 30: 246-254.
24. Courtney DT, Zora RR, Michael RJ, Sohail RR, Rathi VI, Lane Faughnan, et al. Adherence to study medication and visits: Data from the BABY HUG trial. Pediatric Blood & Cancer. 2009.
25. The Management of Sickle Cell Disease. 2002; 1–206.
26. Christakis NA. Prognostication and bioethics. Daedalus. 1999; 128: 197-214.
27. Khalid L. Rehman. Bone marrow transplantation for sickle cell disease. N Engl J Med. 1996; 335: 1845–1846.
28. Kline M Ronald. Pediatric hematopoietic stem cell transplantation. 2006.
29. Hansbury EN, Schultz WH, Ware RE, Aygun B. Bone marrow transplant options and preferences in a sickle cell anemia cohort on chronic transfusions. Pediatr Blood Cancer. 2012; 58: 611-615.
30. Far From the Tree. Scribner. 2012.
31. Bohanek JG, Marin KA, Fivush R, Duke MP. Family narrative interaction and children's sense of self. Fam Process. 2006; 45: 39-54.
32. Toni-Uebari TK, Inusa BP. The role of religious leaders and faith organisations in haemoglobinopathies: a review. BMC Blood Disord. 2009; 9: 6.

Cite this article

Pecker LH, Roth M, Landman S, Cunningham L, Silver EJ, et al. (2015) Communicating Prognosis in Sickle Cell Disease: A Qualitative Study of Adolescents with Sickle Cell Disease, Their Parents and Providers. Ann Pediatr Child Health 3(1): 1031.