THE REAL EFFECTS OF SICKLE CELL DISEASE ON CHILDREN AND ADOLESCENTS

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ABSTRACT

Informal interviews were conducted with four patients with Sickle Cell Disease (SCD) as inpatients in 2013. The interviewers tried to elucidate the psychosocial effects of SCD, including their quality of life between and during hospital stays. Treatment options including hydroxyurea and the idea of multimedia communication were discussed.

KEYWORDS

Paediatric haemoglobinopathy, Hospitalisation, Multimedia support

INTRODUCTION

Sickle Cell Disease (SCD) is the most common genetic disorder in England and occurs in approximately 1 in every 2400 live births (11). In particular sections of the world, as with many African countries, the prevalence can be as high as 30% (10). SCD affects the shape of red blood cells and therefore their ability to move through the body. An alteration in the structure of haemoglobin allows it to stack lengthwise when deprived of oxygen causing the red blood cell to “sickle” (12). Sickled cells become sticky and can adhere to other blood cells and the walls of blood vessels, leading to hypoxia in the microcirculation and sometimes pain in the affected individual (10).

A sickling “crisis” is a significant handicap but individuals are affected to different degrees. Some may be symptom free for many years while others have poor school attendance and multiple hospital admissions (12). A crisis typically begins suddenly and may be precipitated by intense activity, dehydration, cold, or internal stressors such as infections (10).

SCD impacts a youth’s psychosocial well-being. Chronic illness can place significant stress on families emotionally, financially, and physically (13;9). Often communication and social support between family members and the hospitalised child or adolescent can suffer due to other obligations, time or financial constraints. Chronically ill children often have a hard time with
socialisation and adjusting to normal developmental transitions due to time spent in hospital and away from peers (13). Hospitalised and ill children and adolescents need social support and contact from family and peers in order to normalise and transition through development stages while also adapting to life with a chronic illness. Family involvement has been shown to produce better outcomes for the ill child, both emotionally and physically (13;7).

Both interpersonal relationships with family and peers as well as the academic realm of a patient may be compromised. Painful events from SCD can lead to frequent absences from school and long-term absences due to hospitalisation. In England, SCD students have an 8.4% absence rate as compared to the 3.47% average (2). Patients with SCD have been stereotyped by teachers as lazy or lacking motivation (2), when in reality often require more support to balance their absences and success. SCD youth also miss the opportunities the school environment provides to socialise and spend time with peers. Most importantly SCD patients should be identified as ‘people first’ not ‘disease first’ (9). All efforts should be made to not only provide the best medical care and treatment, but also the best psychosocial care. Social support and communication is essential to the overall quality of life for youth with SCD (7;13).

PARTICIPANTS

Four patients with SCD in the paediatric ward at Ealing Hospital NHS Trust consented to being interviewed during the summer of 2013. All had been hospitalised with a painful crisis caused by SCD. The specific complaints were of severe arm or leg pains. Two were adolescents, a 15 year old boy and a 13 year old girl, each with frequent histories of hospitalisations averaging once a month and two were children, aged 5 years, a boy and a girl. It was the 35th hospitalisation for the 5 year old girl but only the 2nd for the 5 year old boy.

METHODS

This was an informal interview process with inpatient children and adolescents with SCD at Ealing Hospital. This is a small district general Hospital in west London with a school within the ward. The local population has a large number of patients with haemoglobinopathies; management of these is regulated by a local network (including community nurse support) and one of the parents has a long-established patient support group. There are local and national websites that provide educational material and a programme of meetings. A child psychology masters student and a medical student collaborated to interview patients on their experience of the Hospital, what they understand about their illness and treatment options, and to explore the social implications of hospitalisation and chronic illness. The two adolescents were able to answer questions on their own, while the two
children had a little help from their mothers, both present during the interviews.

The participants were first asked about their history of SCD and the reason for the current hospitalisation. The interviewers then asked about frequency and duration of hospitalisations. Each participant was then asked to explain what they know about SCD to gauge their understanding and involvement in their illness. They were also asked about others they know with the illness that they can talk to and relate to.

Following the initial questions about their illness, each participant was asked about the social impacts of hospitalisation and SCD. They were asked about social support from family, including siblings, how often they are visited in the hospital and how often could they talk with family. They were asked if their friends know about their illness or hospitalisations and if they were able to speak with them during hospitalisations. They were asked what they miss most during their hospitalisation.

Questions regarding the effects of SCD on their school lives, participation in activities, and socialisation with friends were also put to them. Finally, the participants were asked about current treatments regimens as well as other treatment options. Treatment with hydroxyurea was discussed, including concerns that each participant had that prevented them from using this treatment.

The interviewers concluded with discussion on possible social support options with the patients, specifically the idea of using Skype to communicate with family and friends during hospitalisation.

**Discussions related to Treatment**

Treatment options have been vastly studied to both treat and cure SCD (12). Hydroxyurea, also known as hydroxycarbamide, has been shown to lower the overall number of painful crises and amount of blood transfusions, resulting in decreased hospital admissions (10;5). Hydroxyurea works by increasing the amount of fetal haemoglobin, which is not affected by the disease, and reducing the amounts of other blood cells, which decreases the concentration of cells in the blood (10).

Participants were asked about the use of hydroxyurea. All four of the children referred to their parents as the decision maker in treatment options. Only one of the children was currently taking the drug. The parents each had different understandings of the risks and benefits of the drug. Some were worried that hydroxyurea may be ineffective, or expressed concerns about its side effects, such as commonly associated hair loss, stigma, and painful procedures (1).

One parent of a younger participant claimed not to have heard of the drug as an option for treatment: their child did not meet the criteria for treatment, that is of more than 3 admissions for pain episodes in a 12 month period (11). However, although not hospitalised, the child frequently sat out on activities as soon as he felt any pain. The frequency of pain crises is one of the best
indicators of quality of life (3); reducing the frequency of pain crises and increasing the ability to participate in daily activities increases the quality of life.

**Discussions related to Psychosocial effects**

The four participants and their family members were very willing to engage with the interviewers. Many of the concerns with hospitalisation and social issues were common to the four participants and confirmed published findings. The adolescents both had a moderate understanding of SCD and the factors that contribute to pain crises, typically resulting in hospitalisation. The mothers of the two children only had a basic understanding of SCD pathology but did have a good understanding of the factors that lead to painful crises.

The adolescent boy stated that he did not know anyone else with SCD nor was he interested in talking with anyone else with the illness. The adolescent girl knew of some people with SCD and thought it might be helpful to talk with others who have the SCD. The mothers of the two children did not know anyone else with SCD, other than those met during hospitalisations. In conversations with the participants, it was clear they did not attend the parent support group locally or within London, or had any opportunity to communicate with other SCD peers. Given that children and adolescents with SCD often suffer with feelings different than their peers, the desire to hide their disease to avoid judgment, and face social isolation due to frequent hospitalisations (6;7), better publicity of the social support network among SCD peers would be beneficial. This might provide youth with an outlet to voice their concerns and experiences, interact with others just like them, and rely on each other for social support without fear of judgment.

Aside from examining the idea of social support from other SCD peers, the support and communication with family and siblings was discussed. Each of the four participants was visited daily by a parent, for varying amounts of time based on age and parental obligations. However, each participant discussed missing their siblings often during hospitalisation and how much they enjoyed it when they were able to visit. The frequent hospitalisations can place a lot of stress on the family unit and disrupt typical social interactions between family members. It has been shown that social support, especially from family, is a significant indicator of quality of life and adjustment for SCD children and adolescents (7;13;14).

Childhood and adolescence are critical periods for psychosocial development. Youths begin to build peer social relationships, develop individual identities, and seek autonomy. However, these normal development patterns can be disrupted by SCD (13). The two adolescents discussed how they keep their illness a secret and do not want to involve their friends to avoid drawing unwanted attention or being viewed as different, a common fear among SCD children and adolescents (2). Those frequently hospitalised participants are removed from peers and social settings and missed their friends. The adolescent girl stated that she really enjoyed when she was able
to speak with her friends and wished she could more often. Both adolescents were still very dependent on their parents during a time when typical adolescents are seeking autonomy (9). The two children often missed out on physical activities at school and with friends due to hospitalisations and fear of painful crises. The children described feelings of sadness and fear of getting sick, holding them back from participating in activities with other children. The children were not able to participate in physical education classes or sporting activities like their peers, reducing quality of life.

A further area of the children and adolescents lives that was negatively affected by SCD was school. All of the participants discussed the frequency with which they are absent from school, either staying at home due to pain or admitted to the hospital. These frequent absences cause disruptions to learning and socialisation with peers and teachers (9). The participants all stated that they missed being at school and especially missed their friends. The adolescent girl said that she does her homework while in the hospital and emails with the teacher to keep on track.

CONCLUSIONS

Sickle cell disease has profound effects on the physical health, as well as the psychosocial and emotional health of young patients. Common experiences that all four participants missed most while being hospitalised were their family, friends, television, internet, and video games. Addressing the lack of communication with family and friends is a challenge. The potential to use tablets with video links excited all the patients. Each favored the idea of using video chat to regular voice chat and said it would make them very happy to have such a programme available.

Multimedia communication could also be used to connect hospitalised children with teachers and schools, with other children with the same illness, routine medical appointments that could then be handled from home, and involvement in special events. Some hospitals use multimedia technology to connect health care providers with patients to remotely monitor pain and provide treatment reminders (8;1;14). More research and pilot programmes to evaluate multimedia communication technology to connect hospitalised children with social support networks would be useful. For instance, just how significant are the difficulties with establishing safe internet access to a paediatric ward? How can schools and support groups link more effectively with a widespread group of patients without compromising patient confidentiality and protecting identity? One such pilot programme called KidsKonnected (4) is currently being developed by one of the authors. These resources might benefit the psychosocial issues associated with a chronic illness such as SCD and might have the potential to improve the quality of life for patients.
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REFERENCES


