Advances in Social Sciences Research Journal – Vol.2, No.9
Publication Date: Sep. 25, 2015
DoI:10.14738/assrj.29.1478.
Ani, C., & Garralda, E., & Anie, K. A., & Wilkey, O., & Hodes, M. (2015). Empirical Evaluaton of a Theory of stigma in Childrne and Adolescents with Sickle cell Disease. *Advances in Social Sciences Research Journal,* 2*(9)* 119-126.

Empirical evaluation of a theory of stigma in children and adolescents with sickle cell disease

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Abstract

Background. There is limited research in self-perceived stigma among children and adolescents with sickle cell disease (SCD). We applied the concept of "stigma dimensions" to measure the predictors of self-perceived stigma in young people with SCD.Objective. In accordance with "stigma dimensions", we hypothesized that young people with visible clinical features of SCD (jaundice and leg ulcers), and greater interference with their daily lives and social activities (frequent hospital admissions and school absences) would have increased self-perceived stigma. Patients and methods. 93 children and adolescents with SCD aged 10-19 years in the UK completed questionnaires on self-perceived stigma. Socio-demographic and clinical characteristics of SCD were also obtained. Young people were recruited from paediatric haematology or sickle cell outpatient clinics in London and the Sickle Cell Society (National Charity). Participants with all types of SCD were included; exclusions were those who were not fluent in English or unwell at the time of the survey. Results. Mean age was 14.2 years (SD 2.1 years) with 51% males and 49% females. 66% of the respondents had one or more hospital admissions in the preceding year, 46% were jaundiced, and 5% had leg ulcers. Consistent with our hypothesis and "stigma dimensions", regression analysis showed that those who experienced more disruption in their daily lives (frequent hospital admission and less school attendance) had increased self-perceived stigma. However, the presence of leg ulcers or jaundice was not associated with increased self-perceived stigma. Conclusion. Interventions in SCD that effectively moderate disruption of daily activities and improve quality of life could help reduce self-perceived stigma among young people with SCD.

Key words: Sickle cell, stigma, visibility, hospital admission, school absence

BACKGROUND

Sickle cell disease (SCD) comprises a group of genetic blood disorders that result from abnormal haemoglobin production. The predominant symptom is pain resulting from the blockage of blood vessels due to abnormally shaped "sickle" red blood cells. Other complications include severe anaemia, splenic sequestration, organ damage, stroke, priapism,

leg ulcers, and jaundice. SCD can pose medical and psychosocial challenges to affected individuals and their families [1].

The potential contribution of stigma to psychosocial difficulties in SCD is increasingly becoming recognised [2-5]. Despite these encouraging advances, empirical explorations of SCD-related stigma remains relatively underdeveloped and limited compared with other chronic medical conditions such as epilepsy. This study adopts a theory-driven research strategy [6] to contribute to a growing body of evidence, and explores the application of a specific theory in stigma namely "Stigma Dimensions" to children and adolescents with SCD.

The concept of stigma dimensions [7,8] is one of the extensions of Goffman's original stigma theory. Stigma dimensions help to predict why certain attributes and not others are more likely to become stigmatising [7,8]. This study examines the application of two specific stigma dimensions of "Visibility" and "Disruptiveness" to children and adolescents with SCD. We focused on these two dimensions as they are more readily measurable in SCD.

The dimension of "visibility" refers to the extent to which the attribute is obvious to others, difficult to conceal, and or aesthetically challenging. The prediction is that the more visible and disfiguring an attribute, the more stigmatising it is likely to be. In relation to SCD, some affected persons have easily recognisable physical manifestations such as jaundice, leg ulcers, and delayed physical development in adolescence [1]. In severe cases, especially where the availability of effective medical treatment is limited, deformities of some flat bones may result in dental malocclusion and bossing of the forehead [9].

The stigma dimension of "disruptiveness" describes the extent to which possessing the attribute interferes with the person's personal functioning and interpersonal relationships. Although the clinical course of SCD is variable and many affected persons live relatively healthy undisrupted lives, a proportion require frequent hospitalisation as a result of acute pain episodes and symptoms [1]. These acute episodes may disrupt schooling, employment, and other social activities [10] making concealment more difficult and increasing the potential for stigma.

The primary objective of this study is to explore the theory of stigma within the context of visibility of clinical manifestations in SCD and disruptiveness arising from these complications. We hypothesised that children and adolescents with SCD who have more visible appearances (jaundice and leg ulcers) and or experience more disruptiveness (frequent hospital admissions and school absence) will have increased self-perceived stigma. This type of theory-driven research could further improve understanding of the factors promoting stigma in people with SCD, and inform better interventions to diminish it.

METHODS

This was a cross-sectional questionnaire survey of children and adolescents with SCD aged 10-19 years in the UK. All genotypes were included; exclusions were those who were not fluent in English or medically unwell at the time of the survey. Recruitment was through the Sickle Cell Society (National Charitable Organisation), and three paediatric haematology or sickle cell outpatient clinics in London (Central Middlesex, North Middlesex, and St Mary's Hospitals). Two approaches to recruiting participants were employed. First, study packs were sent by post to families who were members of the Sickle Cell Society, and children and adolescents with SCD were invited to complete and return the questionnaire in a prepaid reply envelope. Second, recruitment from the three outpatient clinics was carried out by an assistant

psychologist or specialist nurse who were trained and supervised by the first author. Families were approached as they attended routine clinic appointments with information about the research. Those who expressed an interest were given a study pack, with the option to consent and complete the questionnaire in the clinic or at home and return by post. The study pack was also posted to those who were not due a clinic appointment soon. Each participant was offered a £10 voucher for completing the questionnaire. The study was approved by the South West NHS Multicentre Research Ethics Committee (06/MRE06/10).

MEASURES

A study questionnaire was constructed from standardised and validated instruments comprising Socio-Demographics, Self-Perceived Stigma, Disruptiveness, and Visible Signs of SCD.

Self-perceived stigma

This was assessed with three items adapted from a previous study of stigma in young people with epilepsy [11]. The latter study presented a methodology for studying stigma in adolescents with chronic conditions. A subsequent study in adolescents who stutter demonstrated the adaptability of the methodology [12]. The adapted questions are shown in Table 1. To maximise the analytical power of the stigma questions, the answers were summed to create a dimensional "stigma scale" (Mean 4.0, Range 3-10; higher scores indicate more self-perceived stigma). The scale showed a good internal consistency (Cronbach Alpha 0.80).

Disruptiveness

Disruptiveness was measured with two items (a) frequency of hospital admissions (defined as an overnight or longer stay in hospital, and (b) frequency of school absence) [13]. These measures were presented as incremental ordinal scales with higher scores indicating more admissions or more school absence. As the two variables correlated significantly ($r = .33$, $n = 88$, $p = 0.003$), principal component factor analysis was used to combine them to create a common factor for "disruptiveness".

Visible signs of SCDs

The questionnaire prompted respondents to indicate if they had jaundice and or leg ulcers at the time of completing the questionnaire.

RELIABILITY OF DATA COLLECTION

Reliability was checked by comparing questionnaire responses on three variables against the medical records of 10 randomly selected participants in one centre. The check found 100% concordance. Furthermore, demographics and sickle cell variables of 10 randomly selected respondents were compared with 10 randomly selected non-respondents. The comparison found more admissions in the preceding 12 months among the respondents (Mean = 2.0, SD =2.11) compared with non-respondents but the difference was not significant (Mean = 0.7, SD = 0.82) ($t = 1.82$, $df = 18$, $p = 0.08$, CI, -0.20, 2.80). The two groups did not differ significantly in age, presence of leg ulcers or prescription of hydroxyurea.

ANALYSIS AND HYPOTHESIS TESTING

Data entry and analysis was conducted with SPSS Version 22. Interval and ordinal data which were normally distributed were summarised with means and standard deviations. Bi-variate comparisons to determine group differences were conducted with chi-square, Fishers exact, and t-tests. Due to small numbers in some cells, the six-group OPCS socio-economic measure was recoded into three groups (I and II, III and IV, V and VI). Hierarchical linear regression

model was used to test the study hypothesis. Stigma scale was entered as the dependent variable. Age, and gender were entered together in the first block, while disruptiveness was entered in the second block. Measures of visibility were not included in the regression model as they were not statistically significantly associated with stigma in bivariate analyses. The pattern of missing data was random and cases were excluded only if they were missing the data required for that particular analysis [14]. Statistical significance was set at the 0.05 level.

RESULTS

A total of 93 young people aged 10-19 years (Mean, 14.2 years, SD 2.1) participated in the study of which 51% were males and 49% females. More respondents (59.1%) completed the questionnaires at home. There was a greater number of respondents recruited from outpatient clinics (73.1%) than through Sickle Cell Society (26.9%). There were no statistically significant differences in the proportion of participants recruited from hospitals clinics or the Sickle Cell Society in gender: (males = 37/46, (females = 29/44, X = 2.43, df = 1, p = 0.12); Socio-economic group: (X = 3.82, df = 2, p = .15), frequency of hospital admissions (M = 2.33, SD = 1.29 vs M = 2.30, SD = 1.61, t(86) = 0.10, p = 0.92 or self-perceived stigma (M = 3.92, SD 1.72 vs 4.04, SD 1.98, t(87) = -0.27, p = 0.78). Two-thirds of the young people had at least one hospital admission in the preceding year. Rates of school attendance showed a high variability with some (9%) having no loss of school days in the previous year while 18% had more than 28 days absence. Nearly half of the respondents (46.1%) were jaundiced but only 5.6% had leg ulcers.

QUESTIONS ABOUT STIGMA

Responses to these questions are presented in Table 1.

Table 1: Questions About Stigma

	N(%)
Do you think that having Sickle Cell affects whether people want to be friends with you?	
Never	65 (73.0)
Rarely	14 (15.7)
Sometimes	7 (7.9)
Often	3 (3.4)
Do you think that having Sickle Cell affects whether people like you or not?	
Never	76 (83.5)
Rarely	8 (8.8)
Sometimes	6 (6.6)
Often	1 (1.1)
Do you think that having Sickle Cell affects whether or not you are invited to people's homes or to parties?	
Never	75 (82.4)
Rarely	8 (8.8)
Sometimes	6 (6.6)
Often	2 (2.2)
Stigma scale – Mean (SD)	4.0 (1.8) Range 3-10

Table 1 shows that most of the young people with SCD were less likely to acknowledge self-perceived stigma. For example, only 11.3% thoughts that having sickle cell affected whether people wanted to be friends with them. Similarly, only 7.7% thought that having Sickle Cell affected whether people liked them or not. Female respondents scored significantly higher on the composite stigma scale (M = 4.35, SD = 2.14) than males (M = 3.58, SD 1.31, t(86) = -2.03, p = 0.046). However, the stigma scales was not significantly associated with age (r = =.09, p = 0.41) or Socio-economic group [F(2,17.16) = 2.21, p = 0.14] (the assumption of homogeneity of variance was violated; therefore the Brown-Forsythe F ratio is reported)

STIGMA AND VISIBLE SIGNS OF SCD

The presence of leg ulcers was not statistically significantly associated with the stigma scale (M = 6.0, SD = 3.1 vs M = 3.85, SD = 1.65 t(4.2) = 1.59, p = 0.19). Similarly, no association was found between the presence of jaundice and stigma (M = 3.77, SD 1.54 vs M 4.15, SD 1.98, t(85) = -0.97, p = 0.33). These findings are not in keeping with the first study hypothesis.

Stigma and Disruptiveness

Disruptiveness was significantly and positively correlated with stigma scale (r = .33, n = 83, p = 0.002), which provides preliminary support for the second study hypothesis.

Multivariate analysis

Hierarchical multiple linear regression was used to explore whether disruptiveness predicted the stigma scale after controlling for age and, gender.(Table 2). We chose hierarchical linear regression so that the relative contribution of disruptiveness to the total variance could be more readily evidenced. In line with Pallant's [14] recommendation to enter confounders first, age and, gender were entered together in Step 1. The model explained 6.6% of the variance in stigma but was not statistically significant [F(2,76) = 2.67, p = 0.076]. Disruptiveness was entered alone in Step 2.The model was statistically significant and explained 11.8% of the variance in stigma [F(1,75) = 4.49, p = 0.037]. In the final model, only the stigma dimension of disruptiveness was statistically significant (beta = 0.41, p = 0.037). . Extensive regression diagnostics showed that the model met the underlying assumptions for linear regression. For example Normal Probability Plot showed points that were on or close to the line, and Durbin-Watson score of 2.23 showed the data met the assumption of independent errors.

DISCUSSION

The main aim of this study was to explore the application of the stigma dimensions of "visibility" and "disruptiveness" to SCD. Consistent with our hypothesis, we found that disruptiveness predicted self-perceived stigma but contrary to our hypothesis, the two measures of "visibility" (jaundice and leg ulcers) were not associated with stigma in this cohort. Thus our overall hypothesis was partially supported.

Stigma dimensions and SCD

The findings of this study provide partial support for the application of stigma dimensions to SCD. As outlined earlier, stigma dimensions are the characteristics that determine the stigma potential of conditions [7,8]. The stigma dimension we found applicable is "disruptiveness". In this study, disruptiveness was derived as a common factor of frequency of hospital admission and school absence. Almost two thirds of the respondents had had one or more hospital admissions in the previous year, and 18% had had more than 28 days absence from school. These episodes of hospital admissions or being away from school are likely to be disruptive to education and social activities. In terms of stigma, these events may prompt curiosity from

peers about the child's absence thus making it more difficult for the young person with SCD to manage information about their condition.

Information management by people with stigmatising conditions involves a variety of strategies including concealment and or preventative disclosure [15,16]. Concealment may provide some relief but the constant threat of potential discovery may lead to preoccupation, obsessive preoccupation and intrusive thoughts about the 'secret [16]. While disclosure may lead to more stigma, preventative disclosure may have the advantage of reducing the negative impact of others finding out in other ways. It also allows an opportunity to provide accurate information and correcting in advance the negative impression that others could form based on misinformation [16].

The likelihood of adopting preventative disclosure as a coping strategy is to some extent a function of the person's perception of the predictability of their illness [16]. Those who feel able to control the manifestation of the physical illness may feel they can influence the social consequences more positively [16]. This study of young people with SCD shows that for some of them, the disruptiveness associated with the illness makes their situations less predictable, thereby reducing the potential for them to gain the advantages of controlled preventive disclosure. This suggests that improving medical and psychosocial management in SCD to limit the frequency of hospital admissions and school absences could contribute to their ability to self-manage their condition and reduce self-perceived stigma. An example of such measures includes avoiding the scheduling of routine medical and or nursing appointments during school hours as much as possible.

Contrary to one of our hypothesis, the study did not support the applicability of the stigma dimension of "visibility" to this cohort. The stigma dimension of visibility refers to the extent that SCD has attributes that are aesthetically challenging to others. While advanced medical treatment in the UK makes gross physical signs of SCD, such as dental malocclusion and bossing of the forehead, uncommon, some people with SCD in the UK still have visible characteristics. In this study, almost half of the subjects (46%) were jaundiced but very few (5%) had leg ulcers. Incidentally, neither the presence of jaundice nor leg ulcers was associated with self-perceived stigma. This is surprising given that an association between visibility and stigma has been shown in other chronic medical conditions [17]. It is possible leg ulcers in particular were not noticeable because these are usually concealed with clothing, and jaundice not visibly enough to affect young people's perception of stigma, thus reducing the risk of uncontrolled disclosure. However, a different outcome might have been seen if the study had been done in other countries where physical features of SCD such as bossing of the forehead are still prominent.

Self-perceived stigma and SCD

In order to put in perspective the level of self-perceived stigma endorsed by the young people with SCD in this study, we compared their responses with those of young people with epilepsy [11] and stuttering [12], where similar methodology was used. For example, in response to the question "Do you think that having Sickle Cell affects whether people want to be friends with you?" 73% of the children and adolescents with SCD in this study stated that this was "never" the case. The proportions of children with epilepsy and stuttering who gave the same answer are 66% [11] and 65% [12] respectively. Similarly, in response to the question, "Do you think that having Sickle Cell affects whether people like you or not?" 84% of the children and adolescents with SCD stated that this was "never" the case. The proportions for children with epilepsy and stuttering are 60% and 63% respectively. These comparisons show that children

and adolescents with SCD in this study consistently endorsed less perception of stigma compared with children with epilepsy or stuttering.

Stigma theory could explain why children and adolescents with SCD in this study reported less self-perceived stigma compared with children with epilepsy and stuttering. Stigma theory suggests that conditions that are more visible tend to be more stigmatizing than those with less visible features. Although SCD, epilepsy and stuttering all have visible features, it could be argued that for most affected persons in developed countries like the UK (with access to advanced medical care), SCD may be less visible than epilepsy and stuttering. While some children with epilepsy may have infrequent seizures, each fit; especially grand mal seizures can be very dramatic and highly visible. Similarly, children with stuttering demonstrate evidence of their difficulty with almost every verbal communication. This suggests that reduced visibility may help children and adolescents with SCD not perceive as much stigma as children with epilepsy or stuttering.

LIMITATIONS

Our findings should be interpreted in the light of some limitations. First, we did not have data on the different SCD types, which are usually associated with the variability in the clinical course and severity. Secondly, socially desirable responding was not assessed but may have contributed to the lower prevalence of self-perceived stigma reported in the study. Third, the cross-sectional design allows only a speculative causal sequence. Fourth, the recruitment process did not allow for calculation of a response rate. Finally, the study was conducted in the UK; therefore these findings may not apply to other countries, where health systems and management of SCD are different.

CONCLUSION

The findings of this study provide partial support for the application of stigma dimensions to SCD. Self-perceived stigma in SCD has the potential to become self-fulfilling whereby the fear of discrimination leads the young person to avoid peers thereby missing opportunities to test out whether in fact their fears of discrimination would have happened. For this reason, it is possible that self-perceived stigma could be reduced with interventions such as cognitive behavioural therapy [18].

In addition, given that some self-perceived stigma may be driven by actual experience of enacted stigma caused by discriminatory behaviour by other people, there is need for public education to improve understanding and attitudes towards SCD. However, it is increasingly being recognised that public education alone may be insufficient to alter public attitudes; hence additional institutional and statutory policies against discriminatory behaviour towards people with SCD may be required to foster change.

ACKNOWLEDGEMENTS

We are extremely grateful to the children and adolescents who participated, and their parents for giving consent for the children to take part. We express our appreciation for funding from The Sir Jules Thorn Charitable Trust, and the Sickle Cell Society for their kind assistance. We acknowledge Sonia Patel, Ursula Johnson, and Phil Daly for patient recruitment and data collection. We are also thankful to Professor Irene Roberts for her support.

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