

# Comparative assessment of quality of life among adolescents with sickle cell disease and sickle cell trait: evidence from Odisha, India

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## Research Article

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# Abstract

The present study aims to assess the quality of life (QOL) of adolescents with sickle cell disease (SCD) and sickle cell trait (SCT) in hard-to-reach regions in Koraput district of Odisha state. 387 adolescents with sickle cell genes (HbSS = 52, HbAS = 135, HbA = 200) were selected through their medical records from southern parts of Odisha. To assess the QOL, a validated and pretested QOL scale was modified. The questionnaires were modified by aiming to describe the proportion of adolescents who feel restricted in different domains along with measuring the extent within each domain or finding an aggregate score of QOL. Furthermore, to explore the expenditure on health, 552 households were selected randomly, of which 72 families had HbS individuals. This study found a significantly lower health-related quality of life (HRQOL) in adolescents with SCD. However, most psychosocial sub-domains, for instance, worry about the illness, frequency of angry days, feeling jealousy toward other normal adolescents, and negative feelings of sadness on some days, are similarly affected in adolescents with SCT and SCD. The overall QOL of SCD individuals is more affected (percentage of affected mean score = 60.93%), followed by SCT individuals (35.63%). Healthy adolescents' QOL is relatively unaffected (13% were affected). The yearly frequency of blood transfusion received ( $1.7 \pm 0.4$ ) and hospitalization ( $2.1 \pm 0.9$ ) was significantly higher in adolescents with SCD. The healthcare expenditure was significantly higher (3.6–81.3% of the family income) in families with HbS than in families without HbS (0.8–19.2%) ( $p < 0.05$ ). The overall QOL was affected in both SCD and SCT adolescents. The focus should be given equally to both SCD and SCT individuals, in spite of only SCD individuals.

## Introduction

Sickle cell hemoglobinopathy includes all the abnormal hemoglobin S (HbS), i.e., sickle cell trait (HbAS), sickle cell disease (HbSS), and an array of miscellaneous heterozygous hemoglobinopathies such as hemoglobin SC disease, hemoglobin SD disease, hemoglobin SO Arab disease, and HbS combined with beta-thalassemia (Hb S/ $\beta$  Th). Around 300 million sickle cell carrier individuals are present worldwide, with concentrations in Africa, the Arab Peninsula, India, the Mediterranean, and the southern United States (Key et al. 2015). The sickle gene is prevalent in numerous tribal communities in India, with a prevalence of 1 to 40 % (Colah et al. 2015), who have a high frequency of socioeconomic disadvantage and are medically underserved (Bindani et al. 2020). SCD has high degrees of comorbidities and mortality and Quality of life (QOL) among adolescents having SCD has not been broadly reported. Furthermore, very few studies were reported concerning the QOL among adolescents with SCT which is relatively asymptomatic, which is central to the present study.

Though it is well established that Odisha is at higher risk for sickle cell hemoglobinopathies, there is a lack of an appropriate approach to initiate a large-scale population-based study that covers all the sickle cell prevalent areas (Bindani et al. 2020; Mohanty & Das 2011). Koraput district of Odisha state, where the present study was conducted, is one of the underdeveloped districts and predominantly inhabited by tribal and scheduled caste people, who have a high dominance of socioeconomic disadvantage and are often medically underprivileged (Bindhani & Nayak 2018). There is no published in-depth population-

based study assessing their quality of life. Also, not a single study was found that focuses on adolescents with sickle cell hemoglobin.

Thus, the present study aims to assess the QOL by exploring specific domains that are most affected in adolescents with sickle cell disease (SCD) and sickle cell trait (SCT) in hard-to-reach regions in Koraput district of Odisha.

## Materials and Methods

The present retrospective cross-sectional study was carried out in Koraput district of Odisha, India. The snowball sampling technique was used with the help of villagers, Anganwadi and health personnel to find villages having a high number of individuals with sickle cell genes. Villages (n= 26), those were remotely situated and where healthcare facilities were not easily accessible, were finally picked up for this study. From these villages, a total of 387 adolescents of both sexes in the age group of 10-17 years were selected from their medical/caregiver records. Out of the 387 recruited samples, 52 had sickle cell disease (SCD), and 135 had SCT. Additionally, 200 age-sex-community-matched normal controls were interviewed to compare their QOL to the adolescents with SCD and SCT.

To assess QOL, a QOL scale was modified, which Patel and Pathan developed based on the multidimensional conceptual framework of QOL by Patrick and Erickson, EUROQOL, and WHO QOL BREF (Patel & Pathan 2005). Further, this scale was empirically validated after a pilot study. This study aimed to describe the proportion of adolescents who feel restricted in different domains and measure the extent within each domain or have an aggregate score of quality of life. Therefore, this validated scale could help to enhance the quality of the study. Adolescents in the age group of 10-17 years old are better at responding to the questionnaire. Also, necessary help was taken from the parents of the subjects during the data collection.

Moreover, to explore the expenditure on health, 552 households were selected randomly, of which 72 households had HbS individuals. Demographic information on adolescents was recorded. The data analysis was done using SPSS-22.

## Results

Of the 387 recruited adolescents, 173 (44.7%) were males and 214 (55.3%) were females. Further, 52 individuals were with SCD (HbSS), 135 were with SCT (HbAS) and 200 were without sickle cell hemoglobin or controls (HbA). The mean age ( $\pm$ SD) was found to be  $15.2\pm 2.8$ . The frequency of blood transfusion received ( $1.7\pm 0.4$ ) was significantly higher in adolescents with SCD than in SCT ( $0.07\pm 0.3$ ) and healthy controls (0) during the last 1 year from the day of the survey. The notable factor here is that individuals with SCT were found to have undergone blood transfusions in some instances. Furthermore, the frequency of hospitalization ( $2.1\pm 0.9$ ) was found to be significantly higher among adolescents having SCD (Table 1).

Figure 1 represents the overall QOL score of respondents with HbSS, HbAS and HbAA, respectively. The higher score represents a better quality of life. The QOL of HbSS individuals was found to be highly affected, followed by that of HbAS individuals. The QOL was better among the individuals without HbS. These scores have significantly differed among the three categories. Analysing the various domains (physical function, psychological function, social function, morbidities and opportunities), revealed that the QOL of all these domains were affected (Figure 2). Further, the QOL in each domain significantly differed among the HbSS, HbAS, and HbAA individuals. Interestingly, the control group, consisting of HbA individuals, was categorized based on the presence of family members with HbS. It was found that the quality of life (QOL) of HbA individuals who have family members with HbS was significantly lower compared to HbA individuals without HbS family members ( $p < 0.05$ ) (Figure 3).

Tables 2 to 7 depict the differences in quality of life among adolescents with SCD, SCT and normal controls in various domains like physical, psychological, cognitive, and social functions, along with their health, comorbidities, and perceptions of opportunities. As expected, all the domains were relatively unaffected in normal adolescents compared to adolescents with sickle cell hemoglobin (HbS). Most of the domains (physical, psychological, cognitive, and social functions) were affected in adolescents with SCD followed by SCT. However, most psychosocial sub-domains, for instance, worry about the illness, frequency of angry days, feeling jealousy to other normal adolescents, and negative feelings of sadness on some days, are similarly affected in adolescents with SCT and SCD. Further, the study reported that adolescents with both SCD and SCT were getting equal support from their teachers and friends. In the domain of health perception, it is noteworthy that adolescents with SCT experienced the common cold more frequently (62.96%) compared to their counterparts with SCD, who had a lower incidence rate of about 32.69%. In the sub-domain of feeling sad, both the groups (SCD and SCT) felt that they are the economic burdens on their families because of extensive hospitalization and health care services, which is another root cause for their sadness and depression. Apart from this, the study found breathing problems and weakness among most respondents having HbS in the studied area. The intensity of pain was significantly more severe in individuals with SCD than in SCT. As expected, the QOL is notably more affected in individuals with SCD. Nevertheless, there's also a distinct impact on the overall QOL among those with SCT to some degree.

The annual healthcare cost in households with HbS was significantly higher (3.6% to 81.3% of the total family income) than in households without HbS (0.8% to 19.2%). The families with only sickle cell carriers had lower spending than families with sickle cell disease. 8.33% of the households having HbS had more than 40% healthcare cost, whereas no households without HbS had more than 40% healthcare cost (Table 8). The annual healthcare cost was significantly higher among the households with HbS individuals ( $5182.5 \pm 1124$ ) than the households without HbS ( $911.4 \pm 315.8$ ).

More importantly, the correlation analysis revealed a positive correlation between QOL and annual income, ageing, pain frequency, and comorbidities ( $p < 0.05$ ).

## Discussion

The present study aims to assess the quality of life (QOL) by exploring specific domains most affected in adolescents with SCD and SCT. Several published studies exist on the QOL of individuals affected by SCD (Goldstein-Leever et al. 2020; Bindhani & Nayak 2018; Dale et al. 2011). Most of these studies reported on specific domains reporting average scores of QOL. Furthermore, the focus has mostly been on individuals with SCD, assuming the benign nature of SCT. However, the present study included adolescents with both SCD and SCT to assess their quality of life.

This study found a significantly lower health-related quality of life (HRQOL) in adolescents with sickle cell genes (HbSS and HbAS) than in healthy adolescents (HbAA), which is similar to the findings of previous studies (Ferro et al. 2021; Hilliard et al. 2020; Dale et al. 2011; Gold et al. 2009; Speechley et al. 2006; Shankar et al. 2005). Individuals with SCT are considered relatively asymptomatic. However, this study revealed that many psychosocial subdomains, for instance, worry about their illness, frequency of angry days, feeling jealousy toward other normal adolescents, and a negative feeling of sadness on some days, were affected almost equally in adolescents having SCT and SCD. This corroborates with a similar study by Patel & Pathan (2005). Also, both SCD and SCT adolescents reported that they were economic burdens on their families because of the extensive hospitalization and health care maintenance, which is another root cause for sadness and depression among the economically poor respondents. This finding is similar to a previous study (Bindhani 2016). The intensity of pain was significantly more severe in SCD than in SCT, which corroborates previous research (Ashorobi et al. 2019; McClish et al. 2005). Further, adolescents with SCT experienced the common cold more frequently (62.96%) compared to their counterparts with SCD, who had a lower incidence rate of about 32.69%.

The frequency of blood transfusion received (1.7) was significantly higher in adolescents with SCD than in SCT. However, to the best of our knowledge, no study has reported blood transfusion among sickle cell carriers (SCT). Furthermore, the frequency of hospitalization (2.1) was higher in subjects with SCD than in SCT. Both the frequencies of blood transfusion and hospitalization among SCD-affected individuals were higher in this study compared to the findings of a previous similar study (Patel & Pathan 2005). More importantly, the frequency of blood transfusion and hospitalization were considerably higher among the studied SCT subjects as compared to the previous study (Patel & Pathan 2005). This study explored the overall quality of life (QOL) score by grouping participants into sickle cell patients, sickle cell carriers, controls with HbS family member(s), and controls without HbS family member(s) for a grassroots-level analysis. Furthermore, the present study reported that breathing problems are common among individuals with SCT, which might have occurred as they generally worked at high altitudes and walked a long distance for schooling. However, no previous study was found related to this type of comorbidities among SCT adolescents.

The percentage of annual household income spent on healthcare overheads ranged from 3.6% to 81.3% in families with sickle cell hemoglobinopathies. In contrast, the expenditure on health in families without sickle cell hemoglobinopathies was 0.8% to 19.2% of their family income. Though previous studies reported a high healthcare expenditure rate in families with sickle cell patients, this was relatively lower compared to the present study (Ngolet et al. 2016; Olatunja et al. 2015). On the other hand, this

proportion was more affected in the studied area due to their comparatively lower family income. The substantial difference in healthcare costs among families with sickle cell carriers was observed because families with only sickle cell carriers had lower expenditures compared to families with sickle cell disease. Although the government of Odisha recently implemented a scheme providing 500 rupees for each individual with sickle cell disease, it is negligible compared to their healthcare costs (Bindhani et al. 2021). Some families' healthcare expenditures were out of pocket. In other words, their healthcare care cost exceeded their total monthly income. A similar research described that in most developing countries, the healthcare cost of households with sickle cell hemoglobinopathies is mainly paid through out-of-pocket expenditure (Olatunya et al. 2015). For instance, during the household survey, a family was found where a widow woman had two children; one child had SCD, and the other one had SCT. The mother was also a carrier of HbS with associated comorbidities. Their monthly income was nearly only 1000 Indian rupees, and many times, this amount was very little to treat themselves. Also, many families were found to have the same status in the studied area. Since sickle cell carriers are associated with comorbidities that increase with age, the quality of life of older individuals may be more affected. Therefore, further studies are needed to explore this relationship in more detail.

In sum, the overall QOL was affected more in adolescents with SCD than in SCT. However, many domains, like psychosocial functions, are equally affected in both SCD and SCT individuals. The studied population's low socioeconomic and educational status is more like an extra burden in their QOL. Interventions to improve QOL should target the affected domains. Enhancing healthcare facilities and awareness of the disease and its manifestation will help to improve the QOL in adolescents with SCT. Community-based awareness and interventional programs should be implemented to elevate the quality of life in individuals with SCD and SCT.

## **Declarations**

The authors declare that no funds, grants, or other support were received during the preparation of this manuscript.

### **Conflict of interest**

The authors declare no competing interests.

### **Ethics approval**

The work was approved by the Ethical Committee, Department of Anthropology, Central University of Odisha.

### **Consent to participate**

Informed consent was obtained from all patients for being included in the study.

### **Consent for publication**

All authors agreed with the content and gave explicit consent to submit.

### **Availability of Data and Materials**

Not Applicable.

### **Code Availability**

Not Applicable.

### **Author Contributions**

All authors contributed to the study conception and design. Material preparation, data collection, and analysis were performed by BKB. The first draft of the manuscript was written by BKB. JKN reviewed it thoroughly and incorporated his input. All authors read and approved the final manuscript.

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## Tables



Table 1

Comparison of sociodemographic and medical history information among individuals with SCD, SCT, and normal status (n = 387)

| Characteristics                               | SCD (n = 52)          | SCT (n = 135)         | Normal/HbA (n = 200)  | p-value |
|---|-----------------------|-----------------------|-----------------------|---------|
| Male (No., %)                                 | 24 (46.15)            | 58 (42.96)            | 91 (45.5)             | 0.878   |
| Female  | 28 (53.85)            | 77 (57.04)            | 109 (54.5)            |         |
| Age (mean $\pm$ SD) (in years)                | 14.7 $\pm$ 2.4        | 15.1 $\pm$ 2.6        | 15.8 $\pm$ 3.5        | 0.891   |
| Educational level (mean $\pm$ SD)             | 5.2 $\pm$ 2.1         | 6.4 $\pm$ 2.4         | 6.7 $\pm$ 2.5         | 0.345   |
| Annual family income (mean $\pm$ SD) (in INR) | 62035.1 $\pm$ 16792.9 | 62642.3 $\pm$ 15163.8 | 63173.9 $\pm$ 18792.1 | 0.427   |
| Medical history                               |                       |                       |                       |         |
| No. of times hospitalized (in last 1 year)    | 2.1 $\pm$ 0.9         | 0.8 $\pm$ 0.4         | 0.3 $\pm$ 0.3         | 0.03    |
| Transfusion received (in last 1 year)         | 1.7 $\pm$ 0.4         | 0.07 $\pm$ 0.3        | 0                     | < 0.001 |

Table 2  
Adolescents' responses for different items of physical function

| Physical function           | HbSS |       | HbAS |       | HbA |       |         |
|-----------------------------|------|-------|------|-------|-----|-------|---------|
|                             | No.  | %     | No.  | %     | No. | %     |         |
| <b>Mobility</b>             |      |       |      |       |     |       |         |
| No problem                  | 36   | 69.23 | 124  | 91.85 | 191 | 95.50 | < 0.001 |
| Problem on some days        | 11   | 21.15 | 11   | 8.15  | 9   | 4.50  |         |
| Unable to do on most days   | 5    | 9.62  | 0    | 0.00  | 0   | 0.00  |         |
| <b>Indoor games</b>         |      |       |      |       |     |       |         |
| No problem                  | 31   | 59.62 | 102  | 75.56 | 193 | 96.50 | < 0.001 |
| Problem on some days        | 14   | 26.92 | 33   | 24.44 | 7   | 3.50  |         |
| Unable to do on most days   | 7    | 13.46 | 0    | 0.00  | 0   | 0.00  |         |
| <b>Attendance in school</b> |      |       |      |       |     |       |         |
| No problem                  | 10   | 19.23 | 68   | 50.37 | 180 | 90.00 | < 0.001 |
| Problem on some days        | 27   | 51.92 | 45   | 33.33 | 19  | 9.50  |         |
| Unable to do on most days   | 15   | 28.85 | 22   | 16.30 | 1   | 0.50  |         |
| <b>Outdoor games</b>        |      |       |      |       |     |       |         |
| No problem                  | 8    | 15.38 | 79   | 58.52 | 188 | 94.00 | < 0.001 |
| Problem on some days        | 23   | 44.23 | 50   | 37.04 | 11  | 5.50  |         |
| Unable to do on most days   | 21   | 40.38 | 6    | 4.44  | 1   | 0.50  |         |

Table 3  
Adolescents' responses for different sub-domains of psychological function

| Psychological function        | HbSS |       | HbAS |       | HbA |       |         |
|-------------------------------|------|-------|------|-------|-----|-------|---------|
|                               | No.  | %     | No.  | No.   | %   | No.   |         |
| <b>Disinterest</b>            |      |       |      |       |     |       |         |
| On most days, I do not        | 27   | 51.92 | 128  | 94.81 | 185 | 92.50 | < 0.001 |
| On some days, I do            | 15   | 28.85 | 7    | 5.19  | 15  | 7.50  |         |
| On most days, I do            | 10   | 19.23 | 0    | 0.00  | 0   | 0.00  |         |
| <b>Worry</b>                  |      |       |      |       |     |       |         |
| On most days, I do not        | 27   | 51.92 | 73   | 54.07 | 194 | 97.00 | < 0.001 |
| On some days, I do            | 11   | 21.15 | 45   | 33.33 | 6   | 3.00  |         |
| On most days, I do            | 14   | 26.92 | 17   | 12.59 | 0   | 0.00  |         |
| <b>Look Perception</b>        |      |       |      |       |     |       |         |
| I don't feel I look different | 16   | 30.77 | 80   | 59.26 | 178 | 89.00 | < 0.001 |
| I look different              | 23   | 44.23 | 21   | 15.56 | 11  | 5.50  |         |
| I have not though about it    | 13   | 25.00 | 34   | 25.19 | 11  | 5.50  |         |
| <b>Feeling sad</b>            |      |       |      |       |     |       |         |
| On most days, I do not        | 27   | 51.92 | 90   | 66.67 | 197 | 98.50 | < 0.001 |
| On some days, I do            | 13   | 25.00 | 40   | 29.63 | 3   | 1.50  |         |
| On most days, I do            | 12   | 23.08 | 5    | 3.70  | 0   | 0.00  |         |
| <b>Jealousy</b>               |      |       |      |       |     |       |         |
| On most, I do not             | 39   | 75.00 | 101  | 74.81 | 182 | 91.00 | < 0.001 |
| On some days, I do            | 9    | 17.31 | 34   | 25.19 | 17  | 8.50  |         |
| On most days, I do            | 4    | 7.69  | 0    | 0.00  | 1   | 0.50  |         |

Table 4  
Responses of the subjects for different factors of social function

| Social function                             | HbSS |       | HbAS |       | HbA |       |         |
|---|------|-------|------|-------|-----|-------|---------|
|   | No.  | %     | No.  | %     | No. | %     |         |
| <b>Entertainment</b>                        |      |       |      |       |     |       |         |
| On most days, not affected                  | 33   | 63.46 | 96   | 71.11 | 187 | 93.50 | < 0.001 |
| Affected on some days                       | 10   | 19.23 | 27   | 20.00 | 12  | 6.00  |         |
| Affected on most days                       | 9    | 17.31 | 12   | 8.89  | 1   | 0.50  |         |
| <b>Teachers support for problems</b>        |      |       |      |       |     |       |         |
| Always supportive                           | 39   | 75.00 | 115  | 85.19 | 179 | 89.50 | < 0.001 |
| Supportive on some days                     | 12   | 23.08 | 19   | 14.07 | 20  | 10.00 |         |
| On most days not supportive                 | 1    | 1.92  | 1    | 0.74  | 1   | 0.50  |         |
| <b>Acceptability by friends</b>             |      |       |      |       |     |       |         |
| On most days, not affected                  | 43   | 82.69 | 119  | 88.15 | 195 | 97.50 | < 0.001 |
| Affected on some days                       | 7    | 13.46 | 16   | 11.85 | 5   | 2.50  |         |
| Affected on most days                       | 2    | 3.85  | 0    | 0.00  | 0   | 0.00  |         |
| <b>Participation in cultural activities</b> |      |       |      |       |     |       |         |
| Can on most days                            | 25   | 48.08 | 112  | 82.96 | 194 | 97.00 | < 0.001 |
| Can on some days                            | 9    | 17.31 | 21   | 15.56 | 5   | 2.50  |         |
| Unable to do on most days                   | 18   | 34.62 | 2    | 1.48  | 1   | 0.50  |         |

Table 5

Respondent's reaction to different items of health perception (collected only from the individuals with SCD and SCT)

| Health perception             | SCD |       | SCT |       |         |
|-------------------------------|-----|-------|-----|-------|---------|
|                               | No. | %     | No. | %     |         |
| <b>Seriousness of illness</b> |     |       |     |       |         |
| Like a common cold            | 17  | 32.69 | 85  | 62.96 | < 0.001 |
| More than a common cold       | 17  | 32.69 | 40  | 29.63 |         |
| It is a severe illness        | 18  | 34.62 | 10  | 7.41  |         |

Table 6  
Adolescents' responses for different items of comorbidity

| Morbidity                        | HbSS |       | HbAS |       | HbA |       |         |
|----------------------------------|------|-------|------|-------|-----|-------|---------|
|                                  | No.  | %     | No.  | No.   | %   | No.   |         |
| <b>Pain frequency</b>            |      |       |      |       |     |       |         |
| Never in pain                    | 2    | 3.85  | 104  | 77.04 | 180 | 90.00 | < 0.001 |
| Sometimes in pain                | 36   | 69.23 | 27   | 20.00 | 20  | 10.00 |         |
| Always(most of the time) in pain | 14   | 26.92 | 4    | 2.96  | 0   | 0.00  |         |
| <b>Weakness frequency</b>        |      |       |      |       |     |       |         |
| Never feel tired                 | 3    | 5.77  | 58   | 42.96 | 168 | 84.00 | < 0.001 |
| Sometimes feel tired             | 32   | 61.54 | 68   | 50.37 | 29  | 14.50 |         |
| Always feel tired                | 17   | 32.69 | 9    | 6.67  | 3   | 1.50  |         |
| <b>Pain intensity reaction</b>   |      |       |      |       |     |       |         |
| No sadness                       | 6    | 11.54 | 70   | 51.85 | 195 | 97.50 | < 0.001 |
| Minimum sadness                  | 8    | 15.38 | 40   | 29.63 | 3   | 1.50  |         |
| Some sadness                     | 12   | 23.08 | 25   | 18.52 | 2   | 1.00  |         |
| Cry                              | 13   | 25.00 | 0    | 0.00  | 0   | 0.00  |         |
| Agony                            | 13   | 25.00 | 0    | 0.00  | 0   | 0.00  |         |
| <b>Weakness intensity</b>        |      |       |      |       |     |       |         |
| No weakness                      | 3    | 5.77  | 57   | 42.22 | 152 | 76.00 | < 0.001 |
| Minimum weakness                 | 14   | 26.92 | 42   | 31.11 | 33  | 16.50 |         |
| Some weakness                    | 19   | 36.54 | 34   | 25.19 | 15  | 7.50  |         |
| Cry                              | 16   | 30.77 | 2    | 1.48  | 0   | 0.00  |         |
| Agony                            | 0    | 0.00  | 0    | 0.00  | 0   | 0.00  |         |

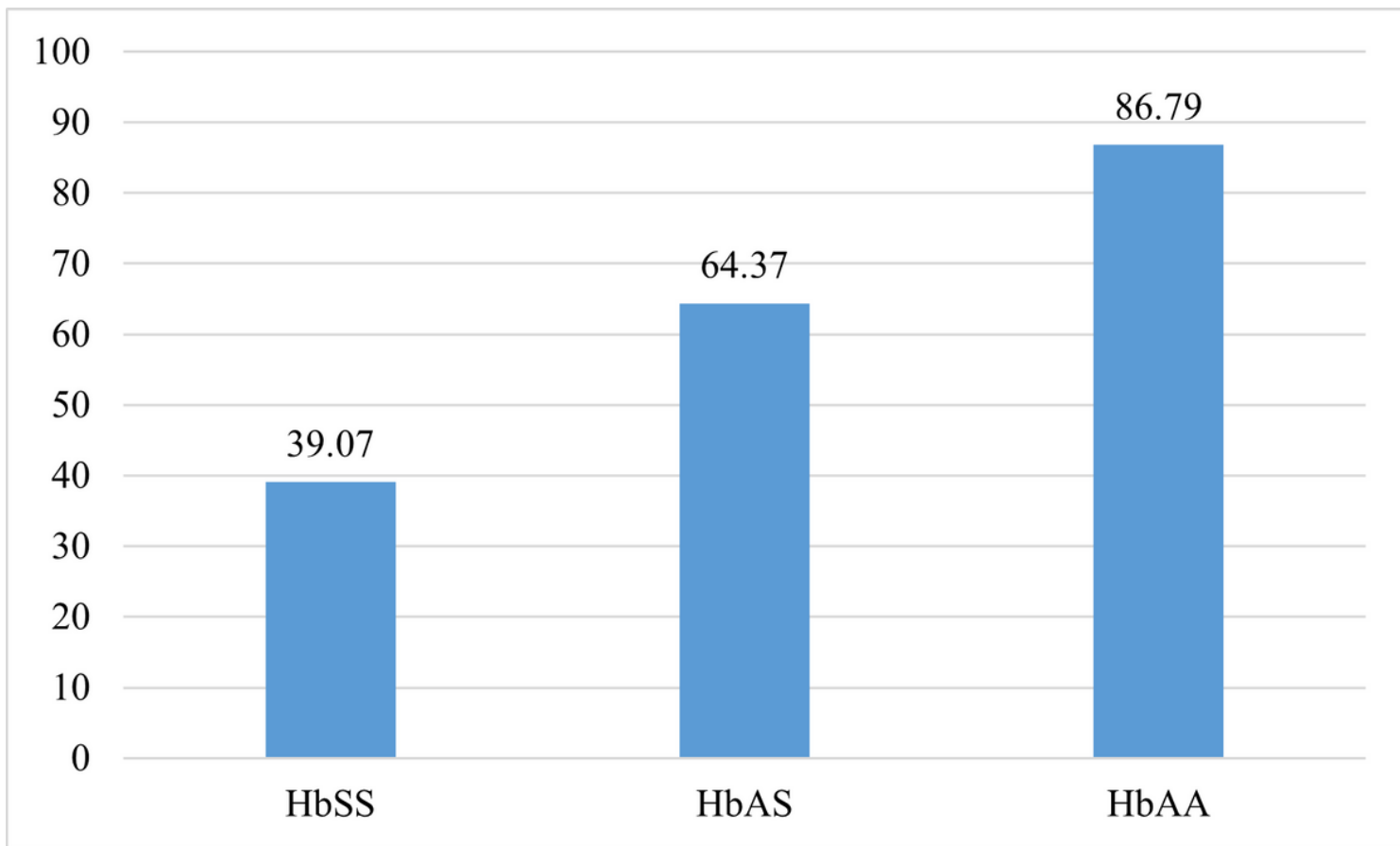
Table 7  
Adolescents' responses for different factors of opportunity

| Opportunity                              | HbSS |       | HbAS |       | HbA |       |         |
|--|------|-------|------|-------|-----|-------|---------|
|  | No.  | %     | No.  | No.   | %   |       |         |
| <b>Optimism in achieving success (O)</b> |      |       |      |       |     |       |         |
| Optimistic in achieving like peers       | 11   | 21.15 | 75   | 55.56 | 160 | 80.00 | < 0.001 |
| Can achieve with difficulty              | 30   | 57.69 | 57   | 42.22 | 37  | 18.50 |         |
| Will not be able to achieve              | 11   | 21.15 | 3    | 2.22  | 3   | 1.50  |         |

Table 8  
Healthcare cost of families with HbA and without HbS

| Healthcare expenditure (% of the family income) | No. of households without HbS (n = 482) |       | No. of households with HbS (n = 70) |       | p-value |
|---|---|-------|-------------------------------------|-------|---------|
|   | No.                                     | %     | No.                                 | %     |         |
| < 10%   | 351                                     | 73.13 | 28                                  | 38.89 | < 0.001 |
| 10% to < 20%                                    | 115                                     | 23.96 | 21                                  | 29.17 |         |
| 20% to < 30%                                    | 12                                      | 2.50  | 8                                   | 11.11 |         |
| 30% to < 40%                                    | 2                                       | 0.42  | 9                                   | 12.50 |         |
| > 40%   | 0                                       | 0.00  | 6                                   | 8.33  |         |
| Mean healthcare cost (Annually)                 | 911.4 ± 315.8                           |       | 5182.5 ± 1124                       |       | < 0.001 |

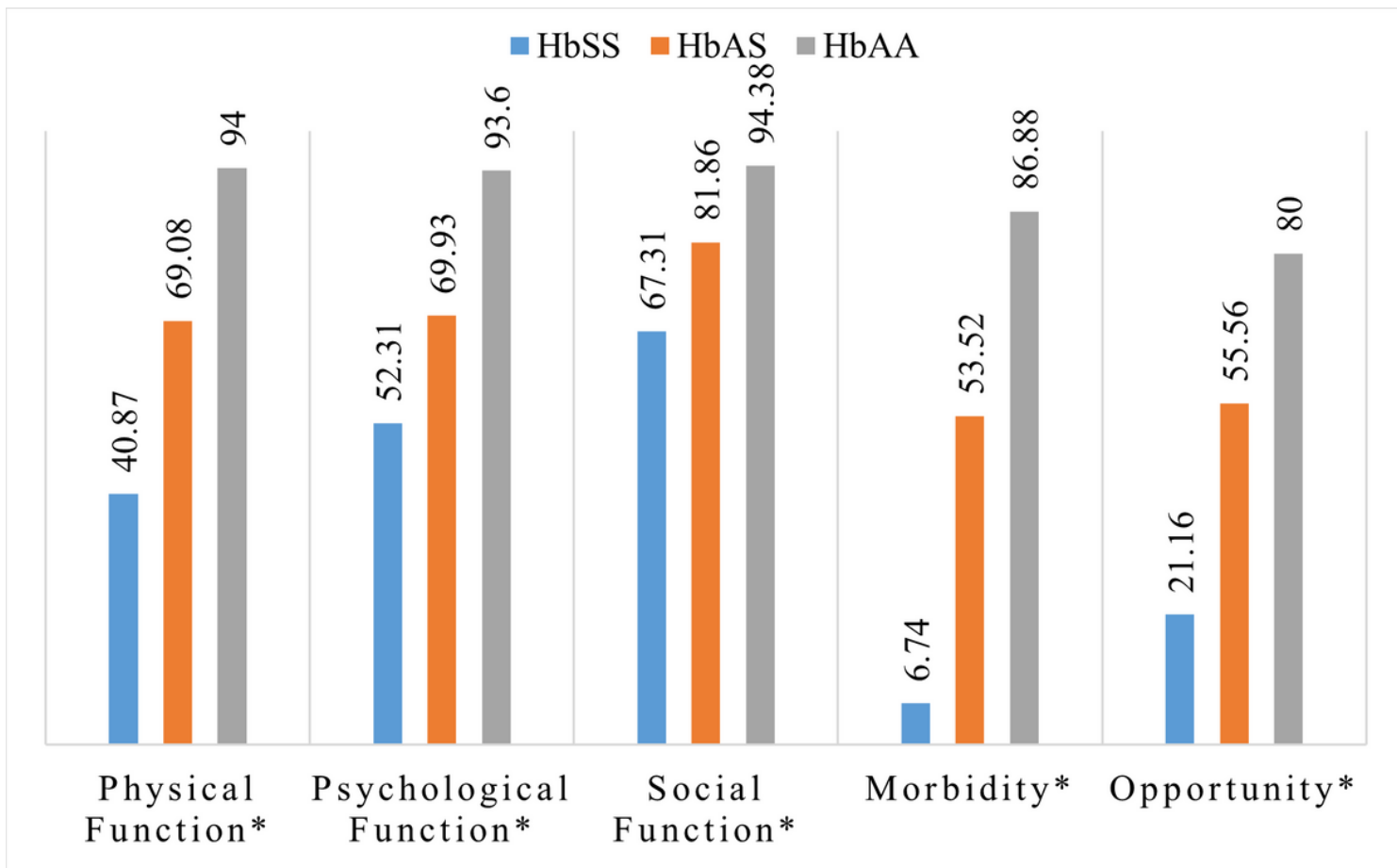
## Figures



**Figure 1**

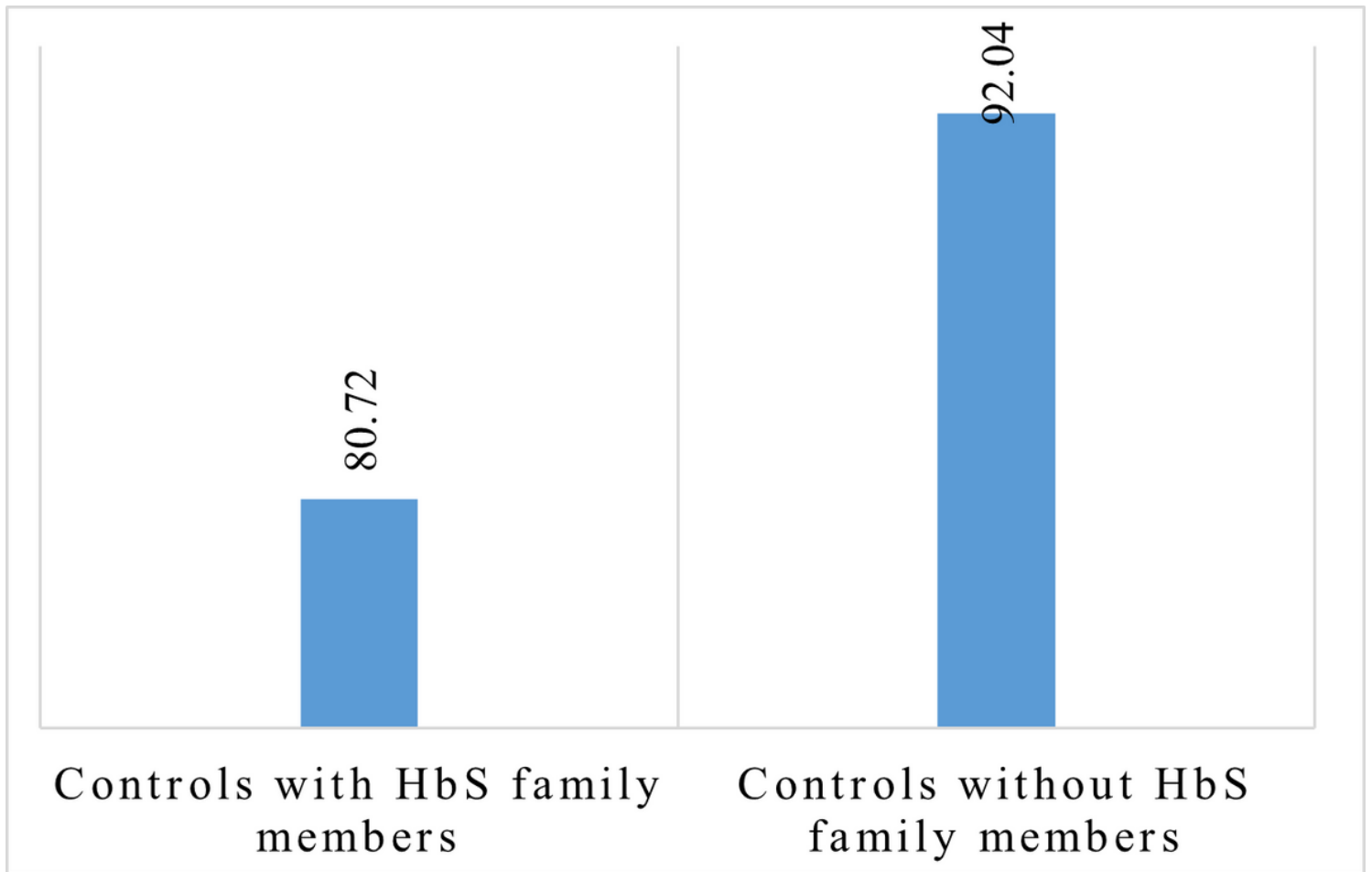
Comparisons of overall QOL score of respondents with HbSS, HbAS and HbAA (p value <0.05)





**Figure 2**

Domain wise comparison of QOL (\*p<0.05)



**Figure 3**

Comparison of QOL score among normal individuals (HbA) with HbS family members and without family members ( $p < 0.05$ )