Research Article

Lung Function Tests in Sickle-Cell Patients in Benin City

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Background. Sickle-cell disease (SCD) is a life-long haematological disorder characterized by red blood cells that assume an abnormal, rigid, sickle shape and with high prevalence in West Africa. Sickle cell disease (SCD) is one of the most common genetic disorders among Africans as indicated in Akinyanju (1989). 61.4% of the children born annually with haemoglobinopathies worldwide have been reported to have SCD and they are from Africa as shown in World Health Organization Report (1987). Hypoxaemia is the hallmark of pulmonary abnormality in SCD patients of all age groups. This was first documented by Klinefelter (1942), and has since been corroborated by other workers. The hypoxaemia of stable SCD patients is said to result from the combined effects of perfusion and diffusion defect.

Materials and Methods. This study was carried out to determine the changes in lung functions in relation to gender, age, and body mass index (BMI) of sickle-cell patients in a stable state. 60 subjects made up of 30 patients and 30 control groups were used in this study. Both test and control groups were 13 men and 17 women, aged 19–35 years. Lung Function Test was done with the aid of a digital spirometer (Spirolab II, Italy). Weight was measured using Avery scales (Avery Berkel, 2003, UK). Height was measured using a wall-mounted stadiometer. Data were collected and analyzed using Student’s t-test and Pearson correlation. P < 0.05 was statistically significant.

Results. The results revealed that adults with SCD had a significantly lower mean forced expiratory volume in one second (FEV1) and forced vital capacity (FVC) compared to the control. The lung function indices were lower in females than males in the sickle cell patients. The FVC, FEV1, and FEV1/FVC correlated positively with BMI statistically in the patients. The lung function declined with age. This work has also shown that the most common pulmonary function test (PFT) abnormality was restrictive disease pattern (76.7%).

Conclusion. The result of this work suggested that lung function differs significantly in subjects with SCD compared with matched controls of a similar age and gender.

1. Introduction

Sickle cell-disease (SCD) is one of the most common genetic disorders among Africans. 61.4% of all children born annually with haemoglobinopathies worldwide have SCD and are from Africa [1].

Sickle-cell disease is a life-long haematological disorder characterized by red blood cells that assume an abnormal, rigid, and sickle shape [2]. The sickling occurs because of a mutation in the hemoglobin gene. Life expectancy is shortened, with studies reporting an average life expectancy of 42 and 48 years for males and females, respectively [3]. Hypoxaemia which is the hallmark of pulmonary abnormality in SCA patients of all age groups was first documented by Klinefelter (1942) and has since been corroborated by other workers [4–6].

Platt et al. [3] reported that acute pulmonary complications, such as acute chest syndrome (ACS), predominate in children, while chronic lung disease is more common in adults with SCD.

Studies of lung function in SCD have demonstrated a restrictive defect [7] and a reduction in total lung capacity (TLC) of 50% has been reported in advanced forms.

During steady-state sickle-cell disease, the major abnormality in pulmonary function is a restrictive ventilatory impairment, characterized by a mild reduction in total lung capacity, and reduced diffusion capacity for carbon monoxide [8]. These abnormalities worsen with age and are associated with increases in pulmonary-artery pressures [9].

The hypoxaemia of stable SCD patients is said to result from the combined effects of both a perfusion and diffusion defect.
VanderJagt et al. concluded that pulmonary function is reduced in Nigerian adults with SCD compared to controls and that for both groups, pulmonary function is directly related to body composition [10].

Lung function tests (also called pulmonary function tests) are useful in assessing the functional status of the respiratory system both in physiological and pathological conditions. Spirometry is also the gold standard for the diagnosis, assessment, and monitoring of COPD [11]. Spirometry is a medical test that measures the volume of air an individual inhales or exhales as a function of time [12].

This study was carried out to determine the changes in lung functions in relation to gender, age, and body mass index levels of sickle-cell patients in a stable state. The objective were to determine the difference in lung volumes between male and female sickle-cell patients lung values, to determine the relationship of body mass index with lung function of sickle-cell patients and to compare the lung function values of sickle cell patients with those of nonsicklers.

It became necessary to carry out this study on sickle-cell patients in Benin City as no such study has been carried out in this region.

2. Materials and Methods

2.1. Study Population. 60 subjects made up of 30 SCD patients and 30 control groups were used in this study. Both test and control groups were 13 men and 17 women respectively, aged 19–35 years. All test subjects were persons with haemoglobin SS were in a stable state at the time of study. The test subjects were known and diagnostically confirmed cases of sickle-cell disease who attend consultant outpatient clinic of University of Benin Teaching Hospital. The control group was healthy hemoglobin AA subjects, selected amongst student nurses of University of Benin Teaching Hospital and lacked previous history of cardiopulmonary disease. The tests were carried out after verbal consent was obtained from the subjects following a detailed information and explanation. Each subject was instructed to sign a consent form after a detailed explanation. Each subject had at least three tests trials before the performance. Values for each subject were obtained from the mean of three satisfactory performances.

2.2. Exclusion Criteria. Subjects who had difficulty standing up straight for accurate measurement as a result of disease process or weakness were excluded. Subjects who had crisis within 12 weeks as at the time of study were also excluded.

2.3. Questionnaire. Well-structured questionnaire to access the lifestyle of the subjects (tests and control) was administered. The drinking and smoking habits were established. The questionnaire also probed specifically into respiratory symptoms associated with irritation of the respiratory tract such as coughing, catarrh, and difficulty in breathing.

2.4. Lung Function Test. This was done with the aid of a digital spirometer (Spirolab II, Italy). The forced expiratory volume in one second (FEV₁), forced vital capacity (FVC), and ratio of FEV₁/FVC were measured.

Test results are given as measured values in litres and as percent’s of predicted values using (American Thoracic Society Standardization of spirometry, 1994) customized in the spirometer.

Each subject was made to sit down upright and erect. The subject then held the mouth piece attached to the spirometer and placed his mouth around the opening of the mouth piece. Care was taken that he did not to block it with the tongue and that the mouth completely went tightly round the opening of the mouth piece to prevent leakage of air. The subject was then asked to inspire maximally and then expire forcibly at once through the mouth piece into the spirometer. Each subject had at least three tests trials before the performance. Values for each subject were obtained from the mean of three satisfactory performances.

2.5. Measurement of Weight. Weight was measured in kilograms using Avery scales (Avery Berkel, 2003, UK). The subjects were made to remove their shoes and any piece of clothes or objects in their possession or on the person were put aside. The subject was asked to mount the scale and the weight was read off the scale in kilograms.

2.6. Measurement of Height. Standing height was measured in centimeter using a wall mounted stadiometer. Standing height was measured without footwear.

2.7. Measurement of Blood Pressure. Subject was seated on a chair with the back supported and the right arms bared and supported at heart level. Measurement was performed on the subject after five minutes of rest and was taken using a mercury sphygmomanometer applied on the right arm of the subject. First and fifth Korotkov sounds were recorded for systolic and diastolic readings, respectively. Two readings separated by two minutes were averaged.

2.8. Measurement of Respiratory Rate. Subject was made to sit down quietly without any form of agitation. The number of breaths per minute was taken by simple observation of the chest movement during respiration. All measurements were taken approximately between 8.30 am to 1.30 pm on clinic days.

2.9. Statistical Analysis. The data collected was subjected to statistical analysis involving computation of mean and standard deviation; t-test, and Pearson correlation using the Microsoft Office Excel 2007.

Determination of BMI. This was by dividing weight in kilograms by height in meters squared (BMI = Wt in kg/Ht in m²).

3. Results

3.1. Patients Characteristics. A total of 60 adult subjects were used in this study with an average age of 27.6 ± 10.3 yr
The Body Mass Index correlates positively with FVC in test subjects with an upward inclination as shown in Figure 3. Also in Figures 4 and 5 BMI correlates positively with FEV/FVC and FEV in test subjects’ upward inclination.

64% of the test group had tertiary education, 18% had secondary and primary education respectively. All the subjects of the control group were student nurses. No subject had received blood transfusion in the last one year (Table 1).

FVC and FEV₁ declined in test group compared to control group. Also, FVC and FEV₁ values in test group are lower than normal while the control group had normal values. The FEV₁/FVC% values both test and control groups are with normal range (see Table 2 and Figure 1).

The female subjects had significantly lower FVC and FEV₁ values by 7.1% and 8.2% respectively than the males however the FEV₁/FVC value was only 0.1% lower in females than male value. (see Figure 2 and Table 3). The Body Mass Index correlates positively with FVC in test subjects with an upward inclination as shown in Figure 3. Also in Figures 4 and 5 BMI correlates positively with FEV/FVC and FEV in test subjects’ upward inclination.

This work has shown that lung function indices were lower in females than males in sickle-cell subjects. The difference was significant except the FEV₁/FVC ratio. This may be due to the difference in stature and in the level of hemoglobin.

Miller et al. [13] had reported similar result but Fawibe [14], reported a contrary result which are related to lack of significant difference in stature between the male and female SCD subjects.

This work has shown that adults with SCD had a significant lower mean FEV₁, and FVC compared to the control but their FEV₁/FVC ratios were not significantly different from

### Table 1: Demographic and clinical characteristics of test and control group.

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Male</th>
<th>Female</th>
<th>Male</th>
<th>Female</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (yrs)</td>
<td>26 ± 4.3</td>
<td>29 ± 4.8</td>
<td>26 ± 4.3</td>
<td>29 ± 4.8</td>
</tr>
<tr>
<td>Systolic B/P mmHg</td>
<td>130</td>
<td>125</td>
<td>120</td>
<td>115</td>
</tr>
<tr>
<td>Diastolic B/P mmHg</td>
<td>90</td>
<td>90</td>
<td>80</td>
<td>70</td>
</tr>
<tr>
<td>BMI</td>
<td>20.4 ± 2.3</td>
<td>22.1 ± 1.8</td>
<td>22.6 ± 2.4</td>
<td>22.3 ± 3.3</td>
</tr>
<tr>
<td>Height cm</td>
<td>166.6</td>
<td>155.3</td>
<td>175.6</td>
<td>166.8</td>
</tr>
<tr>
<td>Weight kilogram</td>
<td>56.4</td>
<td>53.3</td>
<td>69.7</td>
<td>62</td>
</tr>
<tr>
<td>Respiration</td>
<td>20</td>
<td>22</td>
<td>16</td>
<td>18</td>
</tr>
<tr>
<td>Blood transfusion in the last one year</td>
<td>NIL</td>
<td>NIL</td>
<td>NIL</td>
<td>NIL</td>
</tr>
</tbody>
</table>

### Table 2: Mean lung volume values for control and test subjects (n = 60).

<table>
<thead>
<tr>
<th>Index</th>
<th>Test (n = 30)</th>
<th>Control (n = 30)</th>
</tr>
</thead>
<tbody>
<tr>
<td>FVC (litre)</td>
<td>*2.2 ± 0.4</td>
<td>3.0 ± 0.7</td>
</tr>
<tr>
<td>% Predicted</td>
<td>*74.3 ± 17.1</td>
<td>91.5 ± 12.9</td>
</tr>
<tr>
<td>FEV₁ (litre)</td>
<td>*2.07 ± 0.5</td>
<td>2.6 ± 0.6</td>
</tr>
<tr>
<td>% Predicted</td>
<td>*71.8 ± 17.4</td>
<td>91.4 ± 21.4</td>
</tr>
<tr>
<td>FEV₁/FVC%</td>
<td>**87.9 ± 18.4</td>
<td>89.5 ± 16.8</td>
</tr>
</tbody>
</table>

Results are shown in mean ± SD, *Significance (P < 0.05), **Not Significant.

### Table 3: Mean lung volumes values for male and female test subjects.

<table>
<thead>
<tr>
<th></th>
<th>Male (n = 13)</th>
<th>Female (n = 17)</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>FVC (litre)</td>
<td>*2.9 ± 0.5</td>
<td>2.0 ± 0.4</td>
<td>P &lt; 0.05</td>
</tr>
<tr>
<td>FVC% Predicted</td>
<td>*78.1 ± 10.5</td>
<td>71 ± 20.6</td>
<td>P &lt; 0.05</td>
</tr>
<tr>
<td>FEV₁ (litre)</td>
<td>*2.6 ± 0.3</td>
<td>1.7 ± 0.4</td>
<td>P &lt; 0.05</td>
</tr>
<tr>
<td>FEV₁% Predicted</td>
<td>*78.8 ± 14.6</td>
<td>70.6 ± 20.3</td>
<td>P &lt; 0.05</td>
</tr>
<tr>
<td>FEV₁/FVC%</td>
<td>*88.2 ± 16.4</td>
<td>88.1 ± 20.2</td>
<td>P &lt; 0.05</td>
</tr>
</tbody>
</table>

Results are shown in mean ± SD, *Significance (P < 0.05), **Not Significant.

**Figure 1:** Bar chart showing the mean lung function tests values between the test and control groups.

### 4. Discussion

This work has shown that lung function indices were lower in females than males in sickle-cell subjects. The difference was significant except the FEV₁/FVC ratio. This may be due to the difference in stature and in the level of hemoglobin.
those of the controls. Previous studies have suggested that abnormal pulmonary function tests are the first objective sign of chronic sickle cell lung disease and that they could be helpful in patient management [15–17].

This work has also shown that the most common PFT abnormality was restrictive disease pattern (76.7%). Previously, studies have demonstrated a spectrum of PFT abnormalities in adult sickle-cell disease including restrictive physiology, and decreased obstructive pattern [8, 18–20]. Reason given by these workers [8, 18–20] for the observed impaired ventilatory function in these subjects was a reduction of lung compliance which may be due to repeated episodes of Acute Chest Syndrome or sickle-cell-related vasculopathy. Obstructive disease, was relatively uncommon, occurring in 23.3% of the patients. Santoli et al. [21] observed obstructive defect in ventilatory function in a group of stable SCD patients with recurrent ACS. They noted that the obstructive pattern was accompanied by an increase in diffusing capacity and suggested that it might have been related to an increase in lung blood volume. Similar result was observed by Koumbourlis et al. [5] in a group of subjects with SCA. They suggested that obstructive lung disease possibly precedes the development of restrictive lung disease, and that airway reactivity may be part of the pathogenic mechanism.

Our results also show that both male and female test and control, FVC, FEV1, and FEV1/FVC correlated positively with BMI (P < 0.05). VanderJagt et al. [10] reported that pulmonary function correlated positively with BMI in Nigerian young adults with SCD compared to controls.

In conclusion, lung function differs significantly in subjects with SCD compared with matched controls of a similar age and gender. These results have implications for the timing of commencement of treatment aimed at reducing chronic pulmonary morbidity in patients with SCD.

**Recommendation**

In Nigeria, outline spirometry can be used as a screening test for asymptomatic sickle cell chronic lung disease in the followup of SCD.

**References**


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