

EXERCISE PERFORMANCE IN YOUNG SUBJECTS WITH SICKLE CELL DISEASE

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Abstract

The sickle cell trait (SCT) is a genetic abnormality of red blood cells which mainly affects people in Africa and those of African descent. Its prevalence in West Africa is 10–15 % and the same rate of prevalence is observed in sportsmen and in persons with a sedentary way of life. This haemoglobinopathy has been suspected, by various authors, to decrease exercise performance or even to be a risk factor for various accidents in which an environmental unfavourable condition is involved, such as moderately high altitude or high ambient temperature. We found that performance capability was not different between well trained subjects with SCT and those without it. SCT had no effect on the subject's performance during exhausting, at least when its duration did not exceed half an hour.

Key words

Sickle cell disease, Exercise performance, Hot climate

INTRODUCTION

The sickle cell trait (SCT) is a genetic abnormality of red blood cells which mainly affects people in Africa and those of African descent in many other countries. It is due to a mutation in a parental gene coding for haemoglobin (Hb). In haemoglobin with the sickle cell trait (Hb AS), valine is substituted for glutamic acid in the sixth amino acid of the chain β of globin. In sickle cell disease (SCD), the mutation is present in both parental genes and the diseased individual is homozygous for S (HbSS) (1).

Prevalence of SCT in West Africa is within the range of 8–10 % (2) and the rate of prevalence in sportsmen of these countries is identical to that of the sedentary general population (2,3). The sickle cell trait is usually asymptomatic but the substitution of glutamic acid modifies physiological properties of haemoglobin and produces disorders such as lower solubility of the desoxygenated form, reduction in affinity to for oxygen and, eventually, sickling formation. Physiological changes associated with hard physical activities induce regional hypoxemia, acidosis, dehydration, hyperthermia, all of which enhance the tendency

of abnormal haemoglobin to polymerise and, therefore, could have a deleterious influence on the course of physical activities. It is well known that patients with sickle cell disease (Hb SS) suffer from a wide range of serious complications even in usual conditions, but it remains unclear if subjects with Hb AS display any disturbance during anaerobic or aerobic exercises.

Some authors suggested that SCT could affect exercise performance and recovery in sportsmen and become a disadvantage to certain competitive athletes (4). On the contrary, no medical complications were reported in many supervised sporting events in athletes with Hb AS, especially during the Mexico-City Olympic games (5).

The aim of the present study was to determine the performance capability during maximal exhausting exercise in young and trained students with SCT who live in a hot climate.

MATERIALS AND METHODS

SUBJECTS

The study was performed on students of the National Institute of Sports and Education of Sénégal (Institut national supérieur d'Éducation populaire et sportive – INSEPS) in Dakar (Senegal). This establishment trains future teachers of physical and sports education. All of the students are submitted to intellectual and physical tests before enrolment; during the academic terms they perform various physical activities for 20 h a week on average. A total of 24 melanoderm male students participated in this study and the majority of them were athletes in good physical conditions, some of them competing in clubs of Senegal. On entry to the study, students did not know their haemoglobin type, with the exception of some SCT subjects who had been detected in an independent survey. Twelve students with sickle cell trait and 12 control subjects with normal haemoglobin participated in this study; their anthropometric data are presented in *Table 1*.

The experiments were carried out in Dakar during the month of February in morning hours. In the laboratory where the subjects performed the cycloergometer test, the ambient temperature ranged between 25 and 30⁰ C and humidity between 70 and 85 % ; the Luc Leger test was carried out outdoors and the temperature with exposure to the sun was about 50⁰ C. The experimental

Table 1

Anthropometric data in the subjects with the sickle cell trait (SCT) and in the control group

	Controls (n=12)	Subjects with SCT (n=12)	Statistical significance
Age (year)	23.5 ± 1.2	24.5 ± 1.4	NS
Height (cm)	181.2 ± 4.8	181.6 ± 6.4	NS
Weight (kg)	70.8 ± 3.2	72.0 ± 4.1	NS
Body mass index(kg/m²)	21.4 ± 1.9	21.8 ± 2.2	NS

protocol, completed with the Declaration of Helsinki, was approved by the local Ethics Committee. A written informed consent was obtained from each subject prior to their participation.

SICKLE CELL DISEASE DETERMINATION

Before any electrophoresis, the presence of SCT was assessed by Emmel's test (6). Examination for the presence and proportion of haemoglobin AS was performed by drawing venous blood into heparinised tubes and routine haemoglobin electrophoresis was performed at alkaline and acid pH in all blood samples. The proportions of A, A₂, S and F haemoglobins were measured by densitometry and subjects were considered to have SCT if their blood contained 35–50 % of haemoglobin S. The subjects with Hb C were not included in the study.

EXPERIMENTAL PROTOCOL

In order to determine indirectly $\text{VO}_{2\text{max}}$, an incremental test was performed on a Monark cycloergometer at a pedalling frequency of 50 cycles/min. The exercise started with a 5 min warming-up at 100 watts and then the workload was increased by 25 W every three minutes until exhaustion. In these measurements, five or six exercise levels were reached according to each subject's capacity. Power output, heart rate, systolic and diastolic blood pressure were determined at the end of each level. $\text{VO}_{2\text{max}}$ was calculated according to the Astrand classic nomogram. The duration of the whole test ranged between 15 and 20 min.

Another performance test was carried out according to Luc Léger's technique. It consisted of repetitive 40 m runs, without intermediary rest, carried out consecutively at increasing controlled speed until exhaustion; this allows an indirect calculation of $\text{VO}_{2\text{max}}$ and gives values that are generally close to those of the Astrand test. In our study, the whole test duration did not exceed 10 min and only the final heart rate was evaluated. This test allowed us to duplicate the measurement of maximal aerobic capacity.

DATA ANALYSIS

The data are presented as mean(standard deviation). Student's *t*-test on the mean values was used to compare results in the two groups and the difference was considered significant if probability values were lower than 0.05.

RESULTS

Anthropometric data are shown in *Tables 2* and *3*. In haematological measurements, there were no significant differences between SCT and control subjects. Only the haemoglobin concentration and haematocrit values were a little higher in SCT. The results of haemoglobin electrophoresis are given in *Table 3*. As expected, the electrophoretic values for HbA and HbS were much different. For HbS in SCT, the highest value was 47.6 and the lowest one 34.2.

Clinically, all the tests were endured normally and similarly by all the athletes: mean heart rates evolved in the same way in Hb AS and Hb AA during the exercise and recovery and blood pressure were not different between the two groups.

Cardiovascular performance data are presented in *Tables 4* and *5*. They were very similar in the two groups; the greatest similarity was shown by $\text{VO}_{2\text{max}}$ that evaluates the athletic level of subjects. During the recovery phase, a decrease in heart rate was very fast (-45 % in 3 min). The observed values show a good physical training of these students, even if absolute values were not so elevated as expected.

Table 2
Haematological data in 12 subjects with the sickle cell trait (SCT) and 12 controls

	WBC (10 ³ /mm ³)	RBC (10 ⁶ /mm ³)	Hb (g/dl)	HCT (%)	MCV μ ³	MCHC (%)
Controls mean ± SD	4.3 ± 0.7	5.0 ± 0.3	14.0 ± 1.2	43.2 ± 3.3	86.4 ± 7.7	32.3 ± 0.6
SCT mean ± SD	4.6 ± 1.1	5.1 ± 0.4	14.9 ± 1.0	45.2 ± 3.8	87.4 ± 3.8	32.8 ± 0.6
Statistical significance	NS	NS	NS	NS	NS	NS

WBS, white blood cell; RBC, red blood cell; Hb, haemoglobin; HCT, haematocrit; MCV, mean cell volume; MCHC, mean cell haemoglobin concentration; NS, non-significant.

Table 3
The results of haemoglobin electrophoresis (in %)

	HbA 97.2 ± 0.3	HbA2 2.4 ± 0.6	HbS 0	HbF 0.40 ± 0.02
Mean ± SD in control group				
Mean ± SD in SCT group	54.0 ± 4.8	2.8 ± 1.7	42.3 ± 4.3	0.58 ± 0.04

Table 4
Bicycle ergometer test. Cardiopulmonary performance during exercise in the subjects with the sickle cell trait (SCT) and in the controls

	Controls (n = 12)	SCT subjects (n = 12)	Statistical significance
Maximal power (W)	194 ± 30	186 ± 25	NS
Maximal heart rate (c/min)	188 ± 21	182 ± 15	NS
Recovery heart rate (c/min)	103 ± 15	101 ± 13	NS
VO ₂ max (ml/min/kg)	44.7 ± 8.1	44.6 ± 6.9	NS

NS, non-significant

Table 5
The results of the Luc Leger test

	<i>Controls</i> (n = 12)	<i>SCT subject</i> (n = 12)	Statistical significance
Number of stages	10.83 ± 0.98	10.95 ± 1.09	NS
VO ₂ max (ml/min/kg)	53.0 ± 4,6	53.4 ± 4.2	NS

NS, non-significant

The difference between the values obtained from the ergometer test and those from the field test in one and the same individual was an unexpected finding. The difference was attributed to a lower adaptation of track athletes to pedalling on a mechanically braked ergometer and/or to better environmental conditions outdoors as compared with those in the atmosphere of the laboratory.

DISCUSSION

Haemoglobinopathy S has been implicated as a risk factor during physical activities, with sickle cell formation compromising O₂ delivery particularly in extreme conditions (7). An example of such activities is a strenuous exercise carried out at an altitude that induces a low level of PO₂ and severe lactic acidosis in the arterial blood. The sickle erythrocytes would either chemically diminish O₂ exchange capability or mechanically interfere with microcirculation within the exercising muscles (8). This process may lead to clinical manifestations. It is well known that these manifestations can be severe, or even lethal, in sickle cell disease (Hb SS) and some fatal complications were observed in the Hb AS heterozygous form (9). In the majority of cases, however, clinical symptoms are moderate or absent (10) and several studies failed to confirm that SCT is a risk factor or even a disadvantage for sports activities (11,12).

These controversial results could be explained by the facts that the group of subjects studied was non-homogeneous, that there were differences in Hb and HbS levels among the SCT subjects or that the conditions of each trial were slightly different

On the other hand, the two groups of young well-trained men in our study were very homogenous. The subjects participating in this study were matched as closely as possible and their athletic status and level of training were the same; these were high, as shown by the results of the Luc Leger test. The subjects were submitted to similar conditions of elevated ambient temperature and exhausting exercise, both indoors and outdoors that included aerobic as well as anaerobic

metabolisms. Nevertheless, we observed that O₂ tissue uptake was not impaired in the SCT subjects whose performance as well as recovery were similar to those of the controls. So we have no arguments for a decrease in O₂ transport by haemoglobin Hb AS in a dramatic situation, unless the subjects developed a compensatory haemoconcentration, as suggested by *Gozal et al.* (4). According to the literature data, exposure to high ambient temperatures would be an important associated risk factor because the complications reported were frequently diagnosed as a heat stroke (13). Our study cannot provide arguments to support either of the views. On the other hand, our findings confirm the conclusions of previous studies that used different exercise protocols and in which no incidence of SCT in the performance was observed (10,11,14). These data suggest that certain metabolic adaptations exist in aerobic and anaerobic paths and may operate in subjects with SCT because, in this situation, a significant increase in erythrocyte sickling rate was recorded (8); however, genetic traits for this tolerance cannot be excluded. It is possible that strenuous activities of longer duration induce some difference or difficulty for these subjects. Therefore, it is obvious that continuous research is needed in this area.

In conclusion, if there is any impairment in the delivery and utilization of oxygen, as maintained by some authors, these data suggest that certain adaptation processes developed by training may operate in SCT sportsmen during high-intensity exercise. We can state that subjects with SCT may participate in sports competitions similarly to subjects with normal Hb AA. Therefore, the aptitude certificates for usual physical activities and competitions may freely be issued for SCT subject and the eventual successes or failures will only be depending on athletic values.

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FYZICKÉ VÝKONY U MLADÝCH OSOB SE SRPKOVITOSTÍ

S o u h r n

Srpkovitost (SCT) je genetická abnormalita červených krvinek, která postihuje zejména lidi v Africe a pocházející z Afriky. V západní Africe se vyskytuje v 10–15 % a stejný výskyt je pozorován u sportovců a u obyvatelstva se sedavým způsobem života. Různí autoři se domnívali, že tato hemoglobinopatie snižuje fyzické výkony nebo je dokonce rizikovým faktorem pro různé úrazy, když se přidají nepříznivé podmínky okolního prostředí, zejména středně vysoká nadmořská výška nebo vysoká teplota okolí. Můžeme udělat závěr, že výkonnost je rovnocenná u dobře trénovaných osob s SCT a bez ní. Hemoglobinopatie Hb AS nemá žádné závažné důsledky na vyčerpávající cvičení, alespoň když doba jeho trvání nepřesahuje půl hodiny.

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