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ORIGINAL ARTICLE

Evaluation of functional capacity for exercise in children and adolescents with sickle-cell disease through the six-minute walk test☆

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KEYWORDS
Sickle cell anemia;
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Abstract
Objective: To evaluate lung functional capacity (FC) for physical exercise in children and adolescents with sickle cell disease (SCD) through the six-minute walk test (6MWT).
Method: A cross-sectional prospective study was performed to evaluate the FC of 46 patients with SCD through the 6MWT. The following parameters were assessed: heart rate (HR), respiratory rate (RR), peripheral pulse oxygen saturation (SpO_2), peak expiratory flow (PEF), blood pressure (systolic and diastolic), dyspnea, and leg fatigue (modified Borg scale) at rest, in the end of the test, and ten minutes after the 6MWT. The total distance walked was also recorded. For statistical analysis, the parametric variables were analyzed using the paired Student's *t*-test, analysis of variance (ANOVA), and Bonferroni multiple comparisons, with a significance level set at $p \leq 0.05$.
Results: The 46 patients were aged age 9.15 ± 3.06 years, presented baseline Hb of 9.49 ± 1.67 g/dL, and walked 480.89 ± 68.70 m. SCD diagnosis was as follows: group 1-HbSS (n = 20)/HbSβ^0-thalassemia (n = 3) and group 2 - HbSC (n = 20)/HbSβ^+-thalassemia (n = 3). Regarding total distance walked, patients in group 1 walked a shorter distance than patients in group 2 (459.39 ± 57.19 *vs.* 502.39 ± 73.60 m; p = 0.032). There was no statistical difference regarding PEF in the three moments of evaluation. The SpO_2 in ambient air and SpO_2 with O_2 differed between groups 1 and 2 ($p < 0.001$ *vs.* p = 0.002), as well as the RR (p = 0.001).
Conclusion: These patients showed a lower FC for exercise than that predicted for the age range in the literature. Patients diagnosed with HbSS/Sβ0-thalassemia had a lower performance in the

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test than those with HbSC/Sβ⁺-thalassemia regarding total distance walked, RR, and SpO_2 after the 6MWT.

PALAVRAS-CHAVE
Anemia falciforme;
Síndrome torácica aguda;
Exercício;
Estado nutricional

Avaliação da capacidade funcional para o exercício de crianças e adolescentes com doença falciforme pelo teste da caminhada de seis minutos

Resumo

Objetivo: Avaliar a capacidade funcional pulmonar (CF) para o exercício físico de crianças e adolescentes com doença falciforme (DF) pelo teste da caminhada de seis minutos (TC6').

Métodos: Estudo transversal prospectivo avaliando a CF pelo TC6' de 46 pacientes com DF. Foram avaliados: frequência cardíaca (FC), frequência respiratória (FR), saturação de pulso de oxigênio (SpO_2), pico de fluxo expiratório (PFE), pressão arterial (PA) sistólica e diastólica, dispneia e cansaço em membros inferiores (escala de Borg modificada) em repouso, ao término e 10 minutos após o TC6' e a distância percorrida. Análise estatística: test *t*-Student pareado, análise de variância e comparações múltiplas de Bonferroni, significância $p \leq 0,05$.

Resultados: Dos 46 pacientes, a média ± dp da idade foi 9,15 ± 3,06 anos, hemoglobina basal 9,4 ± 1,67 g/dL e distância percorrida 480,89 ± 68,70 m. Diagnóstico da DF: Grupo 1– HbSS (n = 20)/HbSβ⁰-talassemia (n = 3); e Grupo 2 – HbSC (n = 20)/ HbSβ⁺-talassemia (n = 3). O Grupo 1 apresentou menor distância percorrida do que o Grupo 2 (459,39 ± 57,19 *vs* 502,39 ± 73,60m; p = 0,032). Não houve diferença estatística em relação ao PFE. A SpO_2 em ar ambiente e a SpO_2 com O_2 (1 L/min) após o teste foi maior no Grupo 2 ($p < 0,001$ e $p = 0,002$, respectivamente). A FR foi maior no Grupo 1 ao final do TC6' ($p < 0,001$).

Conclusão: Esta amostra apresentou CF para o exercício abaixo do predito para a faixa etária na literatura. Os pacientes com HbSS/Sβ⁰-talassemia apresentaram desempenho inferior na distância percorrida, FR e SpO_2 após o TC6′, comparativamente aos pacientes com HbSC/Sβ⁺-talassemia.

Introduction

Sickle cell anemia (HbSS) is the most common monogenic hereditary disease in Brazil, with an estimated prevalence of heterozygotes for HbS ranging from 2% to 8% in the general population.[1] HbSS, the most severe form of sickle cell disease (SCD) is a hemoglobinopathy resulting from the single amino acid substitution of a glutamic acid for a valine at the sixth position of the beta globin chain, on chromosome 11, giving rise to hemoglobin S (HbS).[2] This alteration in hemoglobin is responsible for the anomalous form of erythrocytes, leading to hemolytic anemia, endothelial vasculopathy, and vaso-occlusive phenomena, followed by tissue ischemia and necrosis, with subsequent organ dysfunction, which are responsible for the high mortality of SCD.[1,2] SCD occurs when HbS combines with another hemoglobinopathy, such as C, D, β-thalassemia, or another HbS.[3]

The lung is a major target organ of acute and chronic complications in SCD; acute chest syndrome (ACS) is the second most frequent cause of hospitalization in this population, with high rates of morbidity and mortality.[4–6] It is an acute complication usually triggered by a clinical picture of infection. It can be defined by a combination of signs and symptoms, which include dyspnea, chest pain, fever, cough, and a new pulmonary infiltrate.[7] The proliferative vasculopathy that occurs in sickle cell disease is the main cause of the chronic pulmonary alterations that occur in these patients.[8]

The chronic alterations and recurrent episodes of ACS decrease the functional capacity (FC) in patients with SCD. MacLean et al.,[9] when assessing lung function in children with SCD through spirometry, observed a restrictive pulmonary pattern and a progressive reduction in lung volume. Another prospective study,[10] with patients aged 10 to 26 years, found alterations in pulmonary function, with a predominance of mixed or combined pattern.

Thus, the evaluation of FC should be part of outpatient monitoring of these patients. However, studies assessing and addressing the FC in children and adolescents with SCD are limited.[11] A simple and effective method to evaluate the FC is to apply the six-minute walk Ttest (6MWT), which provides information about functional status, oxygen consumption, exercise tolerance, and patient survival according to test performance.[12,13] The 6MWT assesses the individual's submaximal effort, similar to the effort made in some daily life activities, representing their FC to exercise.[12]

Although the 6MWT has not been widely studied in patients with hemoglobinopathies, especially in pediatric patients, preliminary data suggest that there may be a good correlation among this test, maximum oxygen uptake, and severity of pulmonary hypertension in adults with SCD.[14] There are no published articles in the literature evaluating the FC in children and adolescents with SCD; thus this study aimed to evaluate the FC to exercise in children and adolescents with SCD using the 6MWT.

Methods

This was a prospective cross-sectional study evaluating 46 children and adolescents with SCD aged between 6 and