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ORIGINAL ARTICLE

Association between morphometric variables and nocturnal desaturation in sickle-cell anemia☆

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KEYWORDS
Sickle-cell anemia;
Desaturation;
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Abstract
Objective: to evaluate associations between morphometric variables, cervical circumference (CC), and abdominal circumference (AC) with the presence of nocturnal desaturation in children and adolescents with sickle-cell anemia.
Methods: all patients were submitted to baseline polysomnography, oral cavity measurements (maxillary intermolar distance, mandibular intermolar distance, and overjet), and CC and AC measurements.
Results: a total of 85 patients were evaluated. A positive correlation was observed between the height/age Z-score and CC measurement (r = 0.233, p = 0.031). The presence of nocturnal desaturation was associated with CC (59.2± 9.3 *vs.* 67.5 ± 10.7, p = 0.006) and AC measurements (27.0 ± 2.0 *vs.* 29.0± 2.1, p = 0.028). There was a negative correlation between desaturation and maxillary intermolar distance (r = -0.365, p = 0.001) and mandibular intermolar distance (r = -0.233, p = 0.037).
Conclusions: the morphometric variables of CC and AC may contribute to raise suspicion of nocturnal desaturation in children and adolescents with sickle-cell anemia.

PALAVRAS-CHAVE
Anemia falciforme;
Dessaturação;
Maxila;
Mandíbula;
Apneia do sono

Associação entre variáveis morfométricas e dessaturação noturna na anemia falciforme

Resumo
Objetivo: avaliar associações entre variáveis morfométricas e circunferências cervical (CC) e abdominal (CA) com a presença de dessaturação noturna em crianças e adolescentes com anemia falciforme.

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Métodos: todos os pacientes foram submetidos à polissonografia basal, medidas da cavidade oral (distância intermolar da maxila, distância intermolar da mandíbula e *overje*), CC e CA.
Resultados: foram avaliados 85 pacientes. Foi observada correlação positiva entre o escore Z altura/idade e a medida da circunferência cervical (r = 0,233 p = 0,031). A presença da dessaturação noturna associou-se com as medidas da circunferência cervical (59,2 + 9,3 *vs* 67,5 + 10,7; p = 0,006) e abdominal (27,0 + 2,0 *vs* 29,0 + 2,1; p = 0,028). Houve correlação negativa entre a dessaturação e a distância entre os segundos molares da maxila (r = - 0,365, p = 0,001) e da mandíbula (r = - 0,233, p = 0,037).
Conclusões: as variáveis morfométricas e circunferências cervical e abdominal podem contribuir para a suspeita da dessaturação noturna em crianças e adolescentes com anemia falciforme.

Introduction

The main aspect of sickle-cell anemia (SCA) physiopathology is the vaso-occlusive crisis, also called a painful crisis, resulting from the polymerization of hemoglobin S that causes red blood cells to assume a ''sickle'' shape, resulting in the obstruction of small-caliber blood vessels, tissue hypoxia, necrosis, and severe pain.[1]

Among the consequences of red blood cell sickling are painful vaso-occlusive crises, stroke,[2] and chronic hemolytic anemia.[3] Pulmonary complications account for 20% to 30% of deaths in adults with SCA, followed the pulmonary lesions caused by upper airway (UA) obstruction.[4]

Regarding children with SCA, an important causal factor of airway obstruction is adenotonsillar hypertrophy (ATH). Salles et al.[5] observed a prevalence of 55.3% of obstructive ATH in children and adolescents with SCA. When this UA obstruction is associated with obstructive sleep apnea syndrome (OSAS), it can precipitate episodes of hypoxemia,[6] increasing the risk of vaso-occlusive crises.[7]

The nocturnal arterial oxyhemoglobin desaturation in SCA occurs in up to 83% of children with the disease, and may result from OSAS or pulmonary disease, or may be secondary to the decreased oxygen affinity of hemoglobin S.[8] Oxyhemoglobin desaturation is common during sleep, which is associated with hypoventilation and is exacerbated by UA obstruction;[6] in patients with SCA, there may be increased risk for airway obstruction due to ATH.[9]

Factors that worsen UA obstruction are ATH and the craniofacial consequences caused by obstructive ATH itself, such as predominant mouth breathing; vertical, horizontal, and transverse maxillary alterations, incompetent lip seal; and unbalanced orofacial muscles, mainly the mandibular elevator and depressor muscles.[10,11]

Thus, the present study aimed to evaluate associations between morphometric variables, cervical circumference (CC), and abdominal circumference (AC) with the presence of nocturnal desaturation in children and adolescents with SCA.

Material and Methods

This was a cross-sectional, contemporary cohort study with sequential allocation of 85 patients with SCA enrolled in a referral center for hematology and transfusion medicine, between May of 2007 and May of 2008. The following inclusion criteria were used: diagnosis of SCA attained through the quantitative analysis of hemoglobin by hemoglobin electrophoresis or high-performance liquid chromatography (HPLC), performed with Variant II equipment (Bio-Rad, USA); age between 2 and 19 years; clinically stable; completed the questionnaire; allowing pediatric and otorhinolaryngological evaluation; and undergoing nocturnal polysomnography. The following exclusion criteria were used: other genetic syndromes, debilitating diseases, acute hepatitis, previous treatment for OSAS or recent craniofacial trauma; using hypnotic drugs; having been treated with corticosteroids; pregnancy; and presence of infection during the evaluation.

The PEPI-Sample program (Sagebush Press,USA) was used to calculate sample size, and the following parameters were used: a confidence level of 95%, and the prevalence of OSAS in children/adolescents of 5% (4.9% as an acceptable prevalence difference).

The sample was obtained from a population of approximately 1,000 children and adolescents with SCA, registered at a referral center for hematology and transfusion medicine. Therefore, to meet the objectives, the calculated sample size consisted of 71 patients. Considering 10% losses, the total (n) consisted of 78 patients.

Age was measured in full years, according to the birth date. Ethnicity was self-reported, according to the official nomenclature of demographic censuses, using skin color as reference (white, mixed-race, or black).

Weight was measured using a mechanical scale (model 131; Filizola - Brazil). Length was measured with a stadiometer. These measurements were compared to the growth charts of the National Center for Health Statistics and converted into Z-scores for body mass index (BMI), weight/age, and height/age based on age and gender, using the Epi-Info software (release 3.4.1;CDC- USA).

The oral cavity assessment was performed by a single otolaryngologist in the Frankfurt position, using a standard tool. The measures of the oral cavity, except the overjet (OJ), were made with the tongue in a relaxed position and with a mouth opening angle of 20° to the mandibular condyle. For that purpose, a dry-point, 20° fixed-aperture compass was used, which was placed on the topography of the temporomandibular joint, the tip of its upper leg aligned with the upper central incisors and the lower compass leg aligned with the lower central incisors, to yield the desired mouth opening.