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ORIGINAL ARTICLE

Evaluation of bone mineral density in children with sickle cell disease☆

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Received 17 November 2013; accepted 3 April 2014
Available online 25 April 2015

KEYWORDS
Bone mineral density;
Densitometry;
Sickle cell disease;
Low bone mineral density;
Osteoporosis

Abstract
Objective: To evaluate bone mineral density (BMD) in children with sickle cell disease (SCD) in the Community of Madrid.
Materials and methods: The BMD was estimated in 40 children with SCD, and with an age range between 3 and 16 years, using densitometry (DXA), as recommended by the International Society for Clinical Densitometry (ISCD).
Results: The mean age at the time of the study was 7.97 ± 3.95 years, the mean value of the DXA expressed in *Z*-score was −0.91 ± 1.46 with a range of minimum values −5.30 and 2.30 maximum. More than half (57.5%) of all the children had normal BMD (*Z* > −1), 25% had low BMD (*Z* between −1 and −2), and 17.5% showed abnormal *Z*-score values of osteoporosis (*Z*-score < −2). The Pearson linear correlation was statistically significant between *Z*-score value and the haemoglobin level ($r = 0.368$, $p = .019$), finding no correlation with the levels of 25 (OH) vitamin D.
Conclusion: Prospective studies are needed with a larger number of patients to understand the future implications of bone densitometry changes and associated risk factors.

PALABRAS CLAVE
Densidad mineral ósea;
Densitometría;

Evaluación de la densidad mineral ósea en pacientes con enfermedad de células falciformes

Resumen
Objetivo: Evaluar la densidad mineral ósea (DMO) en niños con enfermedad de células falciformes (ECF) de la Comunidad de Madrid.

☆ Please cite this article as: Garrido Colino C, Beléndez Bieler C, Pérez Díaz M, Cela de Julián E. Evaluación de la densidad mineral ósea en pacientes con enfermedad de células falciformes. An Pediatr (Barc). 2015;82:216–221.

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Enfermedad de células falciformes;
Osteopenia;
Osteoporosis

Material y métodos: Se valora la DMO en un total de 40 niños con ECF y rango de edad entre 3-16 años, mediante densitometría (DEXA) siguiendo las recomendaciones de la Sociedad Internacional de Densitometría Clínica (ISCD).
Resultados: La edad media en el momento del estudio fue de 7,97 ± 3,95 años; el valor medio de la DEXA expresado en Z-score es de -0,91 ± 1,46 con un rango de valores mínimo de -5,30 y máximo de 2,30. Un 57,5% de los niños tiene DMO normal (Z > -1), un 25% tienen DMO baja (Z entre -1 y -2) y un 17,5% presentan Z-score patológico con valores de osteoporosis (Z-score < -2). Los estudios de correlación solo encuentran una correlación lineal de Pearson significativa estadísticamente entre valor de Z-score y valor de Hb (r = 0,368, p = 0,019), no encontrando correlación con los niveles de 25 (OH) D.
Conclusión: Se necesitan estudios prospectivos, con mayor número de enfermos para conocer las implicaciones futuras de la densitometría alterada y los factores de riesgo asociados.

Introduction

In recent years, bone mass alterations or low bone mineral density has been the subject of ongoing attention. The peak bone mass acquired by the end of maturation is a good predictor for the risk of future fractures. Clinicians are generally familiar with the terms ''osteopaenia'' (decrease in bone mass) and ''osteoporosis'' (a more severe loss of bone mass associated with a greater risk of fracture).[1–3] The diagnosis of osteoporosis in children is defined as low bone mineral density (BMD) and a fracture history with long bone fracture of the lower extremities, vertebral compression fracture, or two or more long bone fractures of the upper extremities.[1]

The earliest studies that analysed the role of bone health in fractures in the paediatric age group were conducted in New Zealand.[4,5]

A prospective study in healthy African children 5–9 years of age that analysed the relationship between vitamin D and fractures concluded that a significant number of children with fractures also have low vitamin D levels (59%).[6] Vitamin D deficiency seems to play a previously unknown role in fractures in the paediatric age group.

Furthermore, there is evidence that vitamin D supplementation in healthy children and adults with deficiency leads to a significant increase in BMD, decreasing the risk of fractures.[7,8]

Children with sickle cell disease (SCD) often develop bone complications manifested as vaso-occlusive bone pain crises, dactylitis, osteomyelitis, avascular necrosis or vertebral deformity. In this population, the literature describes a decrease in BMD secondary to chronic anaemia and bone marrow hyperplasia and associated with a higher risk of osteopaenia and osteoporosis.[9,10]

Recent studies show that children with SCD frequently have severe vitamin D deficiency. For instance, a cross-sectional study of 78 children with SCD performed in our hospital found that only 20.5% had 25-hydroxyvitamin D (25(OH)D) levels above 30 ng/mL, the threshold considered optimal for bone health.[11]

Despite the higher risk of bone disease in this population, there is little information on BMD in children with SCD and its association with 25(OH)D levels and the risk of fractures. Our study assessed the prevalence of BMD abnormalities in children with SCD from the autonomous community of Madrid using densitometry (dual-energy X-ray absorptiometry [DXA])[12,13] and analysed potential risk factors associated with low BMD.

Materials and methods

We conducted a cross-sectional study between October 2009 and February 2011 in a cohort of 120 children with SCD residing in the autonomous community of Madrid and who were being followed up at the paediatric haematology department of the Hospital Gregorio Marañón (HGUGM) to evaluate BMD in this population by means of DXA.

We included a total of 40 patients, starting at age 3–4 years to avoid the need for sedation during DXA. We included all possible patients who made at least one visit to the hospital during the study period. We excluded children known to have conditions affecting growth or nutritional status, with chronic liver or kidney function abnormalities, undergoing treatment with drugs that affect the skeleton, or who had had SCD complications the previous month (vaso-occlusive crisis, fever, acute chest syndrome).

The study was approved by the board of ethics of our hospital, and we obtained the signed informed consent from all participants.

We collected the demographic and anthropometric data in the course of routine follow-up visits to a physician in the team.

Bone mineral density was assessed by means of DXA. We measured BMD at the posterior–anterior spine, and not at the hip and proximal femur due to the great variability of the latter areas during growth, adhering to the International Society for Clinical Densitometry (ISCD) recommendations for children. The instrument used for this study was a LUNAR DPX-IQ 5539 machine that met the requirements for research in paediatrics. The results were interpreted by an