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Original article

Bone mineral density evaluation among patients with neuromuscular scoliosis secondary to cerebral palsy☆

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ABSTRACT

Objective: To evaluate bone mineral density among patients with neuromuscular scoliosis secondary to quadriplegic cerebral palsy.
Methods: This was a descriptive prospective study in which both bone densitometric and anthropometric data were evaluated. The inclusion criteria used were that the patients should present quadriplegic cerebral palsy, be confined to a wheelchair, be between 10 and 20 years of age and present neuromuscular scoliosis.
Results: We evaluated 31 patients (20 females) with a mean age of 14.2 years. Their mean biceps circumference, calf circumference and body mass index were 19.4 cm, 18.6 cm and 16.9 kg/m^2, respectively. The mean standard deviation from bone densitometry was −3.2 (z-score), which characterizes osteoporosis.
Conclusion: There is high incidence of osteoporosis in patients with neuromuscular scoliosis secondary to quadriplegic cerebral palsy.

Avaliação da densidade mineral óssea em pacientes portadores de escoliose neuromuscular secundária a paralisia cerebral

Palavras-chave:
Escoliose
Neuromuscular
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RESUMO

Objetivo: avaliar a densidade mineral óssea em pacientes portadores de escoliose neuromuscular secundária à paralisia cerebral tetraespástica.
Métodos: estudo prospectivo, descritivo, em que se avaliaram, além da densitometria óssea, dados antropométricos. Como critério de inclusão, adotamos pacientes com paralisia cerebral tetraespástica, cadeirantes, entre 10 e 20 anos e com escoliose neuromuscular.

☆ Work developed at Hospital Santa Casa de Misericórdia de Vitória, ES, Brazil.
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Resultados: avaliamos 31 pacientes, 20 do sexo feminino, cuja média de idade foi de 14,2 anos. A média da circunferência bicipital, da panturrilha e do IMC foi de 19,4 cm, 18,6 cm e 16,9 Kg/m^2, respectivamente. O desvio padrão médio encontrado na densitometria óssea foi de –3,2 (*z-score*), o que caracteriza osteoporose.

Conclusão: existe elevada incidência de osteoporose em pacientes portadores de escoliose neuromuscular secundária à paralisia cerebral tetraespástica.

Introduction

Many neuromuscular diseases lead to development of spinal deformities. Among these, cerebral palsy is the most frequent: its incidence may range from 25% to 100% of such patients, depending on the degree of neuromuscular involvement.[1] Its etiological origin is secondary to imbalance between the muscle forces in the axial skeleton,[2] caused by lesions in the upper and lower motor neurons.[3] Scoliosis usually presents a C-shaped format, in association with pelvic obliquity, and it frequently progresses even after skeletal maturity has been reached.[4]

Thus, in cases of severe deformity, or in those in which progression of the curve is detected, surgical treatment becomes necessary, with the aims of avoiding progression and restoring or maintaining the sagittal and coronal balance and the capacity to sit, thereby leading to a great improvement of the patients' quality of life.

In these cases, despite the need to perform surgical treatment, the complication rate is very high and is directly related to the impairment of cardiorespiratory and gastrointestinal function and the nutritional grade shown by the patient.[5] Among all the possible complications arising from surgery, infection and loosening of the synthesis material used for correcting the deformity are the ones most frequently observed.[6]

Failure of fixation of the pedicle screws in the spine may occur due to osteoporosis of the vertebra, caused by factors such as the severity of the neurological impairment, increasing difficulty in eating and use of anticonvulsants.[7]

Very few studies have analyzed the bone mass of patients with tetraspastic cerebral palsy. Many complications can result from loosening of the synthesis material in such patients. This can be prevented through correct analysis of bone metabolism and early treatment of patients who present low bone mass. We conducted the present study with the objective of analyzing the bone mass of patients with cerebral palsy who also had neuromuscular scoliosis. Through this, it might become possible to adopt appropriate preventive measures for avoiding the development of osteoporosis and consequently to achieve improvement of their quality of life.

Sample and method

This was a prospective study of descriptive nature for which data covering the period from February 2012 to January 2013 were gathered. The inclusion criteria were that the patients needed to present neuromuscular scoliosis due to cerebral palsy, with a tetraspastic component, and were using a wheelchair. Patients aged less than 10 years and over 20 years and patients whose scoliosis was not of neuromuscular origin due to cerebral palsy were excluded.

A convenience sample was used, which was formed as the patients came to the orthopedic outpatient clinic of a philanthropic hospital in Vitória. In total, 31 patients were evaluated (20 females), with a mean age of 14.2 years. Subsequently, each patient's bone mass was determined by means of bone densitometry on the lumbar spine, on the Lunar Prodigy Advance densitometer, model PA+41606, which produces digitized densitometry scans by means of X-rays, from a special constant 76 kV source with an efficient *k*-edge dose filter. The densitometry was also computer-assisted, by means of the Windows-based Prodigy Bis software.

The results were represented numerically by means of absolute values and percentages and were documented in accordance with protocols. The data analysis was performed using the software Microsoft Office/Excel 2007® and GraphPad Prism® (San Diego, CA, USA).

In addition to bone mass, anthropometric data were evaluated, such as measurements of estimated height, weight, body mass index (BMI), biceps circumference and calf circumference. Specific data such as whether the patient had undergone gastrostomy, was doing physiotherapy or was using an adapted wheelchair were also ascertained. Furthermore, laboratory tests such as hemogram, TSH, free T4, potassium, calcium, serum iron, ferritin, transferrin, C-reactive protein (CRP), total proteins and albumin were performed.

To calculate BMI, the formula used was $BMI = W/H^2$, in which W = weight and H = estimated height. The estimated height was calculated by means of the following formula: $H = (2.69 \times KH) + 24.2$, in which KH was the distance from the knee to the heel.[8]

Results

Among the 31 patients analyzed, 11 were constantly doing motor physiotherapy. Only 11 were using adapted wheelchairs; the remaining 20 were using conventional wheelchairs.

The anthropometric measurements were: weight, 28 kg; height, 143.6 cm; biceps circumference, 19.4 cm; calf circumference, 18.6 cm; and BMI, 16.9.

Table 1 – Mean values from laboratory tests.

	Mean value	Reference range
Hemoglobin	13.52	12–14 g/dL
Hematocrit	39.94	38–42%
Total proteins	7.21	6.0–8.0 g/dL
Albumin	4.42	3.5–5.5 g/dL
TSH	2.38	0.350–5.50 UI/mL
Free T4	1.16	0.70–2.0 ng/dL
Serum calcium	8.89	8.4–10.6 mg/dL
Potassium	4.26	3.5–5.2 mEq/L
Ferritin	34.01	10–140 ng/mL
Serum iron	92.15	Men: 60–150 mcg/dL Women: 40–145 mcg/dL
Transferrin	218.19	200–360 mg/dL
CRP	5.29	<6 mg/L

Table 2 – Bone density analysis.

Bone density	Normal	Osteopenia	Osteoporosis	Total
No. of patients	1 (3.3%)	5 (16%)	25 (80.7%)	31

The mean values from the laboratory tests were within the normal ranges, as shown in Table 1.

On analyzing the bone densitometry results, we found that the mean Z-score in the lumbar spine was −3.2, with a minimum of −6.0 and a maximum of 2.1. Thus, among the 31 patients analyzed, 25 presented osteoporosis, five had osteopenia and one had normal density, as shown in Table 2.

Regarding these patients' BMI, we found a strong relationship between low BMI and low bone mineral density, which was a statistically significant result ($p = 0.005$, i.e. <0.05), as shown in Fig. 1. We observed that the patients' degree of malnutrition was directly related to low bone mass.

Discussion

Surgical treatment for neuromuscular scoliosis is often difficult because of the various complications that are possible and because of the constant need for a multidisciplinary team.[9] Therefore, when surgical treatment is indicated, the utmost caution is required in order to avoid these possible complications.

The malnutrition observed among these patients, which is due to difficulty in eating, use of anticonvulsants and neurological impairment, may lead to diminished bone mass, which implies a higher rate of loosening of the synthesis material.[6] In these cases, new surgical interventions are needed in order to decrease the pseudarthrosis rate.

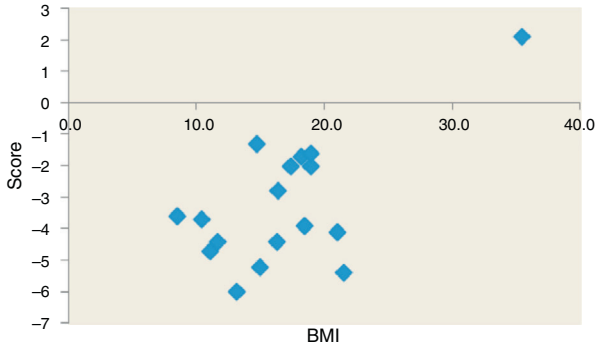

Fig. 1 – Relationship between BMI and Z-score.

Although the possibility of low bone mass among these patients is already known, there is no preoperative routine for assessing it. We did not find any studies in the literature that discriminate among these patients' mean bone mineral density (BMD) values.

To evaluate BMD and diagnose osteoporosis in children and adolescents, we used the Z-score, which is the number of standard deviations resulting from comparing a child's BMD value with the mean BMD value of a standard population of the same sex and age. The values are considered to be abnormal when the Z-score is less than −1. In these cases, the thresholds for osteopenia and osteoporosis are not very well defined or validated, but it is considered that children present osteoporosis when their Z-score is lower than −2.[7]

In a study by Henderson, Lin and Greene, which included 139 children and adolescents with varying severities of cerebral palsy, BMD was evaluated in the proximal femur and lumbar spine, and the mean Z-score for the lumbar spine was -0.92 ± 0.14.[10] However, in that study, a heterogenous group of patients was used, and the factor that best correlated with low BMD was the capacity to walk. In our study, a homogenous group in which all the patients were non-walkers was evaluated, and we only used the BMD of the lumbar spine. In this manner, we observed that there was greater loss of bone mass. The mean Z-score was −3.19.

Children with mental deficiencies may present difficulties in communication, oromotor and postural dysfunctions, food intolerances and alterations of appetite caused by their medication, which frequently interferes qualitatively and quantitatively with nutrient intake and is reflected in their nutritional status.[11] Most of the studies available present lower anthropometric indicators than those of children without deficiencies. They also show that adequate macro and micronutrient intake is rare, including calcium and iron intake.[12]

Sullivan et al.[12] reported that these children have a diet consisting of milk-based drinks and milk products, mainly because the liquid or pasty consistency of these products makes them easier to consume. The low variety and quantity of nutrients may contribute towards a situation of malnutrition and consequent spoliation of electrolytes, thereby comprising the water-electrolyte balance. Another important factor that may be related to malnutrition among these patients is chronic treatment with certain anticonvulsants that are considered to be hepatic enzyme inducers.[13] These act on the enzymatic activity of the P450 system and thus decrease the availability of vitamin D in the organism and consequently interfere with calcium and phosphorus absorption.[14] However, in our study, the mean serum potassium, calcium, iron and albumin values remained within the limits of normality, even though the patients presented low bone mass.

There is a strong association between low BMI and osteoporosis, which may influence the final result from treating vertebral deformities. Therefore, we judge that it is essential to conduct a preoperative nutritional evaluation on these

patients, which makes the surgical treatment safer, with a lower complication rate.

Conclusions

There is high incidence of osteoporosis among patients with neuromuscular scoliosis secondary to tetraspastic cerebral palsy.

Conflicts of interest

The authors declare no conflicts of interest.

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