Bone mineral density in Marfan syndrome

Sir, We read with interest the article of Carter et al. [1], who demonstrate reduced axial and femoral neck bone densitometry (BMD) in men and women with Marfan syndrome (MFS). We have studied hip and wrist BMD of sixty adults patients of both sexes (40 women, 20 men) with Marfan syndrome according to criteria revised in 1996. Because 30% of our patients had previous spine surgery, the lumbar spine was not measured. The results of this study have been published recently [2]. We confirm a significant decrease in BMD both at the hip and the wrist in men and women with MFS.
Although the mean age of our patients (32.9 ± 9.3 yr) was comparable, they had a lower BMI (20.9 ± 3.9) and Z scores at the hip (−1.26 ± 0.9) and wrist (−1.06 ± 1.06) were lower in our patients than in Carter’s group. Men and women had the same reduction in BMD at both anatomical sites. We found significant differences between cortical and trabecular zones in patients with MFS, as expressed by the comparison of 1/3 mean proximal radius evaluating mainly cortical bone versus ultra distal and total femur versus Ward’s triangle.

As part of a multidisciplinary out-patient clinic, our patients have a systematic clinical examination. The prevalence of previous fracture was 10% in our group of adults, most being traumatic fracture. We now have 120 patients in the cohort and the prevalence of fracture events has not changed. We also investigated a possible relationship between phenotypic severity of MFS (as defined by previous eye, vascular or skeletal surgery) and did not find a significant relationship with BMD.

MFS patients have a particular skeletal and muscular morphology, i.e. tall height, low fat mass and long bones. In our patients, there was a correlation between BMD and body mass index. These characteristics may produce some bias when BMD data of these patients are compared with the manufacturer’s (HOLOGIC, Waltham, MA, USA) database [3].

We had the opportunity to obtain hip BMD of a group of normal tall men (mean height 185.8 ± 3.7 cm) and compared their BMD with that of men with MFS. Significantly reduced femoral neck BMD in men with MFS in comparison with normal tall controls was confirmed.

The clinical relevance of low BMD in MFS is not clear. Our data on fracture rate indicate that the rate does not exceed 10% in a population of young adults. A preliminary study of children and adolescents with MFS suggests that abnormal acquisition of peak bone mass is an important factor contributing to low BMD in MFS.

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Reply

We are very interested in the results obtained by this group in their cohort of adult patients with Marfan’s syndrome. In our original paper, bone mineral density was reduced at the lumbar spine and the hip, but we were uncertain of the significance of the results at the hip, given the atypical skeletal proportions of patients with Marfan’s syndrome.

This study would appear to show that even when compared with normal individuals with tall stature and skeletal proportions more closely resembling those of patients with Marfan’s syndrome than the manufacturer’s controls, bone mineral density does indeed appear to be reduced. However, the observation that fracture rates do not appear to exceed those seen in a population of young adults is important, and it remains to be seen whether the small reduction in bone mineral density at the hip is of clinical significance.

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