Introduction

Myelomeningocele (MMC) is a congenital malformation of the neural tube, and accounts for as many as 4.7 in 10,000 live births\(^1\). Before 1960, as few as 10% of these infants survived, most dying from infection and/or hydrocephalus\(^2\). Today, after shunt improvements and more aggressive treatments, at least 60%-80% of patients born withMMC will survive into adulthood. The main clinical problems are difficulty or inability to stand, walk, voluntarily control of bladder and bowel functions. Often they have lower extremity sensory and motor deficits that impair a normal ambulation and lead to variable degrees of physical inactivity, osteoporosis, and development of pathologic fractures\(^3,4\). Despite this latter risk, little is known about the body composition, bone mineral density (BMD) and its relationship with deambulation and fractures.

Neurological level of spinal cord involvement and ambulatory status, may affect bone mineral density of both upper and lower extremities as measured by \(^{125}\)I photon absorptiometry of the distal radius, tibia, and first metatarsal, respectively\(^5\). In MMC patients, the body composition is different in the upper and lower body. The portion of the body above the neurological lesion is normal in strength, dexterity and skin sensitivity. With a proper training, the upper body may compensate the paralysis of the lower extremities. Body composition, therefore, plays an important role in the functional ability of these patients. It has been reported that the amount of muscle mass in a child with MMC is lower than normal, due to the complete or partial enervation of mus-
cles below the neurological lesion. Such enervation results in a decreased physical activity and a subsequent weight gain. After the age of four, body cell mass and lean mass tissue become increasingly replaced by adipose tissue. Moreover, the percentage of body fat is significantly greater in those with high and mid-lesions, and in non-ambulatory patients. Obesity can cause additional problems, including physical and psychological morbidity, ambulatory deterioration, transference difficulties, higher incidence of pressure sores, difficulty with hygiene and stomal care, and more complications after surgery. Dietary habits and caloric intakes should be evaluated in children with MMC whose weight exceeds the 25th percentile for age. Positive effects of physical activity on fat mass (FM) and bone mineral density (BMD) in normal subjects and in those with disability, are reported in many studies. Sport activity improves lean body mass and reduces fat mass.

The aim of this study was to examine and compare body composition and bone mineral density measurements between different subgroups of patients with MMC.

**Materials and Methods**

**Patients**

Because known bone mineral density standards in children are available for the age group 6 to 19 yrs, of approximately 350 patients referred to the Spina Bifida Center of the Catholic University of Rome, we enrolled 64 patients with MMC aged between 5 and 14 yrs. Four patients were excluded because their parents didn’t give permission. Therefore, we enrolled 60 consecutive patients (mean age 10.4 ranging form 5 to 14 yrs, 34 males and 26 females). Informed consent was obtained from all parents or guardians. Before examination, we acquired personal data by asking each patient or parent, if the patient was younger than 10 years, to fill in a case form. Questions specifically concerned the following issues: educational level (e.g., school frequency, presence of assistant teacher for handicapped), urological aspects (incontinence, urethral catheterization, self-made catheterization, urine loss between the catheterizations, pharmacological therapy, infections) and physical activity (if they performed physiotherapy and/or sport activity, and how many hours during the week). Medullar and brain magnetic resonance imaging (MRI) were acquired (e.g., site and type of lesion). Ventriculoperitoneal shunt implantation was performed on all children. Detailed clinical history, careful clinical examination (from the neurological and physical rehabilitation point of view) were always performed (e.g., tendon reflexes of the four limbs, cutaneous sensitivity, muscle strength assessed according to British Medical Research Council scale, joint function, trophysm, dysmetria). The level of spinal cord lesion was: 10 thoracolumbar, 35 lumbosacral and 15 sacral. The ambulatory status of individuals within this group varied greatly and was categorized into two groups, defined as follows: (1) non-ambulator: full-time wheelchair; (2) full-time ambulator: individual who walks with or without tutorial aid. Of the studied patients, 38 were in the first group and 22 in the second one. Regarding sport activity we observed: 28 subjects practised sport activity. Anthropometry was performed using standard techniques (measurement of skinfolds by a calliper, of waist, hip, upper and middle thigh). Body fat as percentage of body weight was calculated from a 4-skinfold thickness using Brook’s method.

**Bone Densitometry Measurements**

The Bone Mineral Content (BMC) and BMD at the lumbar spine and femoral neck levels were performed in all patients by DEXA using a Hologic QDR 2000 analyser (Hologic, Waltham, MA, USA). Data were expressed as deviation or Z-score to allow comparison with age and gender. For reference values of BMD we used data from Zanchetta et al.

**Statistics**

Z-scores were calculated for bone mineral density measurements via the following formula: 

\[ Z-score = \frac{\text{patient’s measurement} - \text{population mean}}{\text{standard deviation (SD)}} \]

For the statistical analysis was used the software STAT-OF (OK-USA). Mann-Whitney U tests were used to compare two independent samples with a skewed distribution. Spearman’s correlation coefficient was calculated to test the association between variables with a non-normal distribution. A \( p \) value <0.05 was deemed to be significant.

**Results**

Characteristics of sample are resumed in Table I.
Weigth, Height and Body Fat

A scatter g of weight and height showed that these subjects were underweight (mean 42th percentile) and underheight (mean 17th percentile) compared with reference data (Table I). The reduction of weight and height was significantly \((p<0.01)\) evident in the non-walking group (38th and 10th percentile respectively). Body fat as percentage of body weight was greater than expected, ranging from 13 to 42\%, with no differences between the ambulatory group (26\%) and non-ambulatory one (25\%) (Table II).

Bone Mineral Density (BMD)

Mean Z-score of bone mineral density for patients is shown in Table I. Lumbar Z-score of patients was lower than that in normal sample (-0.8 ± 0.3; \(p<0.08\)). Similarly neck Z-score was significantly lower in MMC patients (-1.4 ± 0.4; \(p<0.01\)).

BMD and Walking Ability

BMD both in normal children and in patients increases with age. We examined the relationship of BMD respect age in boys and girls with MMC separately, but no differences were observed. Regarding walking ability we observed that lumbar and neck Z-scores were significantly lower in non-walking group (mean lumbar Z-score: -1.02 ± 0.89 vs -0.36 ± 0.75; \(p<0.01\); mean neck Z-score: -1.71 ± 0.66 vs -0.69 ± 0.55; \(p<0.001\)) (Figure 1).

BMD and Sport Activity

BMD at lumbar and neck sites was better in patients who practised sport activity in comparison with who did not. In fact, in walking and sport group we observed a better BMD respect only walking group (mean lumbar Z-score: -0.30 ± 0.18 vs -0.66 ± 0.25); similarly results were found at neck site (-0.61 ± 0.36 vs -0.88 ± 0.32 respectively). Also in non-walking group we observed better BMD at neck and lumbar sites in patients practised sport activity (mean lumbar z-score: -1.02 ± 0.52 vs -1.36 ± 0.66; mean neck z-score: -1.55 ± 0.42 vs -1.89 ± 0.61) (Figure 2).

Body Fat and Sport Activity

Correlation between body fat (as percentage of body weight) and sport activity, showed better results in patients practised sport activity respect other patients with the same disability (21\% in patients walking and practising sport respect 27\% in only walking patients; 22\% in non walking and practising sport respect 26\% in the sample of only non-walking) (Figure 3).

Discussion

In this study we assessed BMD and body composition in children with MMC and correlations with walking ability and sport activity. Literature evidenced that patients with MMC had higher risk of osteoporosis and pathologic bone fractures\(^{13-16}\). James\(^7\) reported 44 lower extremity fractures occurring in 22 of 122 patients, and Drennan and Freehafer\(^8\) reported 58 fractures occurring in 25 of 84 children\(^6\). All Authors felt that disuse of the limbs and overall physical inactivity contributed greatly to the increased risk

<table>
<thead>
<tr>
<th>Patients</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender (M/F)</td>
<td>34/26</td>
</tr>
<tr>
<td>Age (mean ± SD)</td>
<td>10.4 ± 4.1</td>
</tr>
<tr>
<td>Site of lesion (L/LS/S)</td>
<td>10/35/15</td>
</tr>
<tr>
<td>Physical ability (walking/non-walking)</td>
<td>22/38</td>
</tr>
<tr>
<td>Sport activity (Practised/non practised)</td>
<td>38/22</td>
</tr>
<tr>
<td>Weight percentile (mean)</td>
<td>42</td>
</tr>
<tr>
<td>Height percentile (mean)</td>
<td>17</td>
</tr>
<tr>
<td>Lumbar Z-score (mean ± SD)</td>
<td>-0.8 ± 0.3</td>
</tr>
<tr>
<td>Neck Z-score (mean ± SD)</td>
<td>-1.4 ± 0.4</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Weight percentile(^{\circ})</th>
<th>Height percentile(^{\circ})</th>
<th>% Body fat</th>
<th>Lumbar Z-score(^{*})</th>
<th>Neck Z-score(^{**})</th>
</tr>
</thead>
<tbody>
<tr>
<td>Walking</td>
<td>55th</td>
<td>28th</td>
<td>26th</td>
<td>-0.36 ± 0.75</td>
</tr>
<tr>
<td>Non walking</td>
<td>39th</td>
<td>10th</td>
<td>25th</td>
<td>-1.02 ± 0.89</td>
</tr>
</tbody>
</table>

Table I. Characteristics of sample (SD: Standard Deviation; L; Lumbar; LS: Lumbosacral; S; Sacral).

Table II. Characteristics of walking and nonwalking groups (\(^{*}p<0.01\); \(^{*}p<0.01\); \(^{**}p<0.001\)).
of bone fractures. In our children population with MMC, we did not have any lower extremity fractures but we evidenced a significant bone mineral density reduction and consequently an higher risk of osteoporosis and pathologic fractures. Many Authors reported that BMD has been correlated with bone strength and incidence of fractures. Our data agree with these studies. In fact, they showed that BMD reduction is more evident at neck site where higher is the risk of pathologic fractures. Level lesion, because is the main cause of walking ability, is a very important cofactor for bone density and for risk of fractures. We have observed different BMD in our subgroups: the non-ambulator group (full-time wheelchair) had worse BMD respect the full-time ambulator (individual who walked autonomously or with tutorial aid). Due to loss of motor function, adolescents with MMC were restricted in their everyday activities, which led to an increasingly sedentary life style. This, in turn, led to a decrease in physical fitness and an increase in body fat. Conversely, MMC children who practised sport activity had a higher BMD and a better Fat Mass respect patients with the same degree of disability. Deficits in lean muscle tissue in children with motor impairments were associated with a high prevalence of obesity and suboptimal physical fitness. Obesity was 2.5 times more evident among individuals with lower extremity disabilities than among those without any disability. Obesity exacerbates problems associated with disability, such as limited ambulation, difficulty in transferring, risk of pressure sores, psychological morbidity, and possible

Figure 1. Correlation between BMD at lumbar and femoral sites and walking ability (*p<0.01; **p<0.001).

Figure 2. Correlation between BMD at lumbar and femoral sites, walking ability and sport activity.
complications during surgery. Several studies have found that risk of morbidity from coronary disease and atherosclerosis was increased among men and women who had been overweight adolescents. Recent scientific information suggests that the most effective interventions to address excess weight and obesity in children should include behavioural modification and education to improve eating habits and increase energy expenditure. Roberts et al. reported that in MMC after the age of four, body cell mass and lean mass tissue become increasingly replaced by adipose tissue. The percentage of body fat was significantly greater in those with high-lesions and mid-lesions, and in those in the non-ambulatory group. Shurtleff et al. concluded that body fat composes a greater proportion of body mass in subjects with MMC in comparison with controls and that excess adipose tissue bears a statistical relationship to the degree of mobility and the lesion level. Dietary habits and caloric intakes should be evaluated in patients whose weight exceeds the 25th percentile for age. The effects of sedentary lifestyle and physical inactivity on bone fractures and BMD have been examined in several studies. James found that in the asymmetrically affected patients, the nonactive or paralytic limb was more likely at risk for fracture compared with the active limb. Lin et al. examined hemiplegic patients with cerebral palsy and found that BMC and BMD of the nonactive limb were 5.6% and 21% lower than measures for the active limb. The present study confirms that MMC patients have a higher risk for obesity and osteoporosis but shows that it is possible during childhood, to improve quality of life reducing risk of coronary disease, atherosclerosis and pathologic fractures, with active lifestyle and particularly by practising sport. Sport activity improves lean mass, reduces fat mass and causes behavioural modification and education of eating habits.

In conclusion, patients with MMC have a higher risk of pathologic fractures due to osteoporosis. We suggest to perform BMD measurement in each child with MMC particularly in those with higher level of lesion and non-walking, particularly at high risk for pathologic bone fractures. To reduce the risk of obesity and pathologic fractures, the patients may practice sport activity which reducing fat mass and improving bone density, self esteem and quality of life.

References


