INTRODUCTION

Primary osteoporosis (POP) is a rare and important problem in childhood that can cause serious skeletal deformities and morbidity.

AIM

- To reveal the spectrum of POP in childhood
- To assess the effectiveness and safety of bisphosphonates in increasing BMD, reducing fractures, and improving quality of life.

METHOD

- Patients with POP and received at least one course of pamidronate (PA) or zolendronic acid (ZA) between 2000 and 2020 were included in the study.
- Patients were divided into two groups as OI and other POP patients.
- We retrospectively enrolled demographic, anthropometric, clinical, laboratory and radiological findings.
- Bone densitometer parameters, activation scores, pain status, deformity status, number of fractures per year were evaluated in all patients at admission, and after the treatment.

RESULTS

All study subjects had normal serum calcium and phosphate levels at the start of the treatment, and no significant changes in these levels were observed during the treatment.

- The activation score increased and the fracture incidence, presence of bone pain and deformity severity score improved significantly.

Table 1: Characteristics of the patients in all groups.

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Pre-Treatment</th>
<th>After Treatment</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (year)</td>
<td>6.95 ± 5.53</td>
<td>11.95 ± 5.55</td>
<td>-</td>
</tr>
<tr>
<td>Gender</td>
<td>F:17 (%54.8)</td>
<td>F:17 (%54.8)</td>
<td>-</td>
</tr>
<tr>
<td>Height SD</td>
<td>2.1 ± 2.2</td>
<td>2.3 ± 2.2</td>
<td>0.03</td>
</tr>
<tr>
<td>BMD Z</td>
<td>98.75 ± 23.88</td>
<td>103.65 ± 22.04</td>
<td>0.74</td>
</tr>
<tr>
<td>Purity Stage</td>
<td>Pubertal(9) (29%)</td>
<td>Pubertal(8) (27%)</td>
<td>-</td>
</tr>
<tr>
<td>Deformity Severity</td>
<td>1.87 ± 0.79</td>
<td>1.16 ± 1.05</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Activation score</td>
<td>2.81 ± 1.47</td>
<td>3.16 ± 1.48</td>
<td>0.24</td>
</tr>
<tr>
<td>Number of Fractures</td>
<td>2.28 ± 0.67</td>
<td>0.28 ± 0.62</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

Figure 1: Distribution of all subjects.

Figure 2: Means of height adjusted BMD Z-score

- All study subjects had normal serum calcium and phosphate levels at the start of the treatment, and no significant changes in these levels were observed during the treatment.
- The activation score increased and the fracture incidence, presence of bone pain and deformity severity score improved significantly.

Table 2: Characteristics of the patients in Osteogenesis Imperfecta (OI) group

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Pre-Treatment</th>
<th>After Treatment</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (year)</td>
<td>5.29 ± 3.97</td>
<td>10.89 ± 5.72</td>
<td>-</td>
</tr>
<tr>
<td>Gender</td>
<td>F:15 (%71.4)</td>
<td>F:15 (%71.4)</td>
<td>-</td>
</tr>
<tr>
<td>Height SD</td>
<td>-2.14 ± 2.66</td>
<td>-2.46 ± 2.69</td>
<td>0.79</td>
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<tr>
<td>BMD Z</td>
<td>93.21 ± 16.93</td>
<td>97.94 ± 14.70</td>
<td>0.26</td>
</tr>
<tr>
<td>Purity Stage</td>
<td>Prepubertal(16) (76.2%)</td>
<td>Prepubertal(8) (38.1%)</td>
<td>-</td>
</tr>
<tr>
<td>Deformity Severity</td>
<td>1.81 ± 0.79</td>
<td>1.00 ± 0.83</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Activation score</td>
<td>2.62 ± 1.53</td>
<td>3.14 ± 1.42</td>
<td>0.03</td>
</tr>
<tr>
<td>Number of Fractures</td>
<td>5.14 ± 0.50</td>
<td>0.19 ± 0.05</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

Figure 3: Medians of height adjusted BMD Z-score of all patients

- Median BMD height adjusted Z score increased from -3.50 (IQR 6.20) (-0.89) to -1.04 (IQR 3.45) (2.85) (p<0.001, 99.9%). The change was the same in OI group (p<0.001, 99.9%).
- Change in lumbar spine (L5) BMD Z-scores and each L1 L2 L3 L4 BMD (g/cm2) evaluated separately. Improvement was shown in POP and OI group (p<0.001).

Figure 4: Medians of height adjusted BMD Z-score of patients with OI

- In terms of treatment regimen, both PA and ZA following ZA therapy caused BMD improvement in POP and OI as shown in the Figure 3, 4.
- Three of all pamidronate treated patients had adverse reactions such as flu-like symptoms, fever and leukopenia. Three of ZA treated patients had same adverse effects flu-like symptoms, fever (n=3). In follow up, no long-term adverse effects were observed with bisphosphonates.

CONCLUSIONS

POP is a heterogeneous problem that most often arises as a result of OI.
Those with OI were diagnosed at an earlier age with severe deformity and fractures. Bisphosphonate treatment (PA or ZA) increased both BMD and improved quality of life in all with POP.

REFERENCES


CONTACT INFORMATION

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