THE USE OF PAMIDRONATE IN PEDIATRIC PATIENTS WITH DISEASE OF OTHER THAN OSTEOGENESIS IMPERFECTA: THE EXPERIENCE OF OUR CENTER

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Background: Bisphosphonate treatment for bone fragility has expanded beyond children with osteogenesis imperfecta (OI) to those with other causes of low bone mass. Pamidronate is effective in a variety of conditions such as Paget's disease, hypercalcaemia of malignancy, osteolytic bone metastasis, steroid-induced osteoporosis and idiopathic osteoporosis. The experience with bisphosphonates treatment other than OI in children is limited although there are a growing number of publications showing their usefulness in several bone and metabolic diseases.

Material and Methods: The intravenous administration of pamidronate in children in our institute is analyzed: 2 Mc Cune-Albright (MCA), 1 Tripple A syndrome (TA), 1 glycogen storage disease (GSD) type 0. Pamidronate 0.5 mg/kg/day,3-consecutive-days were given for 3 months interval (6 mg/kg/years). We have 2 cases (21 and 7 old ages) of fibrous dysplasia due to MCA who had severe bone pain showing remarkable clinical improvement with pamidronate. We used pamidronate treatment 18-aged- male with TA syndrome that had osteoporosis (BMD-Z score was -3.0) and severe back pain. His BMD-Z score was -2.0 after the treatment
We also treated 9 aged-girl with GSD type 0, who had osteopenia (BMD-Z score.-1.2 end of the therapy z score was: -0.5) and chronic bone pain because of restricting diary product in her diet.

All patients had normal levels of calcium, phosphorus, and vitamin D, and proper nutrient intake. No advers effects of pamidronate treatment were identified and subjective skeletal pain diminished in all patients. All children had a positive response to the treatment, with rapid and marked clinical improvement in their mobility

Diagnosis	Age at diagnosis	Sex	Cause of therapy	Age at the beginning of pamidronate	Dose (mg/kg /year)	BMD: after pamidronate (gr/cm3)	Additional symptoms
Mc Cune Albright Syndrome	21 months old	Female	Severe left hip pain	5 $^{4/12}$ years old	6	L1-L4: 0.794 2.1 SD	Fibrous dysplasia, Hyperthyroidism, precoccious puberty, cafe au lait spots
Mc Cune Albright Syndrome	4 $^{9/12}$ years old	Female	Severe right hip and thight pain	11 $^{3/12}$ years old 17$^{4/12}$ years old	6 7.5	L1-L4: 3.6 SD	Fibrous dysplasia, Precoccious puberty,hypophospatemic rickets, hypothroidism
Glycogen storage disease type 0	5 $^{3/12}$ years old	Female	Recurrent ankle fractures (left 2 times, right 1 time)	8 $^{6/12}$ years old	3	L1-L4: -0.5 SD	
Triple A syndrome	2$^{2/12}$ years old	Male	Severe back pain and osteoporosis	17	4.5	L1-L4: 0.974 -2 SD	Adrenal Insufficiency, Achalasia, Alacrimia, and Cortical laminar necrosis

CONCLUSION

Our experience with the use of bisphosphonates in pediatric patients with diseases other than OI. Intravenous bisphosphonates are well tolerated, and reduce the risk of fracture and ameliorate bone related clinical symptoms.