OSTEOGENESIS IMPERFECTA : EVOLUTION OF THE BONE MINERAL DENSITY AND EVALUATION OF FRACTURE RISK UNDER BISPHOSPHONATES

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Background: Osteogenesis imperfecta (OI), or Lobstein’s disease, also known as ‘Glass bone disease’ is a rare genetic disorder predisposing to low bone mass with impaired bone microarchitecture and abnormal quality of bone material, causing fracture susceptibility and bone deformities.

Objectives: The main objective of our work was to study the clinical and para-clinical characteristics, and global management, of adult patients and children with OI in a rheumatology department by comparing them with data from the literature.

The secondary objectives were to study the evolution of the bone mineral density and to evaluate the fracture incidence under treatment by Bisphosphonates.

Methods: This is a cross-sectional, observational, descriptive, analytical and monocentric study of subject with OI followed at Fattouma Bourguiba University Hospital in Monastir, Tunisia.

Results: We collected 13 patients with OI, including 5 children and 8 adults. Seven patients were female and 6 were male, including 12 patients with type IIB and one with type IIB. All adult patients had a family history of OI. For children, it was sporadic and the genetic survey did not reveal similar cases in the family. The mean age of clinical onset of the disease was 9 years and 3 months [0-33 years] with a mean age at diagnosis of 15 years and 7 months [14 months - 40 years]. The clinical manifestations were dominated by osteo-articular symptoms. Chronic mechanical bone pain related to fractures was present in 5 patients. Twelve patients had a history of fracture. These were repeated fractures without trauma or minimal trauma in the majority of cases mainly in the vertebrae and in the distal part of the femur.

Conclusion: For children, it was sporadic and the genetic survey did not reveal similar cases in the family. The mean age of clinical onset of the disease was 9 years and 3 months [0-33 years] with a mean age at diagnosis of 15 years and 7 months [14 months - 40 years]. The clinical manifestations were dominated by osteo-articular symptoms. Chronic mechanical bone pain related to fractures was present in 5 patients. Twelve patients had a history of fracture. These were repeated fractures without trauma or minimal trauma in the majority of cases mainly in the vertebrae and in the distal part of the femur.
Background: Scientific interest in the clinical aspects surrounding vitamin D has increased exponentially in recent years. Unfortunately, this interest is not currently associated with a proportional level of consensus. Indeed, the large number of studies in this field, which are often of low quality and secondary importance has confused even experts and driven them towards controversial positions.

Objectives: To shed light on this topic, we arranged a meeting with the aim of collecting the opinions of 50 experts in different specialties (‘D. Battito Group’), including internists, endocrinologists, rheumatologists, pediatricians, geriatricians, dermatologists, gynecologists and nephrologists. This meeting dealt with specific questions regarding the management of hypovitaminosis D and aimed to investigate the opinions of Italian experts on this topic. Six key questions were addressed in the meeting. Methods: After a short lecture and a 30 minutes discussion, all the experts expressed their opinions, which were recorded together with any specific commentary.

Results: The results of the meeting demonstrated the presence of insufficient agreement on many key questions regarding the management of vitamin D deficiency, even among clinical experts.

Conclusion: We hope that the various scientific societies will recognize and analyze the inconsistencies in their own positions and reach adequate consensus, especially on the tenets required for conducting proper and useful investigations on the topic, and that the results of these future new studies will be able to shed the needed light.


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