

Bone Mineralization in Children with Wilson's Disease

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Abstract : Wilson disease, or hepatolenticular degeneration, is an autosomal recessive disease that results in excess copper buildup in the body. It primarily affects the liver and basal ganglia of the brain, but it can affect other organ systems. Musculoskeletal abnormalities, including premature osteoarthritis, skeletal deformity, and pathological bone fractures, can occasionally be found in WD patients with a hepatic or neurologic type. The aim was to assess the prevalence of osteoporosis and osteopenia in Wilson's disease patients. This case-control study was conducted on ninety children recruited from the inpatient ward and outpatient clinic of the Paediatric Hepatology, Gastroenterology, and Nutrition department of the National Liver Institute at Menofia University, aged from 1 to 18 years. Males were 49, and females were 41. Children were divided into three groups: (Group I) consisted of thirty patients with WD; (Group II) consisted of thirty patients with chronic liver disease other than WD; (Group III) consisted of thirty age- and sex-matched healthy. The exclusion criteria were patients with hyperparathyroidism, hyperthyroidism, renal failure, Cushing's syndrome, and patients on certain drugs such as chemotherapy, anticonvulsants, or steroids. All patients were subjected to the following: 1- Full history-taking and clinical examination. 2- Laboratory investigations: (FBC,ALT,AST,serum albumin, total protein, total serum bilirubin,direct bilirubin,alkaline phosphatase, prothrombin time, serum critine,parathyroid hormone, serum calcium, serum phosphorus). 3-Bone mineral density (BMD, gm/cm²) values were measured by dual-energy X-ray absorptiometry (DEXA). The results revealed that there was a highly statistically significant difference between the three groups regarding the DEXA scan, and there was no statistically significant difference between groups I and II, but the WD group had the lowest bone mineral density. The WD group had a large number of cases of osteopenia and osteoporosis, but there was no statistically significant difference with the group II mean, while a high statistically significant difference was found when compared to group III. In the WD group, there were 20 patients with osteopenia, 4 patients with osteoporosis, and 6 patients who were normal. The percentages were 66.7%, 13.3%, and 20%, respectively. Therefore, the largest number of cases in the WD group had osteopenia. There was no statistically significant difference found between WD patients on different treatment regimens regarding DEXA scan results (Z-Score). There was no statistically significant difference found between patients in the WD group (normal, osteopenic, or osteoporotic) regarding phosphorus (mg/dL), but there was a highly statistically significant difference found between them regarding ionised Ca (mmol/L). Therefore, there was a decrease in bone mineral density when the Ca level was decreased. In summary, Wilson disease is associated with bone demineralization. The largest number of cases in the WD group in our study had osteopenia (66.7%). Different treatment regimens (zinc monotherapy, Artamin, and zinc) as well as different laboratory parameters have no effect on bone mineralization in WD cases. Decreased ionised Ca is associated with low BMD in WD patients. Children with WD should be investigated for BMD.

Keywords : wilson disease, Bone mineral density, liver disease, osteoporosis

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