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Commentary

Non-Pharmacological Therapy for Cystic Fibrosis Related Bone Disease

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DESCRIPTION

People with cystic fibrosis who receive highly effective CFTR (Cystic Fibrosis Transmembrane Conductance Regulator) modulator therapy have better quality of life and live longer. Although it is anticipated that the burden of pulmonary illness will decrease, additional long-term effects, such as Cystic Fibrosis Bone Disease (CFBD), could increase the morbidity of CF in the form of fractures, pain, and disability. CFBD's pathogenesis is complex and poorly understood. The treatments must balance the therapeutic advantage with long-term negative consequences as they change during different life stages.

Inadequate bone modeling at this period can have a substantial influence on bone health in adulthood because bone mineral content accretion is strongest throughout childhood and adolescence. The gold standard imaging method for evaluating the health of children's and adults' bones is Dual-energy X-ray Absorptiometry (DXA), which measures both Bone Mineral Content (BMC) and area Bone Mineral Density (aBMD). Between 9 and 32 percent of children and adolescents with CF have low BMD, which is determined by DXA-derived height and age adjusted Z-scores of less than or equal to 2.0 Standard Deviations (SD). Low BMD is linked to poor BMI, low vitamin D levels, and higher disease severity ratings. Low BMD with long bone fractures (up to 2 fractures by age 10 or 3 fractures by age 19) or the presence of a vertebral fragility fracture, regardless of Z-score BMD, are both signs of osteoporosis in this age range. Compared to healthy children and teenagers, small cross-sectional studies show an increased risk of vertebral and rib fracture. While there are now accessible guidelines to help with directing the commencement and monitoring of non-pharmacologic and pharmacologic therapy, fracture risk assessment in this age range is still restricted.

Non-pharmacologic treatments are frequently in line with other health concerns and may be beneficial at preventing CF bone damage. For instance, reducing pulmonary exacerbations is beneficial for CF lung disease and perhaps preventative for CF bone disease. Temporally elevated inflammatory markers such as IL-6, IL-11, and TNF-alpha, elevated levels of N-telopeptide and deoxypyridinoline, and decreased levels of osteocalcin are all associated with CF pulmonary exacerbations, suggesting that the acute inflammatory response to pulmonary exacerbations increases bone resorption and decreases bone formation. Another non-pharmacologic method for promoting bone health is to maintain enough nutrition. Numerous studies show a link between inadequate diet and low aBMD in CF. Low calcium, vitamin D, or vitamin K levels, a lack of protein, and hypogonadism are some of the factors that link malnutrition to low bone mineral density. For CF patients with pancreatic insufficiency, Pancreatic Enzyme Replacement Therapy (PERT) is essential for attaining nutritional goals. Nutritional recommendations provide for 110-200 calories per day, with 35%-40% of those calories coming from fat, in order to maintain a BMI >50% for children and >22 or 23 kg/m^2 for adults, respectively. Achieving prescribed BMI targets is anticipated to improve bone health as well, even though these recommendations were created to enhance pulmonary function. Another non-pharmaceutical method to support bone health in CF patients is physical activity. Pediatric chronic disease affects the healthy bone-muscle unit. Growth and puberty cause a rise in muscle mass and strength, which increases the mechanical strain on the bone and causes a positive change in the bone's dimension and strength.

When treating CF pulmonary exacerbations, glucocorticoids can have a deleterious influence on bone health. By suppressing sex hormones and reducing IGF-1 synthesis, among other mechanisms, glucocorticoids promote bone resorption and prevent bone formation. Numerous studies have shown that taking glucocorticoids reduces BMD, but the severity of the disease can mask this association. BMD among PCF is likely to be preserved by avoiding glucocorticoid medication and using the lowest therapeutic dose when necessary.

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