

## Optimizing Bone Health in Cystic fibrosis: Success is the Sum of Small Efforts

\*SONAL KANSRA AND NOREEN WEST

*From the Sheffield Children's Hospital, Sheffield, UK. \*sonal.kansra@sch.nhs.uk*

**S**urvival in cystic fibrosis (CF) has improved vastly. While proactive treatment of lung disease and malabsorption remain the cornerstones of CF care; recognition and interventions in other aspects of this multisystem disorder can allow these patients to achieve a good quality of life and improve all round general health. Bone disease is probably one of the less well recognized complications of CF, particularly in childhood. Most guidelines recommend screening for CF-related bone disease only towards the second decade of life.

CF-related bone disease is multifactorial and related to direct effects from pancreatic insufficiency resulting in suboptimal nutrition – including vitamin D deficiency – and indirect effects such as raised inflammatory cytokines, glucocorticoid use, insulin deficiency and overt diabetes, and limitation of exercise. The Cystic fibrosis transmembrane conductance regulator (CFTR) chloride channel dysfunction has also been postulated to directly affect the bone structure. CF genotype-phenotype correlations can be variable but inevitably those genotypes conferring more severe disease are associated with a higher risk of bone disease. An imbalance in the bone metabolic state characterized by a mismatched turnover between osteoblast activity and osteoclastic activity is thought to lead to CF-related bone disease [1]. These changes are incremental over the several decades, and may then lead to fractures and kyphosis, and could preclude lung transplantation [2].

In this issue of *Indian Pediatrics*, Gupta, *et al.* [3] present findings of a cross-sectional study comparing bone mineral density (BMD) of children with CF and healthy controls. The study highlights a lower BMD in early life in a cohort of patients of CF, who were diagnosed late and with moderate to severe lung disease. While low lung function is known to have a negative

correlation with BMD, almost a third of these children were also reported to be malnourished, a factor which has a straight impact on BMD. A large proportion of children with CF in the study had low Vitamin D levels known to directly affect bone mineralization. Children with CF had lower activity levels as compared to other studies, and a high proportion of children were taking inhaled steroids – other factor that can affect BMD. The study reflects the multifactorial contributors to low BMD in children with CF as well as the real-world challenges in holistic management of these children.

Although CF-related bone disease requiring treatment is unlikely to be manifest in children and access to screening tools like dual energy X-ray absorptiometry (DXA) scans can be limited; its awareness and optimization of nutritional goals can defer symptomatic bone disease by several years. This study also would support an argument for aggressive prevention of vitamin D deficiency, encouraging an active lifestyle, and judicious use of glucocorticoids, all of which are relatively inexpensive interventions, which may promote bone health. Overall small increments in better management of CF-related conditions can translate to better life expectancy and quality of life for these patients.

*Funding:* None; *Competing interests:* None stated.

### REFERENCES

1. Stalvey MS, Clines GA. Cystic fibrosis-related bone disease: insights into a growing problem. *Curr Opin Endocrinol Diabetes Obes.* 2013;20:547-52.
2. Gore AP, Kwon SH, Stenbit AE. A roadmap to the brittle bones of cystic fibrosis. *J Osteoporos.* 2010;2011:926045. doi. 10.4061/2011/926045.
3. Gupta S, Mukherjee A, Khadgawat R, Kabra M, Lodha R, Kabra SK. Bone mineral density of Indian children and adolescents with cystic fibrosis. *Indian Pediatr.* 2017;54:545-9.