Cystic Fibrosis our focus

Cystic fibrosis and bone health

Factsheet – August 2015

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Introduction

As we get older our bones become thinner and weaker, and may become more susceptible to fracture. However, in people with cystic fibrosis (CF), thinning of the bones can occur at an earlier age. This factsheet describes how bone health can be affected by CF and the implications of this, how bone strength is measured, and options for preventing and treating thinning bones.

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Osteopenia and osteoporosis

Osteopenia and osteoporosis are terms that mean bones are ‘thinner’ than they should be. A more scientific description is that the skeleton has a low bone mass. This means that the bones are more fragile and more likely to fracture.

What is Bone Mineral Density?

Bone mineral density (BMD) is a measure of bone mineralisation – the level of minerals contained in bone. The measurements are an indicator of the bone mass and therefore how strong the bones are.

What are BMD ‘Z’ and ‘T’ scores?

BMD measurements are scored according to how much they fall below or above the population average value. BMD results can be reported as Z- or T-scores, but Z-scores are usually the most appropriate method in people with cystic fibrosis. The Z-score compares a measured BMD value with the average value obtained from a healthy population of the same sex and age as the patient. Generally speaking, BMD is considered low in people with CF when the BMD Z-score in the spine or hip is lower than –2. The T-score compares the measured BMD value to the average value obtained from a healthy young adult population; BMD is highest in young adults.

What happens in normal bones?

- The skeleton provides a rigid framework, a site for the attachment of muscles, protects internal organs and is important in maintaining normal body calcium and phosphate levels.
- Large amounts of bone minerals are acquired in childhood and especially in adolescence during the pubertal growth spurt. Around 90% of adult bone mineral is acquired by late adolescence with peak bone mass reached between 20 to 30 years of age. We usually start to lose bone from about 40 years of age.
- Old bone is always being removed (bone resorption) and replaced by new bone (bone formation). About 10% of adult bone is replaced by this process each year.

What about people with CF?

People with CF are at risk of early thinning of the bones. BMD is usually normal in well-nourished children with CF who have good lung function. Even so, many patients fail to gain bone normally, or experience premature bone loss, in adolescence. About one third of adults with CF have low BMD, which may predispose them to bone fractures.

How is BMD measured?

BMD is usually measured by dual energy X-ray absorptiometry (DEXA).

DEXA absorptiometry

DEXA is used to measure BMD in the lumbar spine (below the chest and above the pelvis), the top of the leg (hip), the wrist, and the whole body. The scan at each site takes approximately two to three minutes and involves a low radiation dose, less than the dose of naturally occurring background radiation and much less than that needed for a chest x-ray. The BMD is scored and Z and T values calculated. It is important to note that the BMD score may not accurately predict the fracture risk in people with cystic fibrosis. A DEXA scan should be performed from about 10 years of age and repeated every one to three years, the frequency determined by clinical need. This should be organised by your CF centre and the results interpreted in liaison with the DEXA scanning department.

What causes low BMD in cystic fibrosis?

CF-related low BMD can have many causes:

1. Bone health is strongly linked to the severity of lung disease and body weight. Almost all severely ill patients have low bone mineral density. Conversely, patients with normal weight and height, and good lung function, have near normal bone density. Lung infection in CF is associated with more intense inflammation and raised levels of chemicals called cytokines in blood and sputum. These cytokines increase the activity of cells which break down bone.

2. Apart from the general malnutrition associated with more severe disease, many aspects of nutrition affect bone status, including vitamins (eg vitamins D and K), minerals (eg calcium) and protein intake.

3. Steroids cause rapid bone loss, especially during the first year of therapy. They reduce calcium absorption from the gut, increase calcium loss in the urine, decrease the number of bone-forming cells and increase bone resorption. Most of the larger studies in individuals with CF have shown an association between oral steroid use and low bone mineral density.

4. A number of studies in people who do not have CF have found an association between greater physical activity and higher BMD levels. However, it is not known if weight-bearing exercise in people with CF can increase peak bone mass, preserve BMD or increase it in those with low bone mineral density.

5. Despite most children with CF achieving normal or near normal growth, puberty is often delayed. Bone mineral deficiencies resulting from late onset of puberty may not be corrected when puberty eventually starts. Low sex hormone levels, oestrogen and testosterone, are associated with low BMD in adults with cystic fibrosis.

6. Low BMD has been associated with CF-related diabetes.

7. There may be a direct link between CF-related low BMD and the abnormal protein produced by the CF gene.

Why is it important to prevent fractures?

Fractures in the spine and ribs are painful and therefore may interfere with effective physiotherapy and airway clearance, resulting in a decline in lung function.

How can low BMD be prevented and treated?

- People with CF should be screened for low BMD with regular DEXA scans and chest X-rays. The latter should be examined specifically for vertebral fractures. Risk factors that can be treated, such as low vitamin D or calcium levels, should be monitored and low levels corrected with extra dietary supplements.
- Lung infection damages bone health. Therefore treatments that prevent the progression of lung disease should be optimised.
- Optimising nutrition through dietary intervention, and specialist dietetic supervision of appropriate and proactive interventions with oral supplements and nasogastric or gastrostomy tube feeds is fundamental to care. Vitamin D and vitamin K status and calcium intake should be optimised through frequent contact with a specialist CF dietitian. Vitamin D and calcium levels should be measured at least annually. There is not yet sufficient evidence to recommend universal vitamin K supplementation for bone health in CF, but supplementation should be considered in certain clinical situations, eg for patients with a low BMD or liver disease. Patients should have access to a specialist dietitian at each admission, outpatient review and annual assessment.
- Weight-bearing physical activity should be encouraged.
- A specialist CF physiotherapist should develop an exercise programme where appropriate for an individual patient's abilities and needs.
- Glucocorticoid treatment (prednisolone and inhaled steroids) should be kept to a minimum.
- Delayed puberty and low blood levels of testosterone and oestrogens should be looked for and treated.
- As long-acting 'depot' and possibly other progesterone-only contraception preparations for women may reduce BMD, particularly in adolescents, alternative contraceptive preparations may be suggested.
- Smoking and alcohol can have damaging effects on bone health and should be avoided.

Are there any specific drug treatments?

- Bisphosphonates are a family of drugs that reduce bone breakdown. Non-CF studies have shown benefit in the treatment of postmenopausal osteoporosis, osteoporosis in males and glucocorticoid-induced osteoporosis. There is also increasing evidence for their efficacy in adults with cystic fibrosis.
- Vitamin D levels should be optimised before using a bisphosphonate.
- Bisphosphonates should not be used during pregnancy and should therefore be used with caution in young women. If you are considering starting a family, let your team know before starting to take a bisphosphonate.
- Bisphosphonates can be given orally or intravenously.
- Oral bisphosphonates are best absorbed in the absence of food and should be taken in the fasting state. This may be difficult for people with CF-related diabetes or for those who feed overnight. Calcium supplements impair the absorption of bisphosphonates and the two should not be taken at the same time. If you have been prescribed both, take your calcium at a different time of day (at least two hours after the bisphosphonate).
- Oral bisphosphonates can cause ulceration in the oesophagus and so should be taken with a full glass of water and people should remain upright for 30 minutes after taking the drug.
- Bisphosphonates can very rarely be associated with a condition called osteonecrosis of the jaw. This occurs most commonly in people with significant dental problems (infection or need for invasive treatment such as extraction or implant) and in those prescribed intravenous bisphosphonates. People with dental problems should see their dentist before starting bisphosphonate treatment.
- Use of intravenous and oral bisphosphonates can be associated with bone pain in people with CF and it occurs more frequently in people with unstable lung disease. Experience suggests that the frequency/severity of bone pain can be reduced by starting bisphosphonate treatment at the end of a course of intravenous antibiotics or following a short course of oral steroids.
- Bisphosphonates are not licensed for use in children and although they appear to be relatively safe even when used for long periods in other paediatric bone disease, experience is limited.
- We recommend that treatment of children with bisphosphonates is supervised by a major CF centre or by a paediatric bone specialist.

Further reading

The Cystic Fibrosis Trust has published a consensus document entitled Bone Mineralisation in Cystic Fibrosis. This can be downloaded from our website or hard copies can be obtained by contacting the Cystic Fibrosis Trust Helpline.

Useful wesbites:

National Osteoporosis Society
www.nos.org.uk

National Osteoporosis Foundation
www.nof.org

National Institute of Health
www.niams.nih.gov/Health_Info/Bone

Further information

The Cystic Fibrosis Trust provides information about cystic fibrosis through our factsheets, leaflets and other publications.

Most of our publications are available through our helpline and can be downloaded from our website or ordered using our online publications order form. Visit cysticfibrosis.org.uk/publications.

The Cystic Fibrosis Trust helpline can help you with a range of issues, no matter how big or small. Our trained staff can provide a listening ear, practical advice, welfare/benefits information or direct you to other sources of support. The helpline can be contacted on 0300 373 1000 or helpline@cysticfibrosis.org.uk and is open Monday to Friday, 9am – 5pm.

Calls to 0300 numbers cost no more than 5p per minute from a standard BT residential landline. Charges from other landlines and mobile networks may vary, but will be no more than a standard geographic call and are included in all inclusive minutes and discount schemes. If you are worried about the cost of the call please let us know and we'll call you back.

You can also find more information at our website cysticfibrosis.org.uk.

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More factsheets available at:
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