Cystic Fibrosis our focus

Cystic fibrosis and bone health

Factsheet – November 2020

Fighting for a Life Unlimited

Cystic fibrosis and bone health

Introduction

As we get older our bones become thinner and weaker, and may be more likely to fracture. In people with cystic fibrosis (CF) this can happen at an earlier age. This factsheet describes how CF can affect bone health, what this means for people with the condition, how bone strength is measured, and ways to prevent and treat thinning bones.

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Contents

Osteopenia and osteoporosis

Osteopenia and osteoporosis mean that 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 likely to fracture.

What is bone mineral density?

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

What are bone mineral density 'Z' and 'T' scores?

BMD measurements compare your bone density with the bone density expected for a young, healthy adult or a healthy adult of your own age, gender and ethnicity. BMD results can be reported as 'Z-scores' or 'T-scores,' but Z-scores are usually the most appropriate method in people with cystic fibrosis.

- Z-scores are the difference between your measurement and that of a healthy person of the same age as you who doesn't have cystic fibrosis.
- If your Z-score is lower than -2 in the spine or hip, it is considered low.
- T-scores are the difference between your measurement and that of a young healthy adult. This is because BMD is highest in young adults.

Being shorter than average can mean that you get incorrectly diagnosed with low BMD. It may be useful to discuss this with your team.

How do bones develop?

The skeleton acts like a frame. Muscles are attached to it, it protects internal organs and it helps to maintain the body's normal calcium and phosphate levels.

Bones get most of their minerals in childhood and especially during puberty, when children grow more quickly. By the time they are in their late teens, humans have about 90% of the total bone mineral they will have as an adult. Humans will reach peak bone mass between 20 to 30 years of age. They will usually start to lose bone mass from about 40 years of age.

Old bone is always being removed. This is called bone resorption. When this happens, it is replaced by new bone. This is called bone formation. About 10% of adult bone is replaced by this process each year.

How are bones affected by cystic fibrosis?

People with CF might experience early thinning of the bones. BMD is usually normal in children with CF with a healthy BMI and lung function. Even so, many people with CF fail to gain bone normally or experience premature bone loss when they are teenagers.

About one third of adults with CF have low BMD, which may mean they are more likely to get bone fractures.

How is bone mineral density measured?

BMD is usually measured by 'dual energy X-ray absorptiometry' (DEXA) scans. DEXA scans are 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 part of the body takes approximately two to three minutes and involves a low amount of radiation. The amount of radiation someone getting a scan receives is much less than what they might receive when getting a chest X-ray.

In a DEXA scan, BMD measurements are taken and Z and T scores calculated.

DEXA scans should be performed from when someone with CF is about 10 years old. They should be repeated every one to three years, depending on the health of the individual. Your CF team will organise these scans, and will work with the DEXA scanning department to understand the results.

It is important to note that Z and T scores may not accurately predict the fracture risk in people with cystic fibrosis.

What causes low bone mineral density in cystic fibrosis?

There are many reasons that people with CF might have low BMD:

Poor overall health

Bone health is strongly linked to body weight and lung function. Almost all people with CF who are very unwell have low BMD. Almost all people with CF who are a healthy weight, average height, and have good lung function have near normal BMD.

Lung infection in CF can cause inflammation, which leads to an increase in inflammation blood proteins. These proteins increase bone loss.

Poor nutrition

Poor nutrition can also lead to lower levels of vitamins D and K, calcium and protein, which can all cause low BMD.

Steroids

Steroids cause bone loss to happen more quickly, especially during the first year of taking them. They make it harder for the body to absorb calcium, increase calcium loss in the urine, decrease the number of bone-forming cells and increase bone resorption. Studies have shown a link between using oral steroids and having low BMD in people with cystic fibrosis.

Low physical activity

A number of studies in people who do not have CF have found a link between greater physical activity and higher BMD levels. However, it is not known if weight-bearing exercise, like walking, running and climbing, in people with CF can increase peak bone mass, preserve BMD or increase it in those with low BMD.

Delayed puberty

Children with CF continue to show an improvement in final adult height but often do not reach their growth potential, which can impact BMD.

Delayed puberty also contributes to changes in BMD in children with CF but this is now far less common. Bone mineral deficiencies resulting from puberty starting late might not be corrected when puberty eventually starts.

Low levels of the sex hormones oestrogen and testosterone are associated with low BMD in adults with cystic fibrosis.

CF-related diabetes

Low BMD has been associated with CF-related diabetes.

The CF gene

There may be a direct link between 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 can make it difficult to do physical activity and airway clearance. If someone with CF stops doing these things, it could cause their lung function to get worse.

How can low bone mineral density be prevented and treated?

- People with CF should be screened for low BMD at regular DEXA scans and chest X-rays. Chest X-rays should particularly look for vertebral fractures (fractures to the bones in the spine).
- Low vitamin D or calcium levels can cause low BMD, and should be monitored. Low levels should be corrected with extra dietary supplements. There is not yet sufficient evidence that taking vitamin K supplements improves bone health in people with CF, but they should be considered for people with low BMD or liver disease.
- Nutrition can be improved through food supplements taken orally or through nasogastric or gastrostomy tubes.
- People with CF should have access to a specialist dietitian when they are admitted to hospital, and at outpatient reviews and annual assessments.
- Lung infection damages bone health. Therefore, treatments that prevent the progression of lung disease should be optimised.
- Weight-bearing physical activity like walking, running and climbing should be encouraged.
- A specialist CF physiotherapist should work with the person with CF to encourage appropriate physical activities that will promote good bone health.
- Glucocorticoid treatment (prednisolone and inhaled steroids) should be kept to a minimum.
- Delayed puberty and low blood levels of testosterone and oestrogen should be looked for and treated.
- Contraceptive injections and progesterone-only contraception may reduce BMD, particularly in teenagers. Therefore, alternative contraception may be suggested.
- Smoking and alcohol can cause poor bone health and should be avoided.

How do bisphosphonates help low bone mineral density?

What are bisphosphonates?

Bisphosphonates are drugs that reduce bone breakdown. Studies in the general population have shown these drugs to help with the treatment of postmenopausal osteoporosis, osteoporosis in men and glucocorticoid-induced osteoporosis. There is also increasing evidence that they may be effective in adults with cystic fibrosis.

How are bisphosphonates taken?

Bisphosphonates can be given orally or intravenously (through IVs).

Oral bisphosphonates should be taken first thing in the morning, on an empty stomach and at least 30 minutes before any food, drink or other medicines. This may be difficult for people with CF-related diabetes or for those who have overnight feeds.

Vitamin D

Vitamin D levels should be optimised before taking bisphosphonates.

Calcium supplements

Calcium supplements should not be taken with bisphosphonates, as they may stop them from working. If you have been prescribed both, take your calcium at a different time of day (at least two hours after the bisphosphonate).

Pregnancy

Bisphosphonates should not be used during pregnancy. If you are considering becoming pregnant, let your CF team know before starting to take bisphosphonates.

Ulcers

Oral bisphosphonates can cause ulcers in the oesophagus and so should be taken with a glass of water. You should remain upright for 30 minutes after taking the drug.

Osteonecrosis of the jaw

Though it is very rare, bisphosphonates can cause osteonecrosis of the jaw (where the cells in the jaw bone die). This is most common in people with significant dental problems like infections, or who need invasive treatments like extractions or implants, and in those prescribed IV bisphosphonates. People with dental problems should see their dentist before starting to take bisphosphonates.

Bone pain

IV and oral bisphosphonates can cause bone pain in people with CF, especially those with poor lung function. This may be reduced by starting bisphosphonate treatment at the end of a course of IV antibiotics or following a short course of oral steroids.

Use in children

Like many drugs, bisphosphonates are not licensed for use in children. They are safely used for long periods in other bone disease experienced in children, but they are rarely needed in children with CF and would only be given under the guidance of experts in bone disease.

Further reading

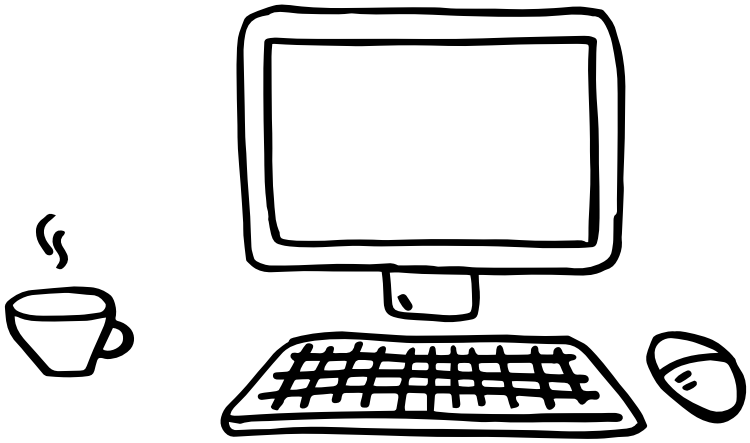
The Cystic Fibrosis Trust has published a consensus document entitled Bone Mineralisation in Cystic Fibrosis. This can be downloaded from our website or hard copies can be obtained by contacting the Cystic Fibrosis Trust Helpline. Details of our Helpline can be found on the back page of this booklet.

Useful websites:

Royal Osteoporosis Society **www.theros.org.uk**

National Osteoporosis Foundation **www.nof.org**

National Institute of Health **www.bones.nih.gov**



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 and benefits information or direct you to other sources of support. The Helpline can be contacted on 0300 373 1000 or by emailing **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:
cysticfibrosis.org.uk/publications