Bone Health and Cystic Fibrosis (CF)

As we get older our bones become thinner and weaker. Thinning of the bones can occur at an earlier age for people with Cystic Fibrosis (CF).

**Osteopenia and osteoporosis**
Osteopenia and osteoporosis are terms that mean bones are “thinner” than they should be. This means that the bones are more fragile and more likely to fracture.

**What is bone mineral density (BMD)?**
Bone mineral density (BMD) is a measure the level of minerals the bones contain. This shows how strong the bones are.

BMD is usually measured by dual energy x-ray absorptiometry (DEXA) scan. The low radiation dose used in the DEXA scan is less than that needed for a chest x-ray.

The scan is taken at these sites on the body for approximately two or three minutes:
- Lumbar spine (below the chest and above the pelvis).
- Top of the leg (hip).
- Wrist.
- Whole body.

It is important to note that the BMD score may not accurately predict the fracture risk in people with CF.

A DEXA scan should be performed from about ten years of age and repeated every one to three years. Check with your doctors when the next one is due.

**What are bone mineral density “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-scores or T-scores.
- The Z-score compares a measured BMD value to the average value obtained from a healthy population of the same sex and age as the patient. Z-scores are usually the most appropriate method in people with CF.
- The T-score compares the measured BMD value to the average value obtained from a healthy young adult population.

**How do I know what these values mean?**
BMD is considered very low in people with CF when the Z-score in the spine or hip is lower than –2.

**What causes low bone mineral density in CF?**
CF-related low BMD probably has many causes:
- Severe lung disease and recurrent lung infections
- Poor nutrition (low body weight, low levels of minerals (e.g. calcium) and vitamins (e.g. vitamin D and K)
- Certain medications (e.g. steroids)
- Delayed puberty
- CF related diabetes

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

**How can low BMD be prevented and treated?**
- Regular DEXA scans to screen for low BMD.
- Optimise lung function and prevent lung infections.
- Optimise nutrition through high energy diets, oral supplements and nasogastric or gastrostomy tube feeds.
- Vitamin D or calcium levels should be checked yearly and low levels corrected with extra dietary supplements.
- Frequent contact with a specialist CF dietitian to gain advice on how best to boost nutrition for bone health.
- There is not yet sufficient evidence to recommend universal vitamin K supplementation for bone health in CF. However, supplementation should be considered in certain clinical situations. Always check with your doctor.
• Weight bearing physical activity is encouraged. A specialist CF physiotherapist can develop an exercise programme where appropriate. This will depend on an individual patient’s abilities and needs.
• Minimise steroid treatment as advised by your doctors.
• Detection and treatment of delayed puberty and low levels of testosterone and oestrogens

Long acting “depo” and some other progesterone-only contraception methods may reduce BMD, particularly in adolescents. Other methods of contraception may be suggested by your doctors.

• Avoid smoking and alcohol which can have damaging effects on bone health.

How much calcium do I need?
Calcium intake in individuals with CF should meet the Recommended Daily Intakes (RDI) for the general population. It should be increased to 1500mg/day for those with low Bone Mineral Density.

<table>
<thead>
<tr>
<th>Category</th>
<th>RDI* (mg/day)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Girls (14yrs -18 yrs)</td>
<td>1300</td>
</tr>
<tr>
<td>Women (19yrs -50yrs)</td>
<td>1000</td>
</tr>
<tr>
<td>Pregnant women (19yrs - 50yrs)</td>
<td>1000</td>
</tr>
<tr>
<td>Breastfeeding mothers (19yrs - 50yrs)</td>
<td>1000</td>
</tr>
<tr>
<td>Boys (14yrs – 18yrs)</td>
<td>1300</td>
</tr>
<tr>
<td>Men (19yrs -70yrs)</td>
<td>1000</td>
</tr>
<tr>
<td><strong>Men &amp; Women with CF with low BMD</strong></td>
<td><strong>1500#</strong></td>
</tr>
</tbody>
</table>

* Based on Nutrient Reference Values for Australia & NZ, NHMRC & NZMOH, 2006
# Based on Australasian Clinical Practice Guidelines for Nutrition in Cystic Fibrosis, 2006

Highest calcium (at least 400mg per serve)
• 250mL (1 cup) calcium fortified milk (e.g. Physical, Lite Start, Shape, Pura tone)
High calcium (at least 300mg per serve)

- 250mL (1 cup) full cream / trim / or skim milk
- 250mL (1 cup) calcium fortified soy milk
- 200g tub yoghurt
- 100g tin salmon with bones or sardines
- 2 slices (40g) of hard cheese
- 250mL (1 cup) custard

Medium calcium (at least 130mg per serve)

- 1 cup of ice-cream (full cream or low fat)
- ½ cup of almonds
- 150g tofu

Low calcium (less than 130mg per serve)

- meats and eggs
- baked beans
- fruit e.g. oranges
- bread and cereals
- soft cheeses e.g. fetta, cottage, ricotta
- green leafy vegetables (e.g. broccoli, spinach)

Vitamin D
Vitamin D is an essential ingredient for maintaining a healthy body. It helps maintain muscle and bone strength and allows your body to absorb calcium.

Sources of vitamin D
The major source of vitamin D in Australia is exposure to sunlight. While small amounts are derived from dietary sources such as oily fish (e.g. Salmon), eggs and fortified foods such as margarine and some milks.

Monitoring & supplementation
Vitamin D levels are screened through a blood test annually. To prevent deficiency, the minimum recommended Vitamin D level is 75 nmol/l.

All patients with vitamin D deficiency and insufficiency should be prescribed vitamin D supplements. Generally, a supplement of 1000 to 5000 IU/day of vitamin D is used in adults. Check with your Cystic Fibrosis treating team, what your individual needs would be.
Are there any specific drug treatments?
Bisphosphonates (e.g., Aledronate, Residronate) are a family of drugs that reduce bone breakdown and may directly stimulate the cells that produce new bone.

Oral Bisphosphonates are best absorbed in the absence of food and should be taken on an empty stomach. 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 together.

For more information, contact your Dietitian or Cystic Fibrosis Centre.

Acknowledgment to CF Trust 2012 for information provided in this education material