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| **Title** | Osteoporosis and Arthropathy |
| **Version** | 1.1 |



## 1. Osteoporosis

- **Osteopenia and osteoporosis** are more common in CF, and may lead to risk of
    - Long-bone fractures
    - Vertebral fractures causing loss of height, change in posture, back pain, and reduced lung capacity

- **Osteoporosis definitions**
    - In CF children, adolescents, or young adults up to the age of 20 years, osteoporosis is defined as having a BMD Z score < minus2 and a significant fracture history (low trauma fracture of a lower limb long bone, vertebral compression fracture, or two or more upper limb long bone fractures).
    - In postmenopausal women or men over the age of 50 years with CF, osteoporosis is defined as having a BMD T-score ≤ minus 2.5.
    - In younger adults, osteoporosis is defined as having a BMD Z score < minus 2 and a significant fracture history.
    - The term osteoporosis can also be applied to adults with CF who have sustained a low trauma fracture as it is an indicator of increased bone fragility.

- **Patients with CF are more at risk of osteoporosis due to:**
    - **CFTR function influences bone homeostasis**
    - Certain medications e.g. steroids
    - Reduced activity levels
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    - Pancreatic insufficiency, vitamin D deficiency and nutrition
    - CF diabetes

      Lung health
- **Monitoring required (ECFS 2011)**
    - Regular dietetic reviews to include calcium intake assessment and advice as required
    - Regular Physiotherapy reviews to include posture assessment as well as exercise routine
    - Annual Review to measure Vitamin D level and note fracture history
    - Assessment for delayed puberty in children
    - Bone Mineral Density (BMD) assessment by DEXA Scans:

      Children and Adults age <50 years with CF:
        - Age 10 yrs (or sooner if at high risk or has sustained low-trauma fracture)

          Then:
        - 5-yearly if BMD Z score is > minus 1, or
        - 2-yearly if BMD Z score is between minus 1 to minus 2, or
        - Annually if
            - BMD Z score < minus 2, or
            - has experienced low-trauma fracture or
            - significant risk factors for low BMD

      Adults with CF after age 50yrs:
        - 5-yearly if the BMD T-score is > minus 1
        - 2-yearly if BMD Z score is between minus 1 and minus 2.5
        - Annually if significant risk factors for low BMD

- **Management**
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    - Nutritional optimisation, Vitamin D supplementation and calcium supplementation
    - Regular physiotherapy review encouraging weight bearing and reistance activities
    - Bisphosphonate therapy is the most common drug therapy– Rheumatology referral required
    - DEXA scan required prior to starting therapy

**2. Cystic fibrosis arthropathy (CFA)**

- **Incidence:** CFA is the most common form of joint pain in PWCF occurring in up to 10% of the population. It can be associated with significant morbidity.
- **Aetiology:** There is little understanding of aetiology, pathogenesis or sequelae. The prominent hypothesis is the formation of immune complexes from excessive antigen load which itself is secondary to chronic persistent bacterial infection. Thus, chronic respiratory infection results in an excess of immune complexes entering the systemic circulation and depositing within the joints. However, the degree of pulmonary disease is often discordant with the joint involvement.
- **Symptoms and Signs:**
    - may vary from a minimally swollen joint to a polyarthritis with swollen, tender joints resembling those seen in rheumatoid arthritis (RA).
    - CFA can follow a palindromic pattern with a remitting and relapsing course and symptoms mostly disappearing between attacks.
    - CFA can affect any joint and while symptoms may be self-limiting, they can also be severe, persistent, and difficult to manage. Arthropathy may also be associated with joint effusions, a flu like illness and in some cases a vasculitic rash.
    - Joint disease in patients with CF may also be associated with adverse drug reactions and gout.
- **Investigation:**
    - Clarification of the diagnosis with a detailed history, joint examination and serology is essential when assessing individuals with CF and joint symptoms.
    - CFA remains a diagnosis of exclusion and other causes fo arthritis such as drug-induced arthropathy, crystal arthritis such as gout and specific inflammatory arthritides such as rheumatoid arthritis need to be considered in the differential diagnosis.
- **Treatment**
    - Most patients will respond rapidly to a non-steroidal anti-inflammatory.
    - If they do not relieve symptoms, then a low dose of prednisolone such as 10mgs per day can be trialled.
    - If these measures fail to improve symptoms, then a rheumatology opinion should be sought.
    - Symptoms also often improve with resolution in chest symptoms.

**3. Hypertrophic pulmonary osteoarthropathy**

- This rarer condition predominantly affecting young adults is characterised by chronic periosteal inflammation of the long bones with or without new bone formation.
- Usually presents with swelling and pain effusions of the knees, ankles and wrists and tenderness and pain over the long bones.
- Associated with a more severe respiratory history and typically worsens during exacerbations.
- Its cause remains unknown.

**4. Other Joint Diseases**

- Gout, sarcoidosis, amyloidosis, and rheumatoid arthritis have all been reported in cystic fibrosis.
- The most common drug induced arthropathy is quinolones causing tenosynovitis of the knees and ankles and patients should be counselled regarding this potential side effect and advised to discontinue treatment and seek medical advice should it occur.

**5. References**

European cystic fibrosis bone mineralisation guidelines. Journal of Cystic Fibrosis Volume 10 Suppl 2 (2011) S16–S23

Fitch G, Williams K, Freeston JE, Dass S, Grainger A, Hodgson R, Horton L, Whitaker P, Peckham D. Ultrasound and magnetic resonance imaging assessment of joint disease in symptomatic patients with cystic fibrosis arthropathy. Journal of Cystic Fibrosis. 2016 Jul 1;15(4):e35-40.

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