Monitoring people with cystic fibrosis

**Monitoring**

NICE recommends that each person with cystic fibrosis is regularly reviewed by a multidisciplinary team at a specialist cystic fibrosis centre.

- Specialist paediatrician or adult physician
- Specialist nurses
- Specialist physiotherapists
- Specialist dietitians
- Specialist pharmacists
- Specialist clinical psychologists

The team should also have access to other professionals, including social workers.

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**Routine reviews**

Regular reviews are critical to prevent or limit symptoms and complications.

**Example schedule**

<table>
<thead>
<tr>
<th>Age</th>
<th>Frequency</th>
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</thead>
<tbody>
<tr>
<td>0–1 month</td>
<td>weekly</td>
</tr>
<tr>
<td>1–12 months</td>
<td>every 4 weeks</td>
</tr>
<tr>
<td>1–5 years</td>
<td>every 6–8 weeks</td>
</tr>
<tr>
<td>5+ years</td>
<td>every 8–12 weeks</td>
</tr>
<tr>
<td>Adults</td>
<td>every 3–6 months</td>
</tr>
</tbody>
</table>

More frequent reviews are needed for those with evidence of lung disease.

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**Annual reviews**

The team is to offer a comprehensive annual review, focussing on long term management of the condition.

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**Basic assessments**

Carry out at all routine and annual reviews

- **Clinical assessment**
  - Review clinical history
  - Medicines adherence
  - Physical examination
  - Measurement of weight and length or height

- **Respiratory secretion samples**
  - for microbiological investigations
  - If possible: Sputum samples
  - Otherwise: Cough swab or Nasal pharyngeal aspirate (NPA)

- **Lung function testing**
  - Spirometry measurements:
    - FEV1: Forced expiratory volume in 1 second
    - FVC: Forced vital capacity
    - FEF: Forced expiratory flow
  - If spirometry is normal:
    - LCI: Lung clearance index

- **Oxygen saturation**
  - Measure oxygen saturation

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**Additional assessments**

Carry out at annual reviews, and as individually needed in routine reviews, for example in relation to specific concerns for that person.

- **Physiotherapy**
  - Airway clearance
  - Nebuliser use
  - Exercise and physical activity

- **Blood tests**
  - White cell count
  - Serum IgE
  - Aspergillus serology

- **Liver disease**
  - Clinical assessment
  - Liver function blood tests

People with liver disease might require treatment with ursodeoxycholic acid and sometimes referral to a liver specialist.

- **Nutrition and malabsorption**
  - Nutritional assessment
  - Vitamin deficiencies
  - Test for exocrine pancreatic insufficiency

- **Cholesterol changes**
  - More detailed pulmonary assessment is recommended at the annual assessment, to monitor chronic respiratory disease

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**Psychological review**

This may include:

- General mental health and wellbeing
- Quality of life
- Adherence to treatment
- Psychosocial problems
- Behaviours that affect health outcomes

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**CF related diabetes**

Annual testing for:

- Age 10+

Additional tests during:

- Pregnancy
- Enteral feeding
- Long term systemic corticosteroid use

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**Other complications**

Be aware of other potential complications of cystic fibrosis, including:

- More common
  - Distal ileal obstruction syndrome
  - Reduced bone mineral density
  - Infertility
  - Osteoporosis
  - Muscle pains and arthralgia
  - Upper airway complications
  - Nasal polyps
  - Sinusitis
  - Meconium ileus

- Less common
  - Arthritis
  - Delayed puberty
  - Renal calculi

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