thebmj Visual summary

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

Routine reviews

Regular reviews are critical to prevent or limit symptoms and complications

Example schedule

Age	Frequency
0–1 month	weekly
1–12 months	every 4 weeks
1–5 years	every 6–8 weeks
5+ years	every 8–12 weeks
Adults	every 3–6 months

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

Annual reviews

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

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

O_2 Measure oxygen saturation

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

Advice on:

- Airway clearance
- Musculoskeletal disorders
- Nebuliser use
- Urinary incontinence
- 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

Chest x ray changes

More detailed pulmonary assessment is recommended at the annual assessment, to monitor chronic respiratory disease

Nutrition and malabsorption

- Nutritional assessment
- Vitamin deficiencies
- Test for exocrine pancreatic insufficiency

Psychological review

This may include:

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

CF related diabetes

Annual testing for: Age 10+

Additional tests during:

- Pregnancy
- Enteral feeding
- Long term systemic corticosteroid use

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

Data source: National Institute for Health and Care Excellence (2017) Cystic fibrosis: diagnosis and management. Available from https: //www.nice.org.uk/guidance/ng78 NICE guidance is prepared for the National Health Service in England. All NICE guidance is subject to regular review and may be updated or withdrawn. NICE accepts no responsibility for the use of its content in this product/publication.

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