The Practitioner

# CPD study pack

**November 2019;262(1820)**
The study pack contains this month's CPD exercise plus all relevant articles:

- Managing mild to moderate asthma in adults
- Diagnosing and managing cystic fibrosis in children
- Managing complications of cystic fibrosis in adults

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- Journal article
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## CME - FILLABLE PDF FORM

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# Practitioner CPD exercise

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**Show impact**
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- Significant event audit
- Practice meeting
- Implement a new service
- Take on new role
- Significant event audits
- Change with feedback from patients and/or colleagues

**Further options for sessional GPs**
- Self audit of own practice:
  - Record-keeping
  - Referrals for investigations
  - Prospective condition based reviews
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  - Review of telephone triage outcomes
- Update drugs in emergency bag

**CPD Framework**

**Online all articles are associated with CPD prompts/frameworks to written reflection:**

**Adobe fillable PDFs**
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- Case review framework
- Audit framework

**Word documents**
- Reflection on a journal article
- Stages of reflection on a situation
- Team reflection

## MILD TO MODERATE ASTHMA IN ADULTS

**1 Which of the statements are true?**

**A** Reducing exposure to house dust mite improves asthma control
**True** ☐ **False** ☐

**B** A quarter of women with asthma experience worsening symptoms during pregnancy
**True** ☐ **False** ☐

**C** All patients who have suffered a life-threatening asthma attack in the past should be under specialist supervision
**True** ☐ **False** ☐

**D** Inhaler technique should be reassessed at every contact with a health professional
**True** ☐ **False** ☐

**E** Cardioselective beta-blockers may impact on asthma control
**True** ☐ **False** ☐

**2 Match the statistic to the statement. In the National Review of Asthma Deaths, the proportion of patients who:**

**A** Did not receive monthly treatment with inhaled corticosteroids

**B** Died without seeking medical attention or before emergency medical care could be provided

**C** Were not under specialist care at the time of their death

**1** 45%
**2** 50%
**3** 57%
**4** 80%

**3 List four characteristics of patients with well controlled asthma.**

**4 List any changes to your clinical practice that you may make having read this article.**

## CYSTIC FIBROSIS IN CHILDREN

**5 Which of the statements are true?**

**A** Cystic fibrosis is an autosomal recessive disorder
**True** ☐ **False** ☐

**B** Young children with cystic fibrosis are vulnerable to infection with *Staphylococcus aureus*
**True** ☐ **False** ☐

**C** Most infants diagnosed by newborn screening have few symptoms
**True** ☐ **False** ☐

**D** Cystic fibrosis related diabetes becomes prevalent in the first decade of life
**True** ☐ **False** ☐

**E** Distal intestinal obstruction syndrome is a surgical emergency
**True** ☐ **False** ☐

**6 Complete the statement:**

Distal intestinal obstruction syndrome is a serious complication that presents with

and

By ensuring children are adequately

and have the appropriate

, gut transit time is improved and the risk of distal intestinal obstruction syndrome reduced.

**7 Most children with cystic fibrosis have malabsorption requiring pancreatic enzyme replacement therapy.**

**A** What dietary supplements may they also require?

**B** What are the symptoms of malabsorption?

»

# CME - FILLABLE PDF FORM

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**8 List any changes to your clinical practice that you may make having read this article.**

## CYSTIC FIBROSIS IN ADULTS

**9 Which of the statements are true?**

**A** The prevalence of *Pseudomonas aeruginosa* infection increases with age
**True** ☐ **False** ☐

**B** Pancreatitis and sinusitis are both clinical features that may support a diagnosis of cystic fibrosis in adults
**True** ☐ **False** ☐

**C** Cystic fibrosis related diabetes is associated with increased morbidity at lower levels of dysglycaemia than type 1 and type 2 diabetes
**True** ☐ **False** ☐

**D** Symptoms of anxiety and depression are twice as high in cystic fibrosis patients compared with the general population
**True** ☐ **False** ☐

**E** Manual chest clearance is the mainstay of physiotherapy
**True** ☐ **False** ☐

**10 Match the statistic to the statement.**

**A** The proportion of CF patients with exocrine pancreatic insufficiency

**B** The percentage of cystic fibrosis deaths caused by progressive respiratory failure

**C** The proportion of men with cystic fibrosis who are infertile

**1** 80%
**2** 85%
**3** 90%
**4** 98%

**11 List the four principles of optimal care in patients with cystic fibrosis.**

**12 List any changes to your clinical practice that you may make having read this article.**

## Answers

**MILD TO MODERATE ASTHMA IN ADULTS**

**1** **A** False **B** False (a third) **C** True **D** True **E** True

**2** **A** 4 **B** 1 **C** 3

**3** No bothersome symptoms during the day or night; require little or no reliever medication; no exacerbations; preserved lung function

**CYSTIC FIBROSIS IN CHILDREN**

**5** **A** True **B** True **C** True **D** False (teenage years) **E** True

**6** Acute abdominal pain; constipation; hydrated; laxatives

**7** **A** The fat-soluble vitamins A, D, E and K; nutritional supplements **B** Meconium ileus; steatorrhoea; faltering growth; distended abdomen; constipation

**CYSTIC FIBROSIS IN ADULTS**

**9** **A** True **B** True **C** True **D** False (up to three times as high) **E** False (a combination of chest clearance techniques, exercise and inhaled mucolytic therapies)

**10** **A** 2 **B** 3 **C** 4

**11** Aggressive treatment of infection; airway clearance; optimisation of nutritional status; close monitoring of complications and comorbidities

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# Managing mild to moderate asthma in adults

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# Managing mild to moderate asthma in adults

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**How** should patients be assessed?

**What** are the treatment options?

**How** can outcomes be improved?

**THE UK HAS ONE OF THE HIGHEST PREVALENCE RATES FOR ASTHMA IN THE WORLD WITH AROUND** 1 in 12 (4.3 million) adults affected.[1] Depending on the exact definition used the vast majority of asthma is considered to be mild to moderate (i.e. symptoms and exacerbation frequency controlled without the need for high-dose asthma therapy) and is therefore managed predominantly in primary care.

However, it is widely acknowledged that asthma is an umbrella term covering a multitude of phenotypes, within which symptoms, airway inflammation and bronchial hyperreactivity vary over time. As such no single test performed at one specific time point can definitively confirm or refute the diagnosis.

Asthma guidelines may contain complex diagnostic pathways[2] (see figure 1, p14), suggesting multiple investigations, some of which remain uncommon in primary care (e.g. exhaled nitric oxide)[2] and even when followed may not result in a clear diagnosis being made for a significant proportion of individuals.[3] This coupled with the fact that a number of other conditions (including non-respiratory causes) may also present with wheeze, cough, chest tightness and episodic breathlessness means there are simultaneous concerns about both the over- and under-diagnosis of asthma. However, what is clear from the UK-based National Review of Asthma Deaths (NRAD) report *Why asthma still kills* is that most patients who died were actually considered to have mild to moderate asthma, with 57% of fatal cases not under specialist care at the time of their death.[4]

**'Most patients who died were considered to have mild to moderate asthma, with 57% of fatal cases not under specialist care at the time of death'**

## MANAGEMENT

### Pharmacological treatment

The *Why asthma still kills* report highlighted that in cases of fatal asthma there was widespread underuse of inhaled corticosteroids (ICS) — 80% of patients did not receive monthly treatment — and an overreliance on short-acting beta-agonists (SABA).[4] In the most extreme case, one patient »

**Box 1**

**The Royal College of Physicians three questions to identify individuals with good, recent symptomatic control of asthma**

In the past month:
- Have you had difficulty sleeping because of your asthma symptoms (including cough)?
- Have you had your usual asthma symptoms during the day (cough, wheeze, chest tightness, or breathlessness)?
- Has your asthma interfered with your usual activities (e.g. housework, work/school, etc)?

**NB** No to all three questions indicates good control

**'Significant over-reliance on reliever medication and underuse of inhaled corticosteroids is a key reason for poor outcomes in mild to moderate asthma'**

who died had received 113 reliever inhalers in the previous 12 months.[4]

It is easy to understand why those with mild to moderate disease might forgo regular maintenance therapy and instead rely solely on as required reliever treatment. However, this may mask deteriorating symptoms while allowing the accumulation of critical airway inflammation, risking a life-threatening deterioration which may be sudden. In the National Review of Asthma Deaths, 45% of those who suffered a fatal asthma attack died without seeking medical assistance or before emergency medical care could be provided.[4] The joint BTS/SIGN guideline therefore no longer considers SABA inhalers an appropriate monotherapy apart from in those few individuals with very occasional short-lived wheeze[5] (and there is debate about whether such cases should be classed as asthma at all).

Asthma is an inflammatory airways disease and should therefore always be treated with a steroid-containing inhaler. All other treatments (including SABA) should be considered additional to inhaled corticosteroids.

Confusingly, national guidelines diverge on the recommended initial add-on therapy to regular low-dose ICS. BTS/SIGN suggests adding in a long-acting beta-agonist (LABA) in the form of a combination inhaler,[5] which may help to avoid non adherence with steroids and is modestly superior in reducing oral corticosteroid-treated exacerbations[6] when compared with the NICE recommendation of an oral leukotriene receptor antagonist. However, the latter may be more cost effective in those patients in whom it works.[2]

Both guidelines suggest considering a maintenance and reliever therapy (MART) regimen before [NICE], or after [BTS/SIGN] progressively escalating the dose of the ICS component of treatment.[2,5]

A MART regimen allows a single ICS/LABA combination inhaler to be used both regularly as a preventer and in between as required as a quick acting reliever. Individuals using a MART regimen receive escalating inhaled steroid therapy automatically when their symptoms increase and this form of management has been shown to reduce unplanned admissions while actually decreasing total steroid burden overall.[7] Only certain ICS/LABA devices and strengths are licensed to be used in this way and in those that are the total number of daily doses may vary by product.

Care must be taken in those sub groups where there is poor correlation between symptoms and airway inflammation – individuals who regularly use their inhalers for episodes of dysfunctional breathing or anxiety may, on a MART regimen, be inadvertently exposing themselves to high-doses of ICS, something that increases the risk of pneumonia.[8]

Use of separate LABA inhalers, which are unlicensed for asthma, is considered unsafe and has been linked to an increased risk of asthma-related death.[9,10] The majority of mild to moderate asthma would be expected to respond prior to the need for high-dose ICS-containing products ± a fourth agent (e.g. long-acting muscarinic receptor antagonist or theophylline), the need for which heralds more difficult or severe asthma and onward referral should be considered.

Consideration must also be given to de-escalation of treatment where possible. It is important to discuss with individuals who have been well controlled for at least three months about reducing treatment. Withdraw oral ± inhaled adjuncts to ICS therapy where possible; aim to reduce any steroid-containing product by no more than 25-50% at a time until the lowest ICS-containing maintenance regimen achievable is found.[11]

**'Consideration must also be given to de-escalation of treatment where possible'**

NICE guidance still suggests that there is a group of patients that might be treated with SABA alone and while it is clear that this is intended to be suitable for only a very small minority of individuals[2] this has proved a controversial decision in light of the NRAD report,[4] particularly as a large-scale Scandinavian study has shown that early implementation of preventer therapy in primary care reduces mortality.[12] Data is beginning to emerge to suggest that reliever inhalers may not be required at all in some forms of mild asthma.[13]

Both major UK guidelines draw attention to the need to check inhaler technique and reinforce good concordance with treatment.[2,5]

**'Pregnant women should be informed that uncontrolled asthma is by far the greatest risk to their developing fetus'**

Importantly, treatment should be continued during pregnancy as large-scale studies have failed to show any increase in congenital malformations associated with asthma therapy. Pregnant asthma patients, a third of whom will experience worsening symptoms during pregnancy, should be informed that uncontrolled asthma is by far the greatest risk to their developing fetus.[5]

### Other medication

Consider whether coprescribed medications may be impacting on asthma

control particularly beta-blockers (even cardioselective ones) and NSAIDs. Cough induced by ACE inhibitors may be mistaken for worsening asthma symptoms. Sensitivity to aspirin is also common, particularly in patients with comorbid nasal polyps, with some series suggesting this impacts on > 20% of cases.[14] If ongoing antiplatelet therapy is still indicated, clopidogrel may be a suitable alternative, particularly as its anti-leukotriene effects have been shown to attenuate airway hyperresponsiveness in animal models.[15]

### Non-pharmacological treatment

While triggers should be identified, forms of aeroallergen avoidance, particularly those aimed at decreasing exposure to house dust mite, have not been shown to be an effective method of improving asthma control and should not be routinely recommended.[5]

Significant weight loss in obese asthma patients may be beneficial,[16] as with any respiratory condition patients often become fearful of getting breathless and restrict their physical activity leading to weight gain and deconditioning which further exacerbate their symptoms.

Up to one in five asthma patients smoke. Smoking is associated with poorer control and higher use of oral corticosteroids[17] and was another preventable factor highlighted in the NRAD report.[4]

### Assessment

According to the BTS/SIGN guideline individuals with well controlled asthma:

- Should not have bothersome symptoms in the day or night
- Require little/no reliever medication
- Do not suffer exacerbations
- Have preserved lung function

Control should be achieved without significant side effects from treatment.[5] Assessment should consider both current symptomatic impact as well as future risk of asthma. Over time patients may become accustomed to ongoing symptoms and fail to complain about less than adequate control. The RCP three questions are a quick, simple and clinically useful screening tool to identify individuals with good recent symptomatic control[18] (see box 1, p12).

Exacerbation frequency, lung function (spirometry or peak expiratory flow rates) and possibly exhaled nitric oxide readings[19] may help to identify individuals at higher future risk and these factors alone may prompt consideration of a need to increase treatment or seek specialist advice in otherwise seemingly well controlled individuals. Men with late-onset asthma in particular may have low symptom perception but high levels of airway inflammation.[20] We feel that exhaled nitric oxide monitoring may be particularly useful to guide the management of this group although there is not yet an established evidence base to support this.

## SELF-MANAGEMENT

Helping patients to understand the need for ongoing use of ICS-containing maintenance therapy, even after symptoms abate is of the utmost importance. Patients are often shocked when told that their blue inhaler does not treat asthma.

**‘Patients are often shocked when told that their blue inhaler does not treat asthma’**

Non concordance with treatment is widespread. Even among patients attending tertiary level asthma services more than a third were found to collect less than half of their medications.[21]

Education on self-management delivered in a primary care setting can be effective in improving asthma control and preventing unplanned use of healthcare services particularly when supported by a written personalised asthma action plan (PAAP).[5] Such plans are fundamental to improving self-management. They should include details of a patient’s maintenance regimen and also what specific actions to take in response to deteriorating asthma control, such as temporarily increasing the ICS component of treatment or using oral steroids. PAAPs may be triggered by either increasing symptoms or decreasing peak flow readings compared with an individual’s predicted or, if known, actual best.

An example of an action plan using a traffic light system is available to download free of charge from the Asthma UK website, (see figure 2, p15). Once individualised, patients should be encouraged to take a picture of their PAAP on their mobile phone so it is readily available to them.

As well as understanding what they should be taking for their asthma and when, patients must also be shown how to use their treatments and their inhaler technique reassessed at every contact with a healthcare professional. Matching devices or using combination inhalers as far as possible when multiple inhaled therapies are required may help avoid unintentional non-adherence, as studies have shown that patients with respiratory conditions who cannot use their inhalers properly experience poorer disease control and more severe exacerbations.[22]

## REFERRAL

A wide range of national and international recommendations exist on which patients should be referred to secondary care,[23] in general we feel the main determinants include the factors shown in table 1, below.

Non-referral of individuals requiring secondary care input has been highlighted as an avoidable factor in cases of fatal asthma.[4] Data, soon to be published, from a retrospective national database study suggest that although »

**Table 1**

**Criteria for referral to secondary care**

**Diagnostic uncertainty or atypical features**

**Clinical findings:** Crepitations, inspiratory or monomorphic wheeze, clubbing, persistent breathlessness, copious sputum production. Rapidly progressive course; significant extra-pulmonary/systemic symptoms

**Investigations:** Chest X-ray changes, restrictive spirometry, marked or persistent eosinophilia

**Coexisting conditions complicating asthma control:** Overlapping COPD; vocal cord dysfunction; bronchiectasis; nasal polyposis. Pregnancy if associated with deteriorating control or significant problems during previous gestational periods

**Possibility of hypersensitivity to inhaled material**

**Including cases of suspected:** Hypersensitivity pneumonitis; occupational asthma; work-aggravated asthma; reactive airways dysfunction syndrome (that follows a single high-volume exposure to an irritant)

**Treatment and exacerbations**

**Need for high-dose ICS to control symptoms:** Refer patients at any stage of treatment in the following circumstances:

- More than two courses of systemic corticosteroids/year required
- Presentation to emergency care providers more than once in the past 12 months
- Following any asthma-related hospital admission

**Patients who have suffered even a single life-threatening asthma attack at any point in the past should be under specialist supervision**

**Figure 1**

**NICE recommendations for interpreting test results for adults with symptoms suggestive of asthma**[2]

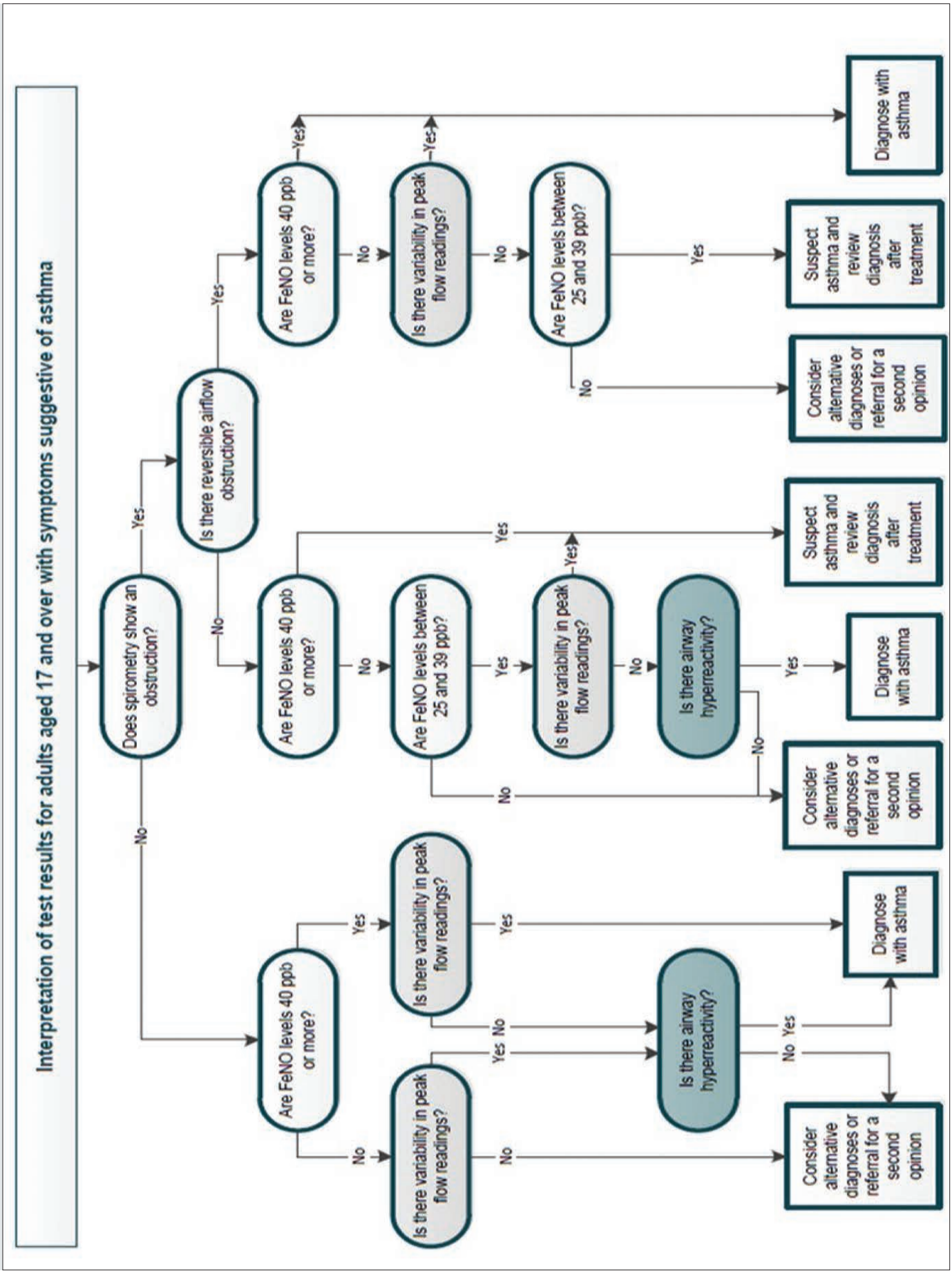


**Figure 2**

**Asthma UK adult asthma action plan using the traffic light system. The template can be downloaded free from the Asthma UK website: www.asthma.org.uk**

## Every day asthma care:

**My asthma is being managed well:**

- With this daily routine I should expect/aim to have no symptoms.
- If I've not had any symptoms or needed my reliever inhaler for at least 12 weeks, I can ask my GP or asthma nurse to review my medicines in case they can reduce the dose.
- My personal best peak flow is: ____

### My daily asthma routine:

**My preventer inhaler** (insert name/colour):

**I need to take my preventer inhaler every day even when I feel well**

I take ____ puff(s) in the morning

and ____ puff(s) at night.

**My reliever inhaler** (insert name/colour):

I take my **reliever** inhaler only if I need to

I take ____ puff(s) of my reliever inhaler if any of these things happen:

- ★ I'm wheezing
- ★ My chest feels tight
- ★ I'm finding it hard to breathe
- ★ I'm coughing

**Other medicines and devices (eg spacers) I use for my asthma every day:**

## When I feel worse:

**My asthma is getting worse if I'm experiencing any of these:**

- My symptoms are coming back (wheeze, tightness in my chest, feeling breathless, cough).
- I am waking up at night.
- My symptoms are interfering with my usual day-to-day activities (eg at work, exercising).
- I am using my reliever inhaler three times a week or more.
- My peak flow drops to below: ____

**URGENT! If you need your reliever inhaler more than every four hours, you're having an asthma attack and you need to take emergency action now.**

**What I can do to get on top of my asthma now:**

If I haven't been using my preventer inhaler, I'll start using it regularly again or if I have been using it...

Increase my preventer inhaler dose to ____ puffs ____ times a day until my symptoms have gone and my peak flow is back to my personal best.

Take my reliever inhaler as needed (up to ____ puffs every four hours).

I carry my reliever inhaler with me when I'm out.

**URGENT! See a doctor or nurse within 24 hours if you get worse at any time or you haven't improved after seven days.**

**Other advice from my GP about what to do if my asthma is worse (eg SMART/MART or rescue steroid tablets):**

## In an asthma attack:

**I'm having an asthma attack if I'm experiencing any of these:**

- My reliever inhaler is not helping or I need it more than every four hours.
- I find it difficult to walk or talk.
- I find it difficult to breathe.
- I'm wheezing a lot or I have a very tight chest or I'm coughing a lot.
- My peak flow is below: ____

### What to do in an asthma attack

1. **Sit up straight** – try to keep calm.
2. **Take one puff of your reliever inhaler (usually blue)** every 30 - 60 seconds, up to a maximum of 10 puffs.
3. **If you feel worse** at any point OR you don't feel better after 10 puffs call 999 for an ambulance.
4. **Repeat step 2 after 15 minutes** while you're waiting for an ambulance.

**After an asthma attack:**

See your GP within 48 hours to make sure you're not at risk of another attack. If you get worse see them urgently. Finish any medicines they prescribe you, even if you start to feel better. If you don't improve after treatment, see your GP urgently.

**What to do in an asthma attack if I'm on SMART/MART:**



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## key points

SELECTED BY

**Dr Peter Saul**
GP with an interest in respiratory medicine, Wrexham and Associate GP Dean for North Wales, UK

**The UK has one of the highest prevalence rates for asthma** in the world with around 1 in 12 (4.3 million) adults affected. Depending on the exact definition used the vast majority of asthma is considered to be mild to moderate (i.e. symptoms and exacerbation frequency controlled without the need for high-dose asthma therapy) and managed predominantly in primary care.

**The UK-based National Review of Asthma Deaths** found that most patients who died were actually considered to have mild to moderate asthma, with 57% of fatal cases not under specialist care at the time of death. In cases of fatal asthma there was widespread underuse of inhaled corticosteroids (ICS) — 80% of patients did not receive monthly treatment — and an overreliance on short-acting beta-agonists (SABA).

**The joint BTS/SIGN guideline no longer considers** SABA inhalers an appropriate monotherapy apart from in those few individuals with very occasional short-lived wheeze. Asthma is an inflammatory airways disease and should always be treated with a steroid-containing inhaler. All other treatments including SABA should be considered additional to ICS.

**Guidelines diverge on the recommended initial** add-on therapy to regular low-dose ICS; BTS/SIGN suggests adding in a long-acting beta-agonist (LABA) in the form of a combination inhaler, which may help to avoid non adherence with steroids and is modestly superior in reducing oral corticosteroid-treated exacerbations when compared with the NICE recommendation of an oral leukotriene receptor antagonist. Both guidelines draw attention to the need to check inhaler technique and reinforce good concordance with treatment.

**According to the BTS/SIGN guideline individuals with** well controlled asthma: should not have bothersome symptoms in the day or night; should require little or no reliever medication; should not suffer exacerbations; and have preserved lung function. Education on self-management, in primary care, can be effective in improving asthma control and prevent unplanned use of healthcare services particularly when supported by a written personalised asthma action plan. As well as understanding what they should be taking for their asthma and when, patients must also be shown how to use their treatments and their inhaler technique reassessed at every contact with a healthcare professional. Reducing exposure to house dust mite, has not been shown to be an effective method of improving asthma control. Significant weight loss in obese asthma patients may be beneficial. Smoking is associated with poorer control and higher use of ICS.

**Referral to secondary care should take place when** there is diagnostic uncertainty or atypical features, the possibility of hypersensitivity to inhaled material or concerns over treatment or exacerbations. Patients who have suffered even a single life-threatening asthma attack should be under specialist supervision.

**Box 2**

**Good practice points in mild to moderate asthma**

- Interrogate asthma registry datasets to identify individuals not receiving ICS-containing treatment – such patients need their diagnosis or treatment reviewing urgently
- Ensure all patients have been taught to use their inhaler correctly and reassess this whenever possible – where feasible match device types, use combination inhalers and avoid blanket switching to other products
- Ensure all asthma patients have an individualised personal asthma action plan and audit this regularly
- Assess prescription pick-up rates in any patient with deteriorating control, prior to any escalation in regular treatment or referral to secondary care
- Consider how frequently reliever medication should be issued – short-acting beta-agonists do not modify disease course and overreliance on them is associated with fatal asthma

a significant proportion of asthma patients fulfill referral criteria, only a small percentage of eligible patients are actually sent for a specialist opinion and even then after some considerable delay. Whether there is adequate capacity to see all eligible patients in secondary care remains unclear.

## IMPROVING OUTCOMES

We have highlighted relatively simple methods that may lead to significantly improved asthma outcomes, particularly the need to ensure regular ICS-containing products are taken as well as the use of written action plans to support self-management. Furthermore, in the *Why asthma still kills* report the following additional recommendations were made:[4]

- GP practices should have a named clinical lead for asthma
- Structured asthma reviews should take place at least annually

The report also suggested that patients prescribed > 12 short-acting reliever inhalers in a year should be called for urgent review.[4] However, a well controlled asthma patient should have infrequent need of SABA – a blue inhaler should last almost a year if used no more than twice a week. We are aware of some primary care teams therefore issuing reliever medication quarterly rather than monthly, making a clinical review necessary for those patients requiring more than four blue inhalers a year and helping to reinforce the importance of maintenance therapy. Likewise patients on a MART regimen who regularly order extra inhalers should be called for a review. Other services routinely check pick-up rates for ICS before escalating treatment or referring to secondary care.

## CONCLUSION

Mild to moderate asthma is very common and generally its impact and future risk can be managed extremely well with simple interventions. However, this patient group may be prone to an acceptance of suboptimal control and underestimate the potential for sudden deterioration of their condition if management is not adhered to. As with other chronic conditions huge potential gains are achievable with simple interventions that could be achieved using expertise and systems already in place in primary care (see box 2, above).

**Competing interests**
Dr Gareth Jones has received honoraria from Astra-Zeneca, GSK, Pfizer and NAPP.
Dr Fatima Khalid has no competing interests.

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## Useful information

**BTS/SIGN asthma guideline (2016)**
**www.brit-thoracic.org.uk/standards-of-care/guidelines/btssign-british-guideline-on-the-management-of-asthma**

**NICE asthma guideline. NG80. (2017)**
**www.nice.org.uk/guidance/ng80**

**Comparing BTS/SIGN and NICE recommendations**
A comparison of the differences between the BTS/SIGN and NICE asthma guidelines
**blogs.bmj.com/thorax/files/2017/12/BTS-SIGN-and-NICE-Asthma-guidelines.pdf**

**Asthma UK**
A downloadable asthma action plan template can be found at:
**www.asthma.org.uk/globalassets/health-advice/resources/adults/adult-asthma-action-plan.pdf**

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# Diagnosing and managing cystic fibrosis in children

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# Diagnosing and managing cystic fibrosis in children

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**FIGURE 1**
A young girl with cystic fibrosis performing chest physiotherapy with a positive expiratory pressure mask

**How** is cystic fibrosis diagnosed in children?

**What** are the mechanisms involved?

**What** are the treatment strategies?

**CYSTIC FIBROSIS (CF) IS AN AUTOSOMAL RECESSIVE GENETIC DISORDER AFFECTING ABOUT 1 IN** 2,500 live births in the UK.[1]

Although all newborns undergo screening for CF, around 15% of infants will present shortly after birth with meconium ileus[2] and some will already have faltering growth when the screening results are available at 3-4 weeks of age. Infants who present with meconium ileus should be treated with a high index of suspicion for CF until proven otherwise. Around 93% of children are diagnosed in the first three months of life and another 5% before the age of 12 months.[2]

CF is a multisystem disease that affects the respiratory and gastrointestinal systems in particular. Respiratory symptoms may be non specific initially including cough and wheeziness, frequent respiratory infections and, in older children, sinusitis. In CF, these symptoms are a consequence of abnormal CF transmembrane conductance regulator (CFTR) function. The CFTR protein regulates ion and water content of epithelial secretions.

Symptoms of malabsorption include meconium ileus, steatorrhoea, faltering growth, distended abdomen and constipation.[3]

Since 2007, CF has been part of the UK newborn blood spot screening programme, with a dried blood spot specimen collected from all infants at 5-7 days of age.[4]

The CF screening test measures immunoreactive trypsin (IRT) levels. The sensitivity of the current IRT protocol is around 97%. If IRT is consistently raised, the same blood sample is tested for the four most common CF gene mutations. If two mutations are found, the baby is identified as 'CF suspected'. If there is only one mutation, an extended panel of up to 31 genes is tested. If a second mutation is not found, the IRT is repeated, with a higher threshold. A result above the threshold is designated 'CF suspected' and a result below the threshold is categorised as 'probable carrier'. The parents of suspected carriers should receive a home visit, from a health professional, within one working day. An information sheet for parents of suspected carriers is available online (see Useful information, p21).

If no mutation is found, a second blood sample may be collected and the IRT repeated. If the IRT remains high, »

the child is identified as 'CF suspected'. If not, the designation is 'CF not suspected'.

Where CF is suspected, the screening centre will notify the CF centre which will arrange immediate care and a confirmatory sweat test.

## GENE MUTATIONS

CF is an autosomal recessive disorder caused by mutations affecting chromosome 7, which encodes the CFTR protein.[5] The most common mutation is the codon deletion encoding phenylalanine at position 508 of the CFTR protein, p.Phe508del (F508del). There are currently 312 known disease causing mutations of the CF gene.[6]

Loss of CFTR function in the lungs, impairs the mucociliary escalator leading to accumulation of mucus in the airways and secondary infection. In the gut, pancreatic function is impaired in most children, leading to malabsorption and faltering growth.

The different mutations affect CFTR function in different ways and there are currently six classes of CFTR mutations.[5]

Many new approaches to treating the basic defect are the subject of current or recently completed clinical trials, including: gene replacement (gene therapy) and RNA repair, which are not dependent on mutation class, and read through agents (circumventing the stop codon in class I mutations). Ivacaftor is a CFTR modulating drug. It is approved by specialist commissioning for patients with CF, aged two years and over, who have at least one copy of the G551D mutation or of one of eight other less common mutations.

## CONFIRMING DIAGNOSIS

The gold standard as recommended by the European Cystic Fibrosis Society is a sweat test and genetic testing. The three pillars of the diagnosis are:

- Clinical features (or family history or positive newborn screening test) and
- Sweat chloride > 59 mmol/L and/or
- Two CF-causing CFTR mutations[7]

## COMMON COMPLICATIONS

Advances in CF treatment have helped improve the survival of patients with CF. However, most children will have malabsorption, requiring pancreatic enzyme replacement therapy (PERT) and a progressive deterioration of respiratory function.

### Respiratory

Most infants diagnosed by newborn screening have few symptoms. However, later on children will experience recurrent respiratory infections.

Symptoms may be as subtle as a wet cough but must be treated to avoid progression to bronchiectasis and ultimately respiratory failure.[8] Antibiotic choice should be guided by previous microbiology results and national guidelines.[9] Some of these infections may become chronic unless treated promptly – a particular problem with *Pseudomonas aeruginosa*. Further investigation by the CF centre should take place if a wet cough persists in spite of a two-week course of oral antibiotics or if sputum cultures are repeatedly negative.

**'Symptoms may be as subtle as a wet cough but must be treated to avoid progression to bronchiectasis and ultimately respiratory failure'**

Allergic bronchopulmonary aspergillosis, another respiratory complication seen in CF, results from a hypersensitivity reaction to *Aspergillus fumigatus* mould which has colonised in the bronchi. The release of IgE and IgG antibodies results in bronchial obstruction through mucus plugging and causes increased cough and haemoptysis.[10] Suspected cases should be discussed with the CF team.

### Gastrointestinal

Most children with CF have malabsorption requiring PERT, fat-soluble vitamins and, in some cases, nutritional supplements. Distal intestinal obstruction syndrome is a serious complication that presents with acute abdominal pain and constipation. Viscous fluid in the intestine causes obstruction in the terminal ileum and is a surgical emergency.

By ensuring children are adequately hydrated and have the appropriate laxatives, gut transit time is improved and the risk of distal intestinal obstruction syndrome reduced.[11]

Liver disease results from damage caused by fatty replacement and interstitial fibrosis resulting from inspissated secretions.[12] CF related diabetes (CFRD) becomes prevalent in the teenage years, often presenting with poor weight gain in a young person who had previously been thriving, and one third of adults with CF have CFRD.[2] Treatment is usually with a long-acting insulin formulation and should be commenced by the CF centre.[7]

Other complications include:[13]

- Nasal polyposis and sinusitis
- Reduced bone mineral density
- CF arthropathy
- Renal and hearing complications secondary to aminoglycoside use
- Delayed puberty
- Psychosocial difficulties
- Male infertility
- Reduced female fertility

## MANAGEMENT

It is important to manage symptoms of CF and slow down the progression of lung disease, to delay the complications described above.

Most newly diagnosed infants with CF will commence once daily physiotherapy for airway clearance delivered by their parents. As children get older they will increasingly be in charge of their airway clearance. Many use positive expiratory pressure devices that require sustained expiration against resistance, followed by a series of 'huffs' and coughs (see figure 1, p19).[7]

Preventing or attenuating respiratory infections in patients with CF helps slow the progression of lung disease. Young children are vulnerable to respiratory infection with *Staphylococcus aureus* and, in the UK, most receive a prophylactic antibiotic, commonly flucloxacillin, until the age of three years.[9,14]

**'Preventing or attenuating respiratory infections in cystic fibrosis patients helps slow the progression of lung disease'**

Children may present to primary care with exacerbations of respiratory symptoms (wet cough, sometimes with sputum). Initial management in primary care, prior to consulting with or referring to the CF centre, can help achieve more rapid resolution of symptoms. A sputum sample should be taken. Alternatively, in children who do not produce sputum, hold a sterile swab at the back of the throat and ask the child to cough (cough swab). For children who live

## key points

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**Cystic fibrosis (CF) is a multisystem genetic disorder** affecting approximately 1 in 2,500 live births in the UK. Although all newborns undergo screening for CF, around 15% of infants will present shortly after birth with meconium ileus and some will already have faltering growth when the screening results are available at 3-4 weeks of age. Infants who present with a meconium ileus should be treated with a high index of suspicion for CF until proven otherwise. Around 93% of children are diagnosed in the first three months of life and another 5% before the age of 12 months.

**Mucociliary dysfunction leads to accumulation of** mucus in the airways and secondary infection. Respiratory symptoms may be non-specific initially and include cough and wheeziness, frequent respiratory infections and, in older children, sinusitis. Pancreatic function is impaired in most children, and symptoms of malabsorption include meconium ileus, steatorrhoea, faltering growth, distended abdomen and constipation.

**Distal intestinal obstruction syndrome is a serious** complication that presents with acute abdominal pain and constipation. Liver disease may result from fatty replacement or interstitial fibrosis due to inspissated bile. CF related diabetes becomes prevalent in the teenage years, often presenting with poor weight gain in a young person who had previously been thriving.

**Most newly diagnosed infants with CF will commence** once daily physiotherapy for airway clearance. At first this is delivered by their parents but as children get older they will increasingly be in charge of their airway clearance. Young children are vulnerable to respiratory infection with *Staphylococcus aureus* and, in the UK, most receive a prophylactic antibiotic, commonly flucloxacillin, until the age of three years. Exacerbations (worsening cough which is wet or associated with sputum) must be treated proactively with antibiotics and increased airway clearance. Children with CF should have antibiotics when a wet cough has been present for more than a week even if there is little to hear on auscultation of the chest. The CF centre should be contacted for advice on the choice of oral antibiotics and whether intravenous antibiotics might be needed. Pancreatic enzyme replacement therapy has been shown to improve fat absorption with concomitant improvement in calorie intake. Supplements of fat-soluble vitamins (A, D, E and K) may also be necessary as these vitamins are poorly absorbed.

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some distance from a CF centre, antibiotics can be prescribed in primary care, following national guidelines[9] or advice from the CF centre. Longer courses, commonly two weeks, of oral antibiotics are required and children should have effective chest physiotherapy. The CF centre should arrange early review to assess the response to treatment.

PERT has been shown to improve fat absorption with concomitant improvement in calorie intake. Children will generally have a daily dose of 10-20 capsules of a PERT formulation (containing 10,000 units of lipase) and so prescriptions should be for larger quantities of PERT to avoid parents running out of this essential medication. Supplements of fat-soluble vitamins (A, D, E and K) may also be necessary as these vitamins are poorly absorbed. [4,15]

## MONITORING

Children should be seen regularly by a specialist centre every few months and have a formal detailed annual review. Each clinic review should involve the multidisciplinary team: CF specialist physician, CF nurse, physiotherapist, dietitian, clinical psychologist, social worker and pharmacist.[3] The annual review should assess all systems and identify any areas where symptom and disease control is not optimal and needs to be addressed.

Respiratory function should be monitored with lung function tests and chest imaging. It is also important to monitor sputum cultures to allow early diagnosis of infections with organisms such as *P. aeruginosa* which will require eradication treatment. *P. aeruginosa* eradication requires combined treatment, usually with oral ciprofloxacin and nebulised colistin for periods of between three weeks and three months.[9,16]

**‘Parents of a baby newly diagnosed with cystic fibrosis should be offered genetic counselling’**

Children should see their GP for regular prescriptions and immunisations, including annual influenza vaccination.

Parents of a baby newly diagnosed with CF should be offered genetic counselling to help with decisions regarding future pregnancies. Testing for older siblings may also be carried out.

**Competing interests**
Professor Alan Smyth has provided consultancy for Vertex, PTC, Roche and Gilead. He has taken part in clinical trials sponsored by Vertex, Raptor and Insmed. He has also given lectures at meetings sponsored by Gilead, Teva and Vertex. Dr Christabella Ng has no competing interests.

*See also Managing complications of cystic fibrosis in adults, p23.*

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## Useful information

**Cystic Fibrosis Trust**
www.cysticfibrosis.org.uk

**Information for parents of suspected carriers**
https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/701859/NBS_screening_cystic_fibrosis_carrier_final_web_240418.pdf

The Practitioner

# Managing complications of cystic fibrosis in adults

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# Managing complications of cystic fibrosis in adults

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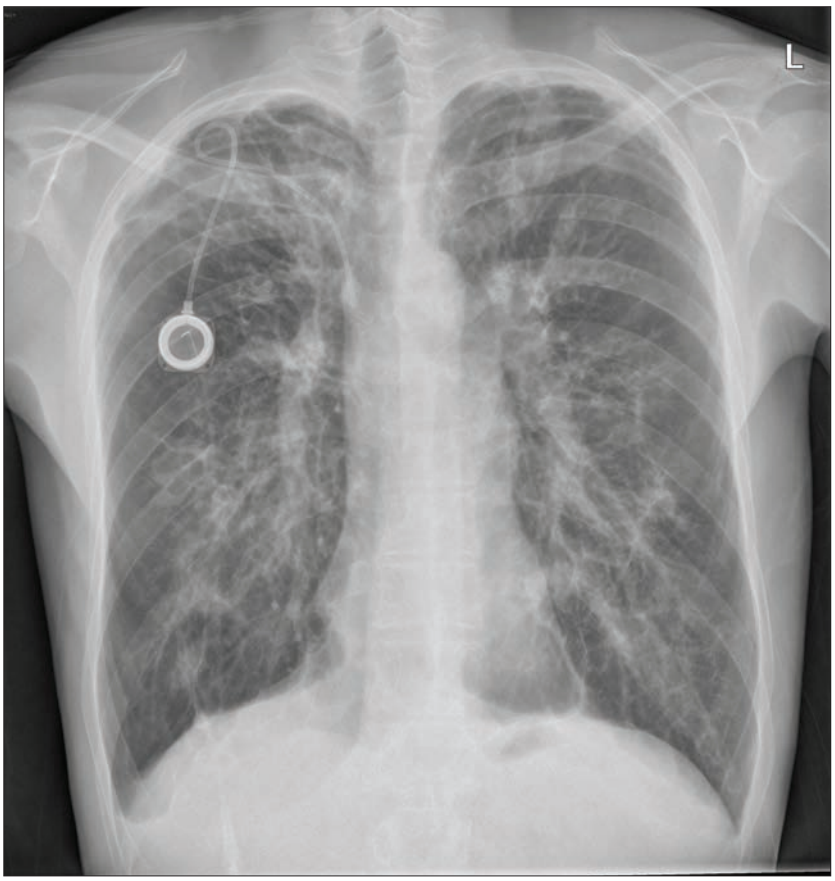



**FIGURE 1**
Plain chest radiograph of an adult with cystic fibrosis. A totally implantable venous access device overlies the right upper zone. There are areas of opacification and changes in keeping with bronchiectasis distributed predominantly in the upper and mid zones

**What** are the common comorbidities?

**How** should patients be monitored?

**What** are the management approaches?

**CYSTIC FIBROSIS (CF) IS THE MOST COMMON LIFE-LIMITING GENETIC CONDITION IN THE WESTERN** world, with more than 10,000 sufferers in the UK alone.[1]

It is caused by mutations in the gene encoding for the cystic fibrosis transmembrane conductance regulator (CFTR) protein that regulates ion and water content of epithelial secretions. Defective CFTR results in thick viscid secretions in many different body systems. CF is a truly multiorgan disease that manifests itself most obviously in the lungs. Impaired mucociliary clearance leads to chronic infection, inflammation and subsequent bronchiectasis (see figure 1, above) with progressive loss of lung function and ultimately respiratory failure.

CF is inherited as a classic Mendelian autosomal recessive trait and the UK has a relatively high prevalence where 1 in 25 Caucasians are CFTR mutation carriers with a disease incidence of 1 in 2,500 live births; ethnic minorities have a lower carrier incidence with correspondingly lower disease rates.

**'Median life expectancy for patients with cystic fibrosis now extends into the fifth decade'**

Overall survival has improved dramatically since the millennium such that median life expectancy for patients with CF now extends into the fifth decade.[1] However, with increasing longevity comes a range of comorbidities, some as complications of the disease itself but others as a consequence of long-term treatments required to manage CF. In 2017 NICE published guidance on the diagnosis and management of CF in both children and adults.[2]

## DIAGNOSIS

Since 2007 a national newborn blood spot screening programme (NBSP) has been in place throughout the UK and most patients are now diagnosed shortly after birth via the heel prick test. However, the NBSP does not detect less common CF-related mutations such that some children with CF still go undiagnosed and along with those born before 2007 may reach adulthood before diagnosis is contemplated.

Furthermore, it is increasingly recognised that the NBSP identifies some infants with no clinical features of CF, but who may be at risk of developing the CF syndrome later in life and have been given the designation of cystic »

**Table 1**

**Clinical features that may support a diagnosis of cystic fibrosis**

**Children**
- Meconium ileus
- Faltering growth
- Malnutrition
- Wet cough
- Symptoms of malabsorption
- Rectal prolapse
- Distal intestinal obstruction syndrome

**Adults**
- Recurrent and chronic pulmonary disease
  — Recurrent infection
  — Bronchiectasis (particularly upper zone distribution)
  — Chronic productive cough
- Pancreatitis
- Sinusitis
- Infertility (male)

**Table 2**

**Complications and comorbidities associated with cystic fibrosis**

**Upper respiratory tract:** Nasal polyps; sinusitis

**Lower respiratory tract:** Chronic infection; bronchiectasis

**Pancreas:** Cystic fibrosis related diabetes; pancreatitis

**Hepatobiliary:** Cystic fibrosis related liver disease; gallstones

**Renal:** Kidney stones; chronic kidney disease

**Gastrointestinal:** Malabsorption; Gastro-oesophageal reflux disease; distal intestinal obstruction syndrome

**Others:** Arthropathy; congenital absence of the vas deferens; subfertility in women

**Table 3**

**Principles of optimal care in cystic fibrosis patients**

- Aggressive treatment of infection
- Airway clearance
- Optimisation of nutritional status
- Close monitoring of complications and comorbidities

fibrosis screen positive inconclusive diagnosis. It is therefore important to maintain vigilance for those whose features suggest CF, see table 1, above. In such cases a referral to a local respiratory service is warranted so that further testing and/or referral on to the specialist CF centre can be facilitated.

The diagnosis of CF can be made based on clinical presentation alone; however, a diagnosis is best supported by genetic screening or an elevated sweat chloride level, see p20.

## LUNG DISEASE

Reduced clearance of thick mucus results in stasis of respiratory secretions, creating a microenvironment ripe for infection and inflammation.

**'Chronic infection with *P. aeruginosa* is associated with increased morbidity and mortality'**

Chronic infection and inflammation eventually cause irreversible bronchiectasis and decline in pulmonary function, and 90% of CF deaths are caused by progressive respiratory failure. Infection, commonly with *Haemophilus influenzae* and *Staphylococcus aureus*, may be detected early in life.

The prevalence of *Pseudomonas aeruginosa* increases with age; by adulthood more than 50% of CF patients are infected.[1] Unfortunately, chronic *P. aeruginosa* infection is associated with increased morbidity and mortality and in order to prevent cross-infection between patients, clinics are segregated based upon microbiology.

## EXTRAPULMONARY COMPLICATIONS

Defective CFTR is expressed in many organs, resulting in multisystem disease contributing to excessive morbidity, see table 2, above.

One of the most common extrapulmonary manifestations is in the reproductive system. Around 98% of males are infertile because of congenital absence of the vas deferens. In women with CF, mucus plugs may act as mechanical barriers to conception but the reproductive tract is otherwise structurally normal.[3]

The pancreas is another commonly affected organ and the term cystic fibrosis was coined from its appearance at autopsy.[4] Viscid pancreatic secretions become static with local autolysis, inflammation and fibrosis with subsequent exocrine and endocrine insufficiency.

Exocrine abnormalities are characterised by malabsorption of fat and fat-soluble vitamins, whereas dysregulated beta-cells struggle to release insulin adequately and cystic fibrosis related diabetes (CFRD) ensues.[5] CFRD is not only associated with aberrant glycaemia but also accelerated pulmonary function decline and a three-fold increase in mortality.[6]

**'Cystic fibrosis related diabetes is associated with a more rapid decline in pulmonary function and a three-fold increase in mortality'**

Elsewhere, stasis of hyperviscous bile can accumulate in the biliary tree causing focal inflammation and fibrosis. In this setting, persistent hepatocyte injury can lead to CF related liver disease, the third most common cause of death in children and adolescents with CF.[7,8] Other common comorbidities include polyposis and chronic sinusitis in the upper respiratory tract,[9] low bone mineral density,[10] gastro-oesophageal reflux[11] and renal disease.[12]

## MANAGEMENT

The management of individuals with CF has evolved dramatically over the past few decades and increasing survival can be attributed to improvements in nutrition, the aggressive treatment of infection, and the recognition of comorbidities, see table 3, above.

Specialist services have also evolved over time and patients should be managed in regional CF centres.

The CF centre model of care revolves around a specialist multidisciplinary team (MDT) with close links to other medical and surgical specialties. The core CF MDT consists of a respiratory physician, specialist nurse, specialist physiotherapist, clinical psychologist, dietitian and pharmacist. CF centres should be organisationally and

## key points

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**Overall survival has improved dramatically for patients** with cystic fibrosis (CF) since the millennium such that median life expectancy now extends into the fifth decade. However, with increasing longevity comes a range of comorbidities, some as complications of the disease itself but others as a consequence of long-term treatments required to manage CF. Chronic infection and inflammation eventually cause irreversible bronchiectasis and decline in pulmonary function, and 90% of CF deaths are caused by progressive respiratory failure. Chronic *Pseudomonas aeruginosa* infection is associated with increased morbidity and mortality, and by adulthood more than 50% of CF patients are infected.

**Around 98% of males are infertile because of** congenital absence of the vas deferens. In women with CF, mucus plugs may act as mechanical barriers to conception but the reproductive tract is otherwise structurally normal. CF related diabetes is associated with accelerated decline in pulmonary function and a three-fold increase in mortality. CF related liver disease is the third most common cause of death in children and adolescents with CF. Other common comorbidities include polyposis and chronic sinusitis in the upper respiratory tract, low bone mineral density, gastro-oesophageal reflux and renal disease.

**Regular surveillance for lung infection is warranted,** ideally with regular sputum sampling. In those with a first or new growth of *P. aeruginosa*, eradication strategies should include nebulised antibiotics in addition to a short course of oral or intravenous antibiotics. Despite eradication being initially successful, infection often returns and treatments targeted toward chronic *P. aeruginosa* infection include long-term nebulised antibiotics such as colistimethate, tobramycin and aztreonam. Exacerbations are characterised by acute deterioration in symptoms and lung function, and although milder exacerbations can be successfully treated with oral antibiotics, many require admission to hospital for intravenous antibiotics, glucocorticoids, optimisation of nutritional status and intensive airway clearance. Chronic macrolide therapy e.g. azithromycin has been shown to reduce pulmonary inflammation and exacerbation rate effectively in both adults and children.

**GPs are responsible for prescribing long-term** medications as advised by the specialist team, providing routine annual immunisation for patients with CF and their families, managing health problems not related to CF, supporting family members or carers, providing illness certification and working in partnership with the CF specialist team at the end of life.

**We welcome your feedback**

If you wish to comment on this article or have a question for the authors, write to: **editor@thepractitioner.co.uk**

structurally designed to minimise the risk of cross-infection between patients.

### Treatment of infection

Regular surveillance for lung infection is warranted from an early age, ideally with regular sputum sampling. Antibiotic strategies are targeted towards chronic suppression of the classical CF pathogens *S. aureus* and *P. aeruginosa*. In younger age groups *S. aureus* is more prevalent and the recent NICE guideline suggests chronic prophylaxis with flucloxacillin may be appropriate.[2] In those with a first or new growth of *P. aeruginosa*, initiation of eradication treatment has been shown to be successful.

Eradication strategies should include nebulised antibiotics in addition to a short course of oral or intravenous antibiotics. Despite eradication being initially successful, infection often returns and treatments targeted toward chronic *P. aeruginosa* infection include long-term nebulised antibiotics such as colistimethate, tobramycin and aztreonam. In the past decade, dry powder formulations of some nebulised antibiotics have become available which are quicker and easier to take but can be associated with increased cough.[13-15]

Chronic pulmonary infection is punctuated by exacerbations characterised by acute deterioration in symptoms and lung function. Although milder exacerbations can be successfully treated with oral antibiotics, many require admission to hospital for intravenous antibiotics, glucocorticoids, optimisation of nutritional status and input from chest physiotherapists for intensive airway clearance.

Exacerbations carry an associated risk of mortality and hence chronic inhaled antibiotics are targeted towards the suppression of infection to prevent exacerbations. In addition, chronic macrolide therapy, e.g. azithromycin has been shown to reduce pulmonary inflammation and exacerbation rates effectively in both adults and children.

### Airway clearance

Historically, physiotherapy for CF was focused solely on manual chest clearance techniques to help mobilise respiratory secretions. However, physiotherapy now includes a combination of chest clearance techniques, exercise and inhaled mucolytic therapies. Physiotherapists work with patients and families to tailor an individualised regimen that is effective and as efficient as possible.

Mucolytic therapies include recombinant human DNA cleaving enzymes such as dornase alpha (nebulised to break down viscid human and bacterial DNA material in CF sputum) and mucus hydrators such as nebulised hypertonic saline and inhaled mannitol.

**‘Physiotherapy now includes a combination of chest clearance techniques, exercise and inhaled mucolytic therapies’**

### Nutritional status

Malnutrition has strong associations with poorer clinical outcomes in CF and hence close attention to nutritional status is important.[16] Increased daily energy demands due to progressive lung disease require increased oral calorie intake including enteral tube feeding if oral supplementation is insufficient. Exocrine pancreatic insufficiency is present in 85% of the CF population where oral pancreatic enzyme replacement therapy (PERT) is given with fat and protein containing meals, snacks and drinks.[17]

Additionally, supplementation of the fat-soluble vitamins A, D, E and K is needed to optimise nutritional status. Poor nutritional status, pulmonary disease and glucocorticoid use can all have a detrimental impact on bone health and low bone mineral density is present in around 20% of individuals with CF.[1] Monitoring of vitamin D levels and DEXA are therefore routinely performed in at-risk patients.

### Close monitoring of complications

CF MDTs should offer a comprehensive annual review of all patients in order to optimise current management, assess pulmonary and nutritional status and screen for other complications.

CFRD is a distinct form of diabetes and is associated with more rapid decline in lung function and increased morbidity at lower levels of dysglycaemia than type 1 and 2 diabetes. It is therefore recommended that the CF MDT performs annual screening for the disease preferably involving continuous glucose monitoring.

An assessment of liver disease, renal disease and exercise capacity and a medication review are also recommended at least annually. »

**Table 4**

**Role of primary care in the management of cystic fibrosis patients**[2]

- Prescribing routine medications in batches of a month's supply or longer if advised by the specialist team
- Providing routine annual immunisation for patients with cystic fibrosis and their families
- Managing health problems not related to cystic fibrosis
- Supporting family members or carers
- Certification of illnesses
- Working in partnership with the cystic fibrosis specialist team at the end of life

People with CF face considerable psychosocial challenges and symptoms of anxiety and depression are up to three times higher than in the general population.[18] CF MDTs must assess mental wellbeing and other psychosocial indicators annually and facilitate CF tailored psychological input or psychiatric input where appropriate. GPs should be aware that the higher prevalence of anxiety and depression extends to family members and carers of people with CF.

### Disease modifiers

Drugs targeting the defective CFTR protein are being developed and are licensed for some genotypes. Ivacaftor is funded for use in patients with certain gating mutations (around 5% of the UK CF population) under the proviso that it can only be prescribed by specialist CF centres.[19]

In those with responsive genotypes, ivacaftor is associated with dramatic improvements in lung function, nutritional status, exacerbation rate and rate of pulmonary function decline.[20]

Licensed products for those with the more common F508del mutation are less effective and are not currently funded for use in the UK, but ongoing trials of other agents appear promising.[21]

## GP SUPPORT

The specialist CF MDT should provide timely information so that GPs can provide appropriate support to patients and their families, see table 4, above. Particularly close support may be required at the end of life, in partnership with the CF centre or local homecare teams, or at the time of transition from paediatric to adult services.

**Competing interests**
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***See also Diagnosing and managing cystic fibrosis in children, p19.***

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## Useful information

**For patients and their families**
**Cystic Fibrosis Trust**
www.cysticfibrosis.org.uk

# CPD prompt and framework
# Journal article

The Practitioner

**Show impact**
- Collection of data
- Practice audit to show improved care

- Case studies
- Implement a protocol
- Prescribing review
- Educational meetings
- Significant event audit
- Practice meeting
- Implement a new service
- Take on new role
- Significant event audits
- Change with feedback from patients and/or colleagues

**Further options for sessional GPs**
- Self audit of own practice:
  - Record-keeping
  - Referrals for investigations
  - Prospective condition based reviews
  - Random case analysis with colleagues
  - Review of telephone triage outcomes
- Update drugs in emergency bag

**Time credits**

**Impact credits**
Quick impact

Later impact

Later impact=

**Total credits**

## Title and reference

Date

What have you learned?

How will this change your practice?

What research/actions will you take that will have further impact on practice?

Draft of your action plan

# CPD prompt and framework
## Case review

The Practitioner

Clinical case for review

Reason for selection of the case for reflection

Evidence base used to reflect on your practice

What went well?

What could have been done better?

What were the causes of the condition/event?

What learning needs arise from this case for (a) you and (b) for the practice?

Summary of any discussion of the case with peers

What changes/actions have resulted to your personal practice?

What changes/actions have resulted to the practice?

How/when will you review these changes?

| Time credits | Impact credits | Total credits |
|---|---|---|

# CPD prompt and framework
## Audit

The Practitioner

Title

Reason for the audit (target to improve or implement)

Define criterion (or criteria) to be met

Set the standard and its basis

Plan the audit process and any involvement of peers

**Time credits** **Impact credits** **Total credits**

Reflection on Table 1 results

What changes were implemented to improve results?

Reflection on Table 2 results and review with peers

**Table 1**

**Results of data 1 analysis** **Date of data collection:**

| Criterion | Number sampled | Achievement (%) | Standard (%) |
|---|---|---|---|
| A | | | |
| B | | | |
| C | | | |

**Table 2**

**Results of data 1 and data 2 analyses** **Date of data collection:**

| Criterion | Number sampled Data 1 | Achievement (%) Data 1 | Number sampled Data 2 | Achievement (%) Data 2 | Standard (%) |
|---|---|---|---|---|---|
| A | | | | | |
| B | | | | | |
| C | | | | | |