The Cystic Fibrosis Association of Ireland

Physiotherapy for Cystic Fibrosis

TREATMENT INFORMATION
INTRODUCTION

PHYSIOTHERAPY FOR CYSTIC FIBROSIS
TREATMENT INFORMATION

This booklet has been written to assist you and your medical advisers. It isn’t intended to replace any advice you may receive from your physiotherapist or doctor. The Cystic Fibrosis Association funds medical and scientific research aimed at understanding, treating and curing Cystic Fibrosis. It also aims to ensure that people with Cystic Fibrosis (PWCF) receive the best possible care and support in all aspects of their lives.

THE CYSTIC FIBROSIS ASSOCIATION OF IRELAND

The Cystic Fibrosis Association of Ireland was set up in 1963 to increase knowledge and awareness of CF and to give advice and support to people with Cystic Fibrosis and to their families.

The Association:

• Provides support and assistance to people with CF and their families.
  – There are 22 branches of the Association throughout the country, with approximately 2,500 members nationally. The membership base comprises of people with CF, parents, family members and friends of the Association.

• Funds Medical Research:
  – The Association is currently funding four Research Projects which are helping in the vital search for a greater understanding of Cystic Fibrosis. The ultimate aim is to find a cure for CF but in the interim, it is imperative that we promote research that will improve on current methods of treatment.

• Supports specialist CF Multi-Disciplinary posts in hospitals around the country.

• Provides a domiciliary physiotherapy service to people with CF (PWCF). This home-based service is provided to young children with CF and promotes training for parents in new breathing techniques.

• CFAI campaigns both locally and nationally for increased and improved services for people with Cystic Fibrosis. Our aim is to ensure that the best possible services are available to all of our members.

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# CONTENTS

The Importance of Physical Treatment 3
The Lungs, CF and Physiotherapy 3
Lobes and segments of the lungs 4
How does CF affect the lungs? 4
Why is chest physiotherapy important? 5
How much physiotherapy should be done? 5
Assessment 6

**PHYSIOTHERAPY TECHNIQUES** 7
Gravity assisted positioning 7
Positions for draining different lung segments 8
The ‘active cycle’ of breathing exercises 11
Breathing Control 11
Thoracic expansion, exercise 12
Forced expiration technique 12
Chest clapping 13
Chest shaking 13
Positions and techniques for breathlessness 14

**APPLICATION OF TECHNIQUES: TREATMENT AT DIFFERENT AGES** 15
Babies 16
Toddlers and young children 18
Children 20
Adolescents and teenagers 22
Adults 23

**Other Physiotherapy Techniques** 24
‘PEP’ (Posture Expiratory Pressure) 25
High Pressure ‘PEP’ 25
Flutter 26
Autogenic drainage 26

**Other Important Aspects of Chest Care** 27
Inhaled medication 28
Exercise 29
Posture and Chest Mobility 30
THE IMPORTANCE OF PHYSICAL TREATMENT

One of the most important parts of the management of Cystic Fibrosis (CF) is physical treatment. The main aim is to keep the lungs clear and to maintain physical fitness - and, if possible, to improve it.

Over the years, physical treatment techniques have developed which are more effective and comfortable than before. There are different techniques for certain ages, for example children can now benefit from active breathing techniques which lead to independent treatment in adult life.

This booklet explains why physical treatment is necessary and gives a general description of chest physiotherapy, exercise and other important aspects of chest care.

THE LUNGS, CF AND PHYSIOTHERAPY

The body needs oxygen, which is carried around the body by the blood. The lungs are the organs which transfer oxygen from the air into the body.

The lungs are made up of a system of hollow tubes, or ‘airways’, which divide up, getting smaller and smaller towards the lungs’ edges. Eventually, these tubes end in air sacs, called alveoli. These are tiny pockets containing oxygen, which is transferred into the blood.

As we breathe in, air (containing oxygen) is sucked through the tubes, or airways, until it reaches the alveoli. As we breathe out, stale air is released.

The lining of the airways continually produces mucus. This keeps the airways moist and if dust, dirt or bacteria are breathed in, they get stuck in the mucus. The mucus can then be cleared from the lungs and swallowed. This is a continuous process and keeps the lungs clean and free of infection.
LOBES AND SEGMENTS OF THE LUNGS

The lungs are divided into separate sections, called ‘lobes’. Each lobe is further divided into small ‘lung segments’.

HOW DOES CF AFFECT THE LUNGS?

Lungs which are affected by Cystic Fibrosis produce mucus which contains less water than it should and is stickier. With healthy lungs, mucus moves naturally to the top of them, where it can be coughed up or swallowed. Lungs affected by CF can’t do this as easily, which means that nature’s way of keeping lungs ‘clean’ is less efficient. Bacteria can build up in the smaller airways and the lungs are more vulnerable to infection.

Infection is the main cause of lung problems for those with CF. The following are typical:

- bacteria build up in the mucus
- the lining of the airways becomes swollen
- the airways react by producing more mucus in an attempt
to get rid of the bacteria
- repeated infections can cause scarring of the lungs.
WHY IS CHEST PHYSIOTHERAPY IMPORTANT?

The aim of chest physiotherapy is to try to prevent the sticky lung secretions building up in the small air tubes. This helps to reduce infection.

HOW MUCH CHEST PHYSIOTHERAPY SHOULD BE DONE?

As far as the amount of treatment is concerned, patients with CF are advised as follows:

• A daily assessment should be made and treatment adapted accordingly (see Assessment on next page)

• The length of treatment sessions should vary according to need. This may be only 10-15 minutes if there are little or no secretions. It could, though, be much longer (perhaps 45-60 minutes) if there are lots of secretions to be cleared

• The number of treatment sessions per day should be varied. Most patients do two per day when all is well, increasing to 3-4 per day as necessary. If no secretions are present, some people may need treatment only once per day

• Adaptations should be made according to what’s noticed when making an assessment. For example, it may be necessary to concentrate on one particular part of the chest for a longer period if this is more productive.

In theory, the more physiotherapy that’s done the greater the possibility that the lungs will stay clear. In practice, though, a sensible amount of treatment should be fitted into a daily routine. Most people with CF have a set routine when they’re well but increase the amount of treatment when they’re coughing more than usual or when there are signs of active infection. It’s obviously essential to step up treatment in these circumstances.
ASSESSMENT

Assessment should be done as part of treatment and consists of the following four parts:
- Asking
- Looking
- Feeling
- Treating

**Asking** Ask yourself or your child whether you feel: unwell, tired, breathless, wheezy?
Have you noticed any other problems?

**Looking** Can you see any changes, such as:
- a faster breathing rate
- chest looking ‘overfull’ of air
- the chest being ‘sucked, in’ slightly when breathing in
- a larger amount of secretions
- stickier, darker coloured secretions

**Feeling** When you place your hands on the chest, can you feel any rattles or ‘crackles’ anywhere when breathing in or out? Does the skin feel warmer than usual indicating a higher temperature?

**Treating** Until you start the physiotherapy you may not notice anything different. Doing the physiotherapy itself is, in fact, part of assessment.

Are there more secretions than usual?
Are secretions harder to cough up?
Are there more secretions coming from a particular area?
Are there more noises from the chest when breathing?
Is treatment more tiring than usual?

If you notice any changes at all, it may be a sign of added infection and you may need to increase physiotherapy treatment and see your doctor.

Mother accessing her child’s chest before physiotherapy
This section describes, briefly, some basic physiotherapy techniques and how these can be applied at different stages. There are a variety of different physiotherapy techniques available but this booklet concentrates on treatment most commonly used in Ireland. It’s important that the techniques described are taught properly and reviewed by a physiotherapist.

**PHYSIOTHERAPY TECHNIQUES**

**Gravity assisted positioning**

Gravity assisted positioning is the use of different body positions to achieve:

- drainage of secretions from a particular area of lung, using gravity

- increased air flow to different parts of the lung Chest physiotherapy techniques should be carried out in different positions, so that each lung segment is kept as clear of secretions - and as fully inflated - as possible.
POSITIONS FOR DRAINING DIFFERENT LUNG SEGMENTS

1. Upper lobes - applicable segments

2. Upper lobes – anterior segments

3. Left upper lobe – posterior segment
4. Right upper lobe – posterior segment

5. Lingula

6. Right middle lobe

7. Lower lobes – apical segments
8. Lower lobes – posterior basal segments

9. Left lower lobe – lateral basal segment

10. Right lower lobe – lateral basal segment

11. Lower lobes – anterior basal segments
THE ‘ACTIVE CYCLE’ OF BREATHING EXERCISES

BREATHING CONTROL

AIMS
- to encourage relaxation and ease breathlessness
- to reduce over inflation of the upper chest and encourage a more normal breathing pattern
- to allow "relaxation" of the airways so that secretions can be cleared more easily

METHOD
- Rest your hand lightly on your tummy
- Try to relax the muscles around the neck and upper chest
- Breathe quietly and gently (as you breathe in, your tummy should swell slightly and, as you breathe out, sink down again). You should feel more movement around the waist and less around the upper chest

All breathing techniques should be taught by a physiotherapist.
THORACIC EXPANSION (DEEP BREATHING) EXERCISES

AIMS
- to loosen secretions
- to keep the chest mobile

METHOD
- Relax the upper chest
- Apply pressure with your hand to the lower part of the ribcage
- Breathe in slowly and deeply, filling up the lungs with air and expanding the lower chest as much as possible
- Release pressure with your hand and breathe out quietly

FORCED EXPIRATION TECHNIQUE (HUFFING & BREATHING CONTROL)

AIMS
- to move secretions from the smaller to the larger airways so that they can be cleared from the lungs more efficiently

METHOD

Stage 1: Huffing
- Take a medium sized breath in
- Squeeze the air out, contracting your tummy muscles and keeping your mouth and throat open. The huff should not be violent
- The breath out should be prolonged but not continued until the lungs are completely empty. This may make you cough as secretions are moved. If secretions do not come up in one or two coughs, try to stop coughing or you will become tired

Stage 2
- Do some breathing control

Stage 3
- Stages 1 and 2 may be repeated until secretions can be felt high in the chest

Stage 4
- When you feel secretions high in the chest, you should take in a deep breath and huff or cough to clear them
CHEST CLAPPING

This technique can also help to loosen secretions. A cupped hand is used to clap the chest firmly (it’s more comfortable through clothing or a towel). A friend or relative can do the chest clapping but many people do it themselves when they’re old enough.

Some people find that this technique is especially helpful in moving secretions if used with breathing exercises, although the technique is effective when used on its own.

CHEST SHAKING

METHOD

- Place your hands on the chest
- On the ‘out’ phase of a deep breath, shake the chest firmly, squeezing out the air in short bursts and applying the pressure inwards

You may not be successful with this method at first but keep trying - it takes practice to do shaking comfortably and effectively.
POSITIONS AND TECHNIQUES FOR BREATHLESSNESS

When you feel breathless, there are certain positions which can help you feel more comfortable. If you adopt one of these positions, and do some gentle breathing control, this can help direct air to the lower part of the chest, reducing overwork for the upper chest.

There may be times when you feel breathless walking uphill, going up and down stairs or exercising. In these cases, you should concentrate on how you’re breathing, because there’s a tendency to over-use the upper chest and to tighten up the muscles around the neck and shoulders.

- Try to relax tense muscles around the upper chest, neck and shoulders.
- Concentrate on ‘breathing control’ with a good breathing pattern; try not to have too much upper chest movement.
- Rhythmical breathing with activities can bring some relief. One example would be to time your breathing with walking, such as breathing in for one step and breathing out for two - or in for one and out for one. Your physiotherapist will be able to help you find out what works best for you.

Two examples of positions which can help ease breathlessness

Relaxed sitting

High side lying supported with pillows

Time breathing in and out with walking up stairs
APPLICATION OF TECHNIQUES TREATMENT AT DIFFERENT AGES

BABIES

Parents are often surprised that their babies like chest physiotherapy. Babies usually enjoy the physical contact and the rhythm of the chest clapping can even send them to sleep. It’s better, though, to treat babies when they’re awake, so that they’re aware that the treatment is being done and so that they become accustomed to it being part of their daily routine. Treatment is usually more effective when they’re awake, because breathing is more shallow during sleep.

Babies can’t do breathing exercises on their own, naturally, so treatment at this young age has to be passive, consisting of gravity-assisted positioning, chest clapping and possibly vibrations (fine shaking of the chest wall on breathing out).

Babies are all individuals and their condition will vary from day to day
BABIES

The following points should be borne in mind:

- Never treat a baby immediately after a feed; it may be sick
- The baby should be dressed in a vest or baby-grow to make the treatment more comfortable.
- The treatment is done most easily on your knee with your baby lying on a pillow, the body and head fully supported.

Father treating his baby’s chest using gravity - assisted positioning on his knee.

1. Lower lobes – posterior basal segments
2. Left lower lobe, lateral basal segment
3. Right lower lobe, lateral segment
4. Lower lobes, anterior basal segments
Hold the baby firmly, with the arms clear of the chest wall, so that the treatment can be done effectively without too much wriggling!

Firm chest clapping to the chest wall, and vibrations if they’re to be used, should be done with the baby positioned for drainage of a particular area of lung. Periods of chest clapping shouldn’t go on too long and there should be pauses for coughing and gentle, quiet breathing.

If the treatment is effective and there are secretions present, the baby will cough when the secretions reach the back of the throat. It doesn’t matter if the baby can’t spit out any mucus, because a good cough will clear secretions from the lungs and the baby will swallow them.

For young babies who can’t sit on their own, it’s important that the programme includes treatment in supported sitting, so that the tops of the upper lobes are drained.

It’s not possible to say how many positions should be used or how long the treatment should last, because each child is an individual and a child’s condition will vary from day to day. You should discuss this with your physiotherapist. However, many babies don’t need more than two 10-15 minute treatments a day when they’re well and their chest is clear.

You don’t necessarily have to stop treatment because the baby cries; unless, that is, the baby is distressed. Crying can be as effective in loosening secretions as breathing exercises.
**TODDLERS AND YOUNG CHILDREN**

Toddlers and young children aren’t the easiest age group to deal with at the best of times - and this applies equally to treatment! However, although it’s often difficult and frustrating to persuade a child to lie still and cooperate with treatment, it’s well worth persevering. Co-operation with treatment in future years is very often dependent on how things are handled at toddler stage.

It’s essential that a child learns from an early age that physiotherapy is something that has to be done every day. With patience and imagination, you can make physiotherapy a time that children enjoy and regard as a special time of day - a time when they have the undivided attention of mum or dad. This can be considered a transitional stage, when children’s treatment is changed and modified as they get older and can learn new techniques.

- When children are too big to be treated comfortably on the knee, they can have their treatment lying over a foam wedge.
- After children are two years old, treatment should progress from ‘passive’ to a more ‘active’ form. In the next couple of years, children can learn new breathing techniques which should be introduced gradually into their physiotherapy programme.
- A variety of ‘blowing games’ can be started. For example:
  - taking big deep breaths in and blowing little pieces of tissue paper
  - blowing bubbles
  - breathing out and steaming up a mirror

Children can play these games whilst on the wedge during physiotherapy sessions, either during or between short periods of chest clapping.
LEARNING TO HUFF

The games can be adapted gradually as the child begins to get used to the feeling of breathing in and out at different depths and rates. Eventually, the Active Cycle of breathing should replace these games, as the child gets older and is able to cooperate more.

- At this age, exercise can be introduced in a more formal way. It’s a good idea to encourage children to be physically active early on, so that they don’t get the idea that they’re not sporty - especially when they get to school. One great idea is to use a mini-trampoline, as young children usually love bouncing.

Bouncing on a mini-trampoline

You’ll need plenty of patience and imagination
CHILDREN

At this age, you should start to explain - in simple language — why physiotherapy is important and how their treatment is effective.

- Treatment can still be carried out on a foam wedge but there are also various physiotherapy ‘beds’ available which older children may find more comfortable.

- Once comfortable in the chosen gravity-assisted position, the Active Cycle of Breathing is performed. The separate parts of the breathing cycle are the same for everyone but the cycle should be tailored for the individual. For example, some people need to spend only 10 seconds on breathing control after a huff, whilst others may need to spend 30 seconds or more to make sure their airways don’t ‘tighten up’. The cycle should be practised with a physiotherapist, so that you and your child can learn to adapt the treatment to suit the child and to ensure that you’re being as effective as possible.

- Chest clapping should be done during the ‘deep breathing’ phase of the cycle. It shouldn’t be uncomfortable or too prolonged (no more than about 30 seconds at a time).

- If secretions can’t be cleared with two or three coughs, it’s better to stop and repeat the breathing cycle. Uncontrollable bouts of coughing aren’t just exhausting, they can force secretions back down again and make wheezing worse.

- The Active Cycle of Breathing should be repeated until the lung segment is as clear as possible. The number of positions treated, and the time spent in each position will depend on the amount of mucus and where it is. This will vary from day to day, and week to week. You should alter length of treatment and positions to meet changing circumstances. Treatment should never follow a rigid programme; constant reassessment is necessary.

By the time children are old enough to start school, they should be participating – actively – in their treatment. At this stage they’ll be old enough to understand the difference between breathing in and breathing out. And they should be able to carry out active breathing techniques.
Once children are old enough (usually from the age of about five) they should be encouraged to spit out the mucus, for the following reasons:

- you can detect the onset of active infection more easily (an increase in the amount of mucus, which is stickier and darker in colour, may indicate this)
- swallowing large amounts of mucus can cause nausea and sickness and suppress appetite
- it’s better to encourage children to get into the habit of spitting out the mucus before they get to an age when they become more self-conscious

It’s important to do the treatment even if there appear to be no secretions. The breathing exercises will be moving tiny little pieces of mucus from the smallest airways and this should help prevent problems developing.

Children should participate actively in their treatment

Mother and child carrying out treatment on specially designed tipping bed
ADOLESCENTS AND TEENAGERS

The onset of adolescence can be a difficult time for any child. Having Cystic Fibrosis can make it more so. As far as physiotherapy is concerned, there are two main areas of concern:

- The first is that it’s very common, for children to become rebellious about doing their physiotherapy at this age. This can be an extremely worrying and frustrating time for parents and it’s not an easy problem to deal with. Thankfully, most children gradually come out of the phase as they grow older, come to terms with their illness and realise the consequences of not doing their treatment properly.

- Sometime compromises have to be reached so that some kind of effective treatment is maintained. This may mean adopting different treatment techniques for a short time, or modifying the daily programme so that it’s acceptable to the individual. Constant nagging is usually counter-productive. It’s important that adolescents and teenagers have all the facts and that the physiotherapist makes sure they understand fully about how CF affects the lungs, why physiotherapy is necessary and how different techniques are effective. This is often a time when there are anxieties about the future and it’s useful if someone outside the family can spend time talking things through.

- The second problem at this age is the process of becoming independent. It’s important that teenagers start to take responsibility for their own treatment and do their own physiotherapy, so that they can look after themselves as adults. This is usually a gradual process and many are initially reluctant. Sometimes the teenager can be keen but the parents find it hard to hand over responsibility. It may take several years before both sides feel comfortable with the change of roles.

Teenager doing her own treatment using the active cycle of breathing, with self chest clapping

Constant nagging is counter productive
ADULTS

An adult with Cystic Fibrosis should be confident in self-care and should be competent in independent physiotherapy techniques. Everybody needs help with physiotherapy from time to time, especially if unwell, but you should feel confident about doing all your treatment yourself on a day-to-day basis. It’s useful to plan your day so that you can fit your treatment around your work and social life.

Starting a job can be difficult, because you may need to do some of your treatment at work. Many people find that it’s possible to choose positions which can be treated more easily at work (such as the upper lobes). Other positions can be left for when you’re at home.

You should maintain an active lifestyle into adulthood. This will require more effort when school activities are no longer available to you. If you find that you get breathless with certain activities, don’t avoid exercise but use breathing techniques.
OTHER PHYSIOTHERAPY TECHNIQUES

There are other physiotherapy techniques available which many patients find useful. In order for them to be used most effectively, they need to be taught by physiotherapists who have had training and experience in their use.
‘PEP’ (POSITIVE EXPIRATORY PRESSURE)

Positive expiratory pressure is usually given through a mask to provide resistance to breathing out. This helps to keep small airways open and to loosen secretions. One of the benefits of PEP is that it’s performed in the sitting position. This makes it convenient to use when space is limited, as it may be at work, in school, or when travelling.

Treatment consists of breathing through a face mask with different sizes of resistors, for about ten breaths. The mask is then removed before the forced expiration technique (huff and breathing control) is performed to clear secretions.

It’s important that a physiotherapist carries out a full assessment, so that the correct level of resistance can be chosen. This will be re-assessed regularly, particularly during a chest infection.

HIGH PRESSURE ‘PEP’

This method uses the same mask as PEP but with differences in breathing technique and higher pressures. For this reason, physiotherapy with lung function testing is essential before this technique can be used.
**FLUTTER**

The flutter is a hand-held device which produces a vibrator effect to the airways during breathing out. The frequency of the vibrations depends on the angle at which the flutter is held.

It’s usually performed in sitting position, with elbows supported on a table. The patient holds the flutter in the mouth, breathing in through the nose and out through the flutter. The breathing exercises consist of slightly deeper breaths through the flutter with the forced expiration technique. A physiotherapist assessment is necessary to find the best breathing cycle and correct flutter angle, to provide maximum, effect.

**‘AUTOGENIC DRAINAGE’**

Autogenic drainage is a breathing technique which needs very controlled breathing at different levels in the lungs. Each breath consists of a slow gentle breath in, a breath hold for three seconds and a fast breath out. Throughout the technique patients must keep their throats open to allow the free passage of air. This Technique needs relaxation and concentration and should be taught by a physiotherapist who’s been specially trained in the technique.
OTHER IMPORTANT ASPECTS OF CHEST CARE

Inhaling ‘nebulised’ bronchodilator drug through mouthpiece
INHALED MEDICATION

Certain drugs can be inhaled straight into the lungs when they’re needed. Some of these, such as bronchodilator drugs (which help to open up the airways), should be given before chest physiotherapy. Others, such as antibiotics, steroids and DNase, should be given after chest physiotherapy, so that the lungs are as mucus-free as possible. If you’re unsure, check with your doctor, nurse or physiotherapist.

There are various ways that inhaled medication can be taken:

**Inhaler devices**

Various inhaler devices are available. Some give the drug as a powder and some as an aerosol. You must make sure that you’re instructed how to use these devices correctly, so that the medication gets into the lungs where it’s needed.

**Nebulisation**

The drug is delivered as a fine mist which is breathed in. You may be given, an air compressor and a ‘nebuliser’ for use at home.
IMPORTANT POINTS FOR TAKING NEBULISED DRUGS:

- A mask can be attached to the nebuliser for babies and small children, although a mouthpiece should be used as soon as the child is old enough.

- Breathe through the mouthpiece (not through the nose) and take deep breaths every few breaths to ensure that the drug gets as far as possible into the lungs.

- Always keep nebuliser pots clean and sterilised; otherwise they may harbour bacteria which could cause infection. Nebuliser pots also have tiny holes in them which can become partially blocked if they’re not kept clean - if that’s the case, they won’t nebulise properly. Instructions for caring for your compressor and cleaning equipment should be provided.

EXERCISE

It has been proven that physical activity can keep people with CF healthy. Exercise should be introduced from an early age, so that children grow up with, as much physical activity fitted into their week as possible.

Many young children enjoy trampolining on mini-trampolines, which can be bought quite cheaply and kept at home. Exercise programmes can be carried out at home but it’s often more enjoyable to get into the fresh air and take part in sports or team games. Choose types of exercise which make you out of breath, such as running, swimming, football or tennis.

It’s important that schools know that exercise is something to be encouraged. Some schools aren’t sure whether exercise is good for people with CF or not.

After leaving school, it’s a good idea to consider joining a sports club, so that sporting activities can be continued.
POSTURE AND CHEST MOBILITY

The spine, rib cage and shoulders should remain fully flexible as far as possible – and it’s important that a good posture is maintained.

Older children or adults may need to do some specific stretching exercises to maintain full movement of the joints and muscles around the shoulders and chest.

Younger children can achieve this with games or activities which involve moving and stretching the trunk and arms. Your hospital physiotherapist should be able to advise on the right type of exercises and activities.

Poor sitting posture showing rounded spine

Sitting with straight back to correct poor posture and stretch the joints and muscles
Games such as ‘wheelbarrows’ can make good stretching exercises, as well as helping to drain secretions
PUBLICATIONS

Other Publications from CF Ireland

Booklets

THE FACTS AND FINDING OUT
An introduction into the causes and effects of Cystic Fibrosis.
A guide for parents of newly diagnosed children with Cystic Fibrosis.

NUTRITION
Eating well with Cystic Fibrosis – A guide for children and parents.

TREATMENT
Physiotherapy for Cystic Fibrosis. Illustrates methods of physiotherapy in CF.

RIGHTS AND ENTITLEMENTS
A guide to rights and entitlements for people with CF and their families.

Leaflets

CFAI GENERAL LEAFLET
Outlines the structures and help available from the Cystic Fibrosis Association

CYBER CAMPUS LEAFLET
Description of online computer course for young adults with CF.

THE FACTS
General introduction to causes and effects of CF.

Magazines

ANNUAL NEWSLETTER
Yearly magazine which looks at the work and achievements of the Association.

FUTURE FORCE
Magazine designed and written by CF adults covering large a range of topics and issues affecting CF adults.

Further enquiries about literature, including booklets produced by the Association can be obtained from

The Cystic Fibrosis Association of Ireland
24 Lower Rathmines Road, Dublin 6
LoCall: 1890-311-211
Phone 01-4962433 / Fax 01-4962201
Email: info@cfireland.ie
Web: www.cfireland.ie