

Cystic **Fibrosis** *our focus*

Steroid treatment in cystic fibrosis

Factsheet – August 2015

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Introduction

Steroids are used for their powerful anti-inflammatory action and can be taken in a number of ways to control inflammation in the airways. This factsheet provides information on how they are used in cystic fibrosis, the different types of steroid available and the possible side effects. When a clinician is prescribing a new medicine such a steroids, they will weigh up the positive actions of the medication over the possible side effects.

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Last reviewed July 2015.

Next review date July 2016.

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What are steroids?

Steroids are a group of medicines that have powerful anti-inflammatory action. There are a number of different types, but those potentially used in people with cystic fibrosis are the group also known as corticosteroids or glucocorticoids. These are not the same drugs that are used by some body builders and athletes.

Steroids can be taken in a number of ways:

- Inhaled into the lungs (inhaled steroids)
- Swallowed by mouth (oral steroids)
- Injected into the blood stream (intravenous steroids)
- Injected into a muscle (intramuscular steroids)
- Sprayed up the nose (nasal steroids).

Inhaled steroids

Names

The most commonly-used inhaled steroids are beclometasone (Becotide, Clenil, QVAR) and budesonide (Pulmicort), which both come as beige and brown inhalers, and fluticasone (Flixotide), which is an orange inhaler.

Sometimes, steroids are combined in the same inhaler with a long-acting medicine that helps relax the muscles of the airways. These are Seretide (fluticasone plus salmeterol), which is a purple inhaler, or Symbicort (budesonide plus formoterol), which is a white turbobalmer with a red base.

How are they given?

As their name suggests, inhaled steroids are inhaled into the lungs. A number of different types of inhaler may be used, and the choice tends to depend on the person's age.

- Inhaled steroids are usually best given by a spacer device to reduce the amount of steroid deposited into the mouth; this can be used at all ages, and is the safest way to take high doses.
- They may also be given in a dry powder form, which is suitable for those aged five years and above.
- They can also be sprayed directly into the mouth by a meter-dose inhaler, but these are more difficult to use, less efficient at getting the drugs into the lungs, and unsuitable for children.
- The mouth must be rinsed out after taking inhaled steroids, particularly if they are taken in powder form.
- The lowest dose possible should always be used to control symptoms.
- They are given twice daily, and sometimes once daily when the dose is being weaned down.
- It is important for people to know the actual dose they are taking (the strength of the inhaler), for example 200 micrograms twice a day, rather than just the number of puffs inhaled.

Uses

- Inhaled steroids may be required for people with wheezing or chest tightness that requires regular bronchodilators, drugs that widen the air passages by relaxing muscles, to improve their symptoms (blue inhalers – salbutamol or terbutaline). Generally, if someone requires a bronchodilator several times a week for symptoms, an inhaled steroid should be started. This is quite common in younger children. It may also be necessary in older children and adults thought to have CF asthma, which is the presence of asthma-like symptoms in someone with cystic fibrosis. There is no extra benefit in increasing the dose during viral colds or wheezy episodes.
- There is no evidence that inhaled steroids are of benefit in treating allergic bronchopulmonary aspergillosis (ABPA).
- They are sometimes given over a longer term to reduce inflammation in the lungs resulting from repeated infections, but it has been difficult to prove that they are beneficial for this.

Side effects

- If high doses are used, growth may be slowed down. This is usually (but not always) reversible. Generally, the final height is unaffected.
- Rare reports have been published of problems affecting production of certain hormones, which can lead to low blood sugar, particularly if the person is acutely unwell. This tends to be with high doses but has been reported in people with CF who were also taking the antifungal antibiotic itraconazole.
- Oral thrush (caused by the fungal infection *Candida*) is occasionally seen, and may be avoided by rinsing the mouth after taking the inhaler.
- Hoarseness of the voice has also been reported, though this is rare.

Conclusions

- Inhaled steroids can be very useful for people having difficulties with tight airways and recurrent wheezing.
- They should be used at the lowest dose possible, and doctors should consider whether the dose can be reduced and whether the person still needs to take it.
- Inhaled steroids are best given via a spacer device, especially at higher doses.

Oral steroids

Names

Prednisolone or dexamethasone.

How are they given?

- Prednisolone Soluble 5-mg tablets (pink) can be dissolved in water.
- Otherwise 5- or 25-mg tablets can be taken, but it is important not to use the enteric-coated ones as they are poorly absorbed.
- Dexamethasone is used less often and comes as liquid medicine or tablets.
- They are usually taken once a day (in the morning), or sometimes every other day.
- They should be taken with or after food.

Uses

- Allergic bronchopulmonary aspergillosis (ABPA) is an allergic reaction to the fungus *Aspergillus*, which is commonly isolated in sputum of people with cystic fibrosis. When this diagnosis is proven, oral steroids are an important part of the treatment, and may be necessary for a number of months.
- Acute, severe wheezing or very tight airways may need a short course of oral steroids, eg three to seven days.
- They are sometimes given to patients having a severe chest exacerbation (worsening of symptoms) who are receiving intravenous antibiotics, but the benefit of this has been difficult to prove.
- Occasionally, oral steroids are used in people with CF with severe joint pains and swelling (CF arthropathy), although usually ibuprofen is enough to treat this.
- They are often used as part of immunosuppression treatment after lung transplantation.
- Long-term use of oral steroids to reduce lung inflammation is not advised as the risk of significant side effects outweighs any small benefit that may be seen.



Side effects

- Side effects are more likely with oral steroids than with inhaled steroids and depend on the dose and length of treatment and some individuals are more susceptible than others. Repeated shorter courses can also be cause side effects.
- It is very important that oral steroids are not stopped suddenly if they have been taken for more than seven to ten days: the dose must be gradually reduced under supervision.
- If someone has been on oral steroids for a while, it is important the dose is increased if they require surgery or are particularly unwell.
- Glucose intolerance: blood sugar levels may be too high and sometimes insulin-dependent diabetes may occur in patients who were on the verge of developing it. People on regular oral steroids should have a urine test for glucose when seen in clinic.
- High blood pressure can develop, so this must also be checked regularly.

- People taking oral steroids can sometimes retain fluid and put on weight, particularly around the face. This goes away when the steroids are stopped.
- Quite a few people find their appetite, and therefore food intake, increase markedly, although this is not permanent.
- Some people can develop skin bruises easily, and some might develop stretch marks on the abdomen.
- Occasionally, people find their mood is altered and they can feel unusually irritable.
- Long-term steroids can cause thinning of the bones and rarely, cataracts have been reported.
- If a child on long-term steroids has not had chicken pox, they should avoid close contact with people who have got chicken pox or shingles. If they are exposed to another child with chicken pox, medical advice must be sought, as an injection of Varicella Zoster Immunoglobulin should be given.

Conclusions

- The use of oral steroids must be fully justified because of the high potential for side effects when given long term.
- Nevertheless, they are sometimes required, especially for treatment of ABPA.
- The lowest effective dose and shortest course should be used.

Intravenous steroids

Names

Hydrocortisone or methylprednisolone.

How are they given?

Intravenously, ie via a cannula (tube) placed in a vein, or via a totally implantable venous access device (eg a port). Hydrocortisone is usually given every four to six hours. Methylprednisolone may be given daily or twice daily for three days every month.

Uses

- Acute, severe wheezing, used in a similar way to someone having a severe asthma attack.
- Severe allergic reactions (anaphylaxis), usually to an antibiotic.
- Episodes of acute rejection after lung transplantation.
- Intractable (hard to treat) wheezing or severe recurrent small airways disease may require short pulses of methylprednisolone.
- Allergic bronchopulmonary aspergillosis (ABPA) usually responds to oral steroids but the use of three-day pulses of methylprednisolone given monthly has been reported.

Side effects

See section on oral steroids.

Conclusions

Intravenous steroids are not used very often, but are occasionally necessary in severe situations.

Nasal steroids

Names

There are a number of preparations that can be taken via the nose, for example Mometasone (Nasonex), Fluticasone (Flixonase), Avamys (Fluticasone), Beclometasone (Beconase) or Budesonide (Rhinocort).

How are they given?

Nasal steroids are usually taken as nasal sprays or drops once or twice a day. It is important that the head is held in the right position to ensure they have the best chance of working.

Uses

- Nasal steroids are used to treat nasal polyps that are causing troublesome symptoms such as a blocked nose, headaches, impaired smell and taste.
- They can also be used to treat hay fever (allergic rhinitis).

Side effects

Side effects are not usually seen with nasal steroids, although caution is advised if they are taken together with inhaled steroids. Side effects are more likely with the nasal drops than sprays.

Conclusions

- It is worth trying nasal steroid treatment for nasal polyps, but success is limited.
- Nasal steroids can be useful against hay fever during the summer months, usually together with an antihistamine.

Additional reading

These Cochrane systematic reviews have plain language summaries and are available on the internet at www.thecochranelibrary.com/view/0/index.html

Balfour-Lynn IM, Welch K. Inhaled corticosteroids for cystic fibrosis. Cochrane Database Syst Rev. 2014 Oct 9;10:CD001915. doi:10.1002/14651858.CD001915.pub4.

Cheng K, Ashby D, Smyth RL. Oral steroids for long-term use in cystic fibrosis. Cochrane Database Syst Rev. 2013 Jun 24;6:CD000407. doi:10.1002/14651858.CD000407.pub3.

Further information

The Cystic Fibrosis Trust provides information about cystic fibrosis through our factsheets, leaflets and other publications.

Most of our publications are available through our helpline and can be downloaded from our website or ordered using our online publications order form. Visit cysticfibrosis.org.uk/publications.

The Cystic Fibrosis Trust helpline can help you with a range of issues, no matter how big or small. Our trained staff can provide a listening ear, practical advice, welfare/benefits information or direct you to other sources of support. The helpline can be contacted on 0300 373 1000 or helpline@cysticfibrosis.org.uk and is open Monday to Friday, 9am – 5pm.

Calls to 0300 numbers cost no more than 5p per minute from a standard BT residential landline. Charges from other landlines and mobile networks may vary, but will be no more than a standard geographic call and are included in all inclusive minutes and discount schemes. If you are worried about the cost of the call please let us know and we'll call you back.

You can also find more information at our website cysticfibrosis.org.uk.

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