factsheet

CYSTIC FIBROSIS TRUST

STEROID TREATMENT IN CYSTIC FIBROSIS

Written by Dr Ian Balfour-Lynn, Director of Paediatric CF Services, Royal Brompton Hospital, London and approved by the Cystic Fibrosis Trust Medical Advisory Committee

This factsheet contains information on steroid treatment in CF, including the different types of steroids, what they are used for, how they are given and their possible side effects.

The information has been written to assist you and your medical advisers. It is not intended to replace any advice you may receive from your Specialist CF Centre or CF Clinic.

Introduction

Steroids are a group of medicines that have powerful anti-inflammatory actions. There are a number of different types, but those potentially used in people with CF are the group also known as corticosteroids or glucocorticoids. These are not the same drugs as those that can cause serious problems in body builders and athletes who ought not to be taking them.

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)
- sprayed up the nose (nasal steroids).

Patron: HRH Princess Alexandra, the Hon. Lady Ogilvy, KG, GCVO President: Duncan Bluck CBE
Registered as a charity in England and Wales (1079049) and in Scotland (SC40196)
A company limited by guarantee registered in England and Wales number 3880213
Registered office: 11 London Road, Bromley, Kent BR1 1BY.

Inhaled steroids

Names

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

Sometimes they 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 turbohaler 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 inhalers may be used, and the choice tends to depend on the person's age.

- Usually inhaled steroids are 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 to six years and above.
- Finally, they can 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, and 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 require regular bronchodilators to improve their symptoms (the 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 allergic bronchopulmonary aspergillosis (ABPA).
- They are sometimes given long term to reduce inflammation in the lungs resulting from repeated infections, but it has been difficult to prove 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 though, the final height is unaffected.
- Rare reports have also been published of problems affecting production of certain hormones, which can lead to low blood sugars, 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 rarely been reported.

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.
- They are best given via a spacer device, especially at higher doses.

Oral steroids

Names

Prednisolone or dexamethasone.

How are they given?

- Prednisolone Soluble 5mg tablets (pink) can be used and dissolved in water.
- Otherwise 5 or 25mg tablets can be taken but it is important not to use the enteric-coated ones otherwise they are poorly absorbed.
- Dexamethasone is used less often and comes as a 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 the 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, e.g., three to seven days, similarly to how they are used in asthmatic patients.
- They are sometimes given to patients having a severe chest exacerbation who are receiving intravenous antibiotics, but benefit has been difficult to prove.
- Occasionally, oral steroids are used in people with CF with severe joint pains and swelling (CF arthropathy), but 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 to happen with oral steroids than with inhaled steroids and depend on the dose and length of treatment, although some individuals are more susceptible than others. Repeated shorter courses can also be a problem.

- 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 the person requires surgery or is 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 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 increases markedly and they eat a lot, although again this is not permanent.
- Some people can develop skin bruises easily, and some can 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 try and 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 i.e., via a cannula (drip) placed in a vein, or via a totally implantable venous access device (e.g., a portacath). 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 antibiotics.
- Episodes of acute rejection after lung transplantation.
- Intractable (hard to treat) wheezing or severe recurrent small airways disease may require short pulses of methylprednisolone.
- 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), 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 e.g., 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 quite helpful for 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:

http://www3.interscience.wiley.com/cgi-bin/mrwhome/106568753/HOME

Balfour-Lynn IM, Welch K. Inhaled corticosteroids for cystic fibrosis. *Cochrane Database of Systematic Reviews* 2009, Issue 1. Art. No.: CD001915. DOI: 10.1002/14651858.CD001915.pub2.

Cheng K, Ashby D, Smyth RL. Oral steroids for cystic fibrosis. *Cochrane Database of Systematic Reviews* 1999, Issue 4. Art. No.: CD000407. DOI: 10.1002/14651858.CD000407.

Further information

If you have any questions that have not been answered in this factsheet, you can contact the Cystic Fibrosis Trust Support Service: ☎ **0300 373 1000**

For further general information and literature published by the Cystic Fibrosis Trust please contact:

Cystic Fibrosis Trust
11 London Road
Bromley
Kent BR1 1BY
☎ 020 8464 7211
Email: enquiries@cftrust.org.uk

To view or download Cystic Fibrosis Trust publications, visit our website:
www.cftrust.org.uk/aboutcf/publications

© Cystic Fibrosis Trust 2010. This factsheet may be copied in whole or in part, without prior permission being sought from the copyright holder, provided the purpose of copying is not for commercial gain and due acknowledgement is given.