

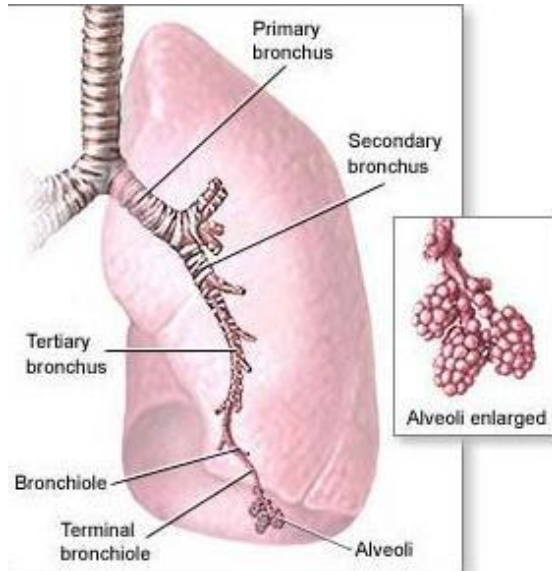
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## BRONCHIOLITIS OBLITERANS

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### What is Bronchiolitis Obliterans?

Bronchiolitis Obliterans (BO) refers to a condition which affects the small airways in the lungs (the bronchioles). It is also known as Obliterative Bronchiolitis (OB). The bronchioles conduct air from the large airways (bronchi) to the alveoli where oxygen enters the bloodstream. The term refers to chronic inflammation within these small airways.



[www.greenfacts.org](http://www.greenfacts.org)

### What causes it?

The causes of BO are varied. It can occur after certain infections (e.g. adenovirus, mycoplasma); after bone marrow transplant or heart-lung transplant; with some types of connective tissue disorders; with gastro-oesophageal reflux with aspiration; after inhalational injury from smoke or other irritants, or after a rare reaction to some infection or medicines which is called "Stevens-Johnson Syndrome". In some cases we can only presume there has been an infection, but can never prove it.

### Is it common?

BO is very rare. There are no reliable figures to say how many cases might be expected in a country the size of New Zealand. However our observation is that there may be up to 50 children suffering from it throughout the country.

### How is it diagnosed?

The diagnosis of BO may be considered after a child has an episode of illness which is known to possibly cause it (e.g. infection with a known risk factor virus; after a bone marrow transplant, after being in a house fire etc). If your child's Respiratory Specialist thinks BO may be the cause of your child's problems, they will ask for a CT scan of your child's lungs.

From these results they may feel confident to diagnose BO. Very occasionally, they may recommend that a biopsy (sample) is taken from your child's lung. More information on lung biopsy can be found on the parent information sheet called "Lung biopsy"

### How is it treated?

Treatment of BO is difficult. There is no standardised scheme of drugs, although most experts would start with oral steroids (prednisolone) in the early stages. Steroids are powerful anti-inflammatory drugs. They are used to “damp down” the inflammation that is damaging the small airways. In some children however, by the time the BO is recognized the inflammation may have settled and the airways are left severely narrowed by scarring. In this situation, steroids cannot do any good, and sometimes can be harmful if they are continued when they are not helping. Your child's Paediatrician will be able to advise whether your child should have steroids or not.

It is also impossible to predict how long any child will need to be on treatment.

Many children with BO also require oxygen at home. This can also be given at school, or via a portable cylinder. Some children show a response to bronchodilators (salbutamol or terbutaline). These can be given, but they only provide temporary, symptomatic relief, and do not affect the overall progress of the disease.

### Are there any problems from the long term treatments?

BO can be a very aggressive illness, with the potential to continue to damage your child's lungs. The underlying cause of the BO can help your child's Chest Specialist to decide how likely it is to progress. In those cases where it is unlikely to progress (e.g. after smoke inhalation) there may only be a need for steroids for a few months. In other cases (e.g. where it occurs after a bone marrow transplant) the chance of continued progression is greater, and so other treatments may be considered. Unfortunately there are no published trials of drug treatments, and no one drug has been shown to be the treatment of choice.

The long term problems from steroids are well documented, and include: raised blood pressure; weight gain; glucose intolerance; poor wound healing; infection risk; poor growth; cataracts. Those underlined are fairly common.

Other drugs which might be considered include methotrexate, hydroxy chloroquine, azathioprine, and cyclophosphamide. Their respective side effects are given below:

- Hydroxy-chloroquine: cataracts, hearing loss, muscle weakness, convulsions. These are usually very rare.
- Azathioprine: muscle weakness, liver disturbance/jaundice, blood disorders/ bone marrow failure.
- Methotrexate: nausea, vomiting, liver function abnormalities, blood disorders, rarely cirrhosis, skin sensitivity to sunlight, hair loss, cough/shortness of breath.
- Cyclophosphamide: fatigue, mild nausea and vomiting, loss of appetite, blood disorders, hair loss, reduced fertility, mouth ulcers, diarrhoea,

### **Can BO be cured by this treatment?**

Usually BO cannot be cured (i.e. go away for ever after a course of treatment). The aim of treatment is to stop the BO from getting worse. However, after the initial injury (infection or other cause) it is usual for there to be a small degree of recovery. Sometime in cases of BO, if there is additional bacterial infection, a different type of lung damage, called "bronchiectasis" occurs. There is an information sheet on bronchiectasis available from the [Kids Health website](#). It may be necessary to start other treatments to limit the effects of further damage from bacterial infection. These additional treatments may include regular physiotherapy, or long term antibiotic treatments.

### **Are there any long term effects from BO?**

This is very difficult thing to predict for an individual child. The more severe the inflammation, and the more difficult it is to get it under control, the more likely your child is to have severe scarring of their lungs. Once scarring has formed, it cannot be reversed. Scarring makes additional infections more likely, which in turn cause even more scarring. Sometimes the scarring itself can cause persistent infection. This is called bronchiectasis.

In some cases, a child's BO may progress to a point where it is life threatening. This is unusual. If your child's BO is progressive, your child's doctors may wish to discuss lung transplant with you. There is another parent information sheet available on "Lung Transplant".

### **What else can parents do?**

It is important that no-one smokes at home. Cigarette smoke will not cause BO, but will aggravate the inflammation, and increase the risk of additional infections.

Your child will need influenza vaccinations (from your GP) every autumn. Depending on your child's age, they may require other vaccinations either before winter, or sometimes throughout the winter period. Your Respiratory consultant will discuss these with you.

There is very little else parents can do to alter the underlying course of the disease. It is very important you make sure your child gets their medications regularly. If you feel your child ought to stop taking their medicines, you *must* discuss this with your child's Respiratory specialist first.

### **Where can I get more information on BO?**

Because it is so rare in children, information is not easy to come by. There are internet sites that have some details (e.g. [www.emedicine.com](http://www.emedicine.com); [www.bpold.co.uk](http://www.bpold.co.uk)). However, when getting information from the internet, it is very important the remember two things:

- Of the almost 300,000 hits on Google for "bronchiolitis obliterans" approximately 200,000 relate to BO in adults. BO in children refers to a **different** group of diseases, and we would advise caution in applying information on adult BO to your child.
- The fact that childhood BO can have many different causes means that any information found on Google (even about childhood BO) may not directly apply to your child.

If you find some information on BO, and you have questions about it, please print it off, and bring it with you to your next clinic appointment. Your child's Paediatric Specialist can then discuss it with you.

This information sheet is produced to answer some of the questions parents ask about Bronchiolitis Obliterans. It is not intended to replace discussion with your child's Respiratory Specialist and you are encouraged to discuss your child's condition with the specialists when you attend clinic.