

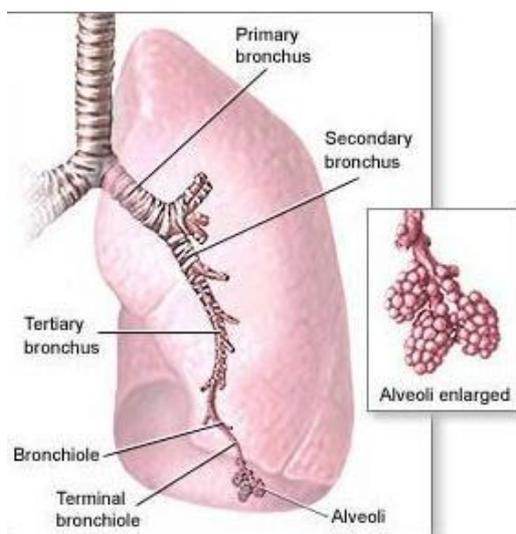
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## INTERSTITIAL LUNG DISEASE (ILD)

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### What is Interstitial Lung Disease?

Interstitial lung disease (ILD) refers to a group of conditions which affect the lungs. ILD causes severe, chronic inflammation within the lungs, which can lead to permanent scarring. This inflammation affects the alveoli (air sacs) and the smallest airways (terminal bronchioles) in the lungs. The alveoli are the bit of the lung which actually get oxygen into the blood, and allow carbon dioxide to leave the blood stream.



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

There are two broad types of ILD seen in children. Older children (usually of school age) can develop forms of adult ILD. This is very rare, and may be in children who have another underlying disease (e.g. problems like scleroderma), or after severe viral infection. The other type of ILD is that seen in pre school children (usually those less than 2 years) which may be due to a viral infection, or may be a problem that the child has been born with. Sometimes a problem a child is born with can take several months to get bad enough to show itself fully.

Some forms of ILD (infant or adult type) are progressive, and may even be fatal if treatment cannot control symptoms. Other kinds of the infant type are self limiting, and resolve themselves after a few years.

### Is it common?

Data from the UK suggests that approximately 3-4 children per 1,000,000 population will have some form of ILD (infantile type or adult type). This would suggest that there may only be 12-16 children with this problem throughout the whole of New Zealand.

### How is it diagnosed?

The diagnosis of ILD is usually made after a biopsy (small sample) of the lung has been studied under the microscope. The possibility of your child having ILD may be considered by your child's chest specialist, after a careful history has been taken from you, and we have examined your child. The specialist will very likely do some initial tests (including a CT scan of the chest), which may show that further information is required. On the basis of the results from these initial tests, a lung biopsy would then be recommended. You may also wish to read our information sheet on lung biopsy.

### How is it treated?

The mainstay of treatment for ILD is steroids. These are strong anti-inflammatory drugs which are used to “damp down” the inflammation that is going on in the lungs. In some children however, the inflammation is too strong for just steroids to help and other more powerful anti-inflammatory drugs are needed. These include hydroxy-chloroquine, methotrexate, azathioprine or cyclosporin. Treatment is likely to be needed for a number of months, if not years. It is impossible to predict how long any child will need to be on treatment. Your child’s specialist will try to reduce the treatment as soon as possible, but if your child gets worse as a result of cutting of down the treatment, then the drugs will need to be stepped up again.

In spite of treatment, some children need oxygen to be given at home (some just need it at night, and some need it for all day and night).

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

ILD is a very powerful disease. If untreated it can be fatal for some children and powerful drugs are needed to fight it. Powerful drugs will always have side effects. One of the decisions you will need to make is the choice between getting proper control of the IPF, and limiting side effects from treatment. Below are some of the more common side effects from the drugs:

- Steroids: raised blood pressure, weight gain, glucose intolerance, poor wound healing, infection risk, poor growth, cataracts. Those underlined are fairly common.
- 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, infertility, mouth ulcers, diarrhoea,

### Can ILD be cured by treatment?

In many cases ILD cannot be cured (i.e. go away for good after a course of treatment). It can be controlled, and in the vast majority of children they can lead an active life, even if they require some medication every day, or every other day. Overall, the cure rate for ILD is not as good as we would like. The fact it is such a rare disease means that it can be difficult to give an accurate idea of what the chances of survival are.

### Are there any long term effects from ILD?

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 chronic 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 lead to chronic infection. This is called bronchiectasis.

If the scarring is sufficiently serious your child may need long term oxygen treatment (either just at night, or sometimes day and night). If the ILD is very aggressive then the dose and duration of the treatments will need to be maximised, and this will increase the risk of drug side effects (as discussed above).

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

It is important that no-one smokes at home. Cigarette smoke will not cause ILD, 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 ILD?**

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 to remember two things:

- Of the >1.5 million hits on Google for "interstitial lung disease" almost all of them will relate to ILD in adults. ILD in children is not exactly the same as in, and has a different range of causes, and survival figures.
- The fact that childhood ILD can have many different causes means that any information found on Google may not directly apply to your child. If you find some information on ILD, and you have questions about it, please print it off, and bring it with you to your next clinic appointment. We can then discuss it with you.

This information sheet is produced to answer some of the questions parents ask about Interstitial Lung Disease (ILD). 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.