Kartagener’s syndrome

What is it?
This condition involves recurrent infections usually affecting the lungs, ears and nose which, if left untreated, may lead to long term lung damage. It is thought to be an inherited syndrome which may consist of several features:

- **Situs inversus** (major organs are on a different side of the body to normal – for example, heart on the right rather than the left).
- **Primary ciliary dyskinesia (PCD)**. This is a condition in which the microscopic hairs (cilia) within organs, such as the nose, ears and lungs, do not function normally, with the result that mucus and bacteria are not cleared effectively. It can cause chronic glue ear, rhinitis, sinusitis and recurrent chest infections.
- **Bronchiectasis** (abnormal dilatation of the airways).

Other terms
Kartagener’s syndrome may also be referred to as Kartagener’s triad, Siewert’s syndrome or Afzelius’ syndrome. Kartagener’s syndrome is a combination of PCD and situs inversus.

Who does it affect?
It is usually diagnosed in infancy or early childhood. It can occur in all races and is equally present in boys and girls. The incidence is thought to be approximately 1 in 40,000.

What causes it?
It is an inherited condition. The genetic defect has still not been identified and your child may be the first in your family to be affected. Genetic counselling may be offered to you.

Signs and symptoms
These can vary in degree and severity between individuals. Not all symptoms will affect every child. They may develop as a consequence of recurrent chest infections.

- Difficulty in breathing
- Productive cough
- Wheeze
- Repeated chest infections
- Nasal congestion and polyps (swellings)
- Recurrent ear infections
- Recurrent sinusitis
**Diagnosis**

Diagnosis is based on a series of investigations including:

- Chest X ray
- Chest CT scan
- Nasal ciliary biopsy (sample of the hair cells in the nose)
- Lung function tests
- Hearing test
- Nasal nitric oxide measurement

**Treatment**

A combination of physiotherapy and a low threshold for antibiotics are recommended to reduce the severity of lung disease.

- **Physiotherapy** - this will be taught to you by a physiotherapist and may be recommended on a daily or intermittent basis. This normally consists of the positioning of your child plus various techniques, which you will be taught. The techniques will allow gravity to aid effective clearance of chest secretions. As children become older it is possible to teach them to do more of the treatments independently.

- **Antibiotics** - these may be given by mouth (orally) or by drip (intravenously). Oral antibiotics may be given daily or intermittently depending on your child’s condition. Intravenous antibiotics are used on an intermittent basis as required. Occasionally children are started on inhaled antibiotics.

- **Sputum/cough swabs** will be taken regularly to detect any infection in your child’s lungs.

- Your child may suffer with **hearing** problems if the cilia in the ears are not sufficiently effective at clearing fluid and secretions. They may need grommets or other aids, especially in early childhood. Grommets are tiny pipes put in the eardrum to help drain any fluid out, and let air into the middle ear (improving the hearing immediately). Generally, hearing tends to improve through childhood and although most adults have a degree of hearing loss, it is not normally severe enough to require any treatment. Your child will be seen regularly by an ENT surgeon to monitor their condition.

**Long term**

If treatments are adhered to, life expectancy should be normal. As yet there is no cure.

Cilia are also found in the reproductive organs, and fertility is often reduced in patients with PCD. Advice about fertility will be offered at an appropriate time.

**Support groups**

There is no specific support group for Kartagener’s syndrome. As all patients have primary ciliary dyskinesia we recommend this support group: [www.p-c-d.org.uk](http://www.p-c-d.org.uk)
We are now a smoke-free site: smoking will not be allowed anywhere on the hospital site. For advice and support in quitting, contact your GP or the free NHS stop smoking helpline on 0800 169 0 169.

Other formats:

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Please note: We do not currently hold many leaflets in other languages; written translation requests are funded and agreed by the department who has authored the leaflet.

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