Children's Interstitial Lung Disease (chILD)

Lung Foundation Australia

when you can't breathe... nothing else matters®

Overview

Children's interstitial lung disease (chILD) is a broad name for a group of rare lung diseases that can affect infants, children and adolescents. It may also be called chILD syndrome or children's diffuse lung disease. "Interstitial" refers to the tissue and space around the air sacs of the lung where oxygen is absorbed into the blood. In chILD this absorption of oxygen is impaired. This results in breathing difficulty and decreases the oxygen supply to other organs in the body.

Diseases included in chILD are:

- Surfactant protein metabolism disorders
- Alveolar capillary dysplasia and other developmental lung disorders
- Pulmonary interstitial glycogenosis (PIG)
- Hypersensitivity pneumonitis
- Neuroendocrine cell hyperplasia of infancy (NEHI)
- Diseases associated with immunological problems.

Symptoms

As there are many types of chILD, the symptoms and severity vary widely. Symptoms can appear at any age during infancy or childhood, and can continue or worsen over a long period of time. Symptoms can be triggered by chest infections (colds and flu), exposure to air pollutants (e.g. dust or smoke) and/or an increase in activity or exercise.

Common symptoms include:

- Fast breathing (tachypnoea)
- Recurrent cough
- Shortness of breath (breathlessness) or wheezing.

Other symptoms may include:

- Poor growth or failure to gain weight at a rate expected for a child's age
- Loss of appetite or feeding issues
- Recurrent chest infections (exacerbations)
- Abnormal chest sounds (wheezing/crackles) heard via a stethoscope
- Using muscles between and under the ribs or in the neck when breathing (retractions/indrawing)
- Abnormal enlargement of the tips of the fingers or toes (clubbing).

Causes

It is often difficult to determine the exact cause of chILD, however some conditions and factors that may cause or lead to chILD include:

- Inherited or genetic conditions
- Impaired development of the lungs
- Immune disorders (auto-immune or auto-inflammatory - where parts of the immune system can mistakenly attack the lungs)
- Exposure or sensitivity to substances in the environment that can irritate the lungs.

Diagnosis

Diagnosing chILD is often difficult because these diseases are rare, and have symptoms that are similar to many other diseases. Before being able to diagnose chILD, doctors may need to rule out other possible conditions. In addition to examining the medical history of a child and their family, diagnostic tests for chILD can include:

- **Chest X-ray**
- **CT scan:** looks at the tissue in the lungs
- **Lung function tests:** breathing tests – this requires a child old enough to cooperate
- **Blood tests:** may also include genetic testing and tests for immune function
- **Lung biopsies:** a small sample of lung tissue that is removed for examination
- **Bronchoscopy:** examination of the lungs by means of a flexible telescope passed via the trachea (windpipe). This may also involve a lavage (squirting fluid into a small part of the lung which is then collected for examination).
- **Sweat test:** to exclude cystic fibrosis
- **Tests to exclude gastro-oesophageal reflux and aspiration:** the inhalation of substances into the lungs such as food, liquid or vomit
- **Overnight sleep test:** to monitor breathing and oxygen in the blood.

Experience

It's important to know each child experiences chILD differently. Some cases are mild with symptoms that slowly improve over time without treatment. In other cases, symptoms can remain severe and may lead to other complications and more complex treatments. Some severe cases may result in death.

In many cases it is difficult to predict the long-term outcomes for children with chILD. More research is needed to learn more about this group of rare and often serious, childhood diseases.

Treatment

Treatment depends upon the specific type of chILD and the length and severity of symptoms. There is currently no cure for chILD, however treatments can be used to lessen symptoms.

Current treatment options include:

- **Medicines**

  Medicines that suppress inflammation are commonly used to treat chILD. Some examples include: anti-inflammatory medications, glucocorticosteroids, hydroxychloroquine, azithromycin and other immune modifying drugs.

- **Oxygen**

  Depending on how severe the symptoms are, sometimes oxygen therapy may be needed either during activity, sleep, or continuously. Oxygen can be supplied in different ways, but usually this is via cylinders or an oxygen concentrator.

- **Nutrition**

  One of the common issues in chILD is poor growth, particularly in infants and younger children. Growth and lung function are closely related, so some children may need extra calories because it takes more work for them to breathe. It is important to discuss this with your doctor as this may mean they need nutritional support. This may include feeding supplements and/or nasogastric feeds (via a tube inserted through the nostril, down the eosophagus, and into the stomach) or gastrostomy feeds (via a tube directly into the stomach). A dietician is an important member of your child's care team if nutritional support is needed.

- **Transplant surgery**

  In very rare cases, complicated treatments like lung transplantation may need to be considered.

Other management options include:

- **Ensuring vaccinations are up-to-date**

  Vaccination prevents infectious diseases that can have serious health consequences for children with chILD. It is important to discuss a vaccination schedule for your child with your doctor, including both routine and specific vaccines. This discussion should include an annual flu vaccination (in children over six months), as well as vaccinating against pertussis (whooping cough) and medicines which help protect against RSV infections.

- **Avoiding exposure to smoke**

  Exposure to second-hand tobacco smoke is harmful to all children in general, but can be particularly harmful to a child with lung disease, and should be avoided.

- **Avoiding infections**

  Although it is almost impossible to prevent all lung infections, it is a good idea to avoid people who are suffering from colds and flus to reduce this risk. Close family members are not expected to avoid each other, but good handwashing and general hand hygiene, is advised.

FURTHER INFORMATION AND SUPPORT

Contact Lung Foundation Australia for more information and to access our support services. You can also join our mailing list for regular updates and the latest news.

Lung Foundation Australia Services

- Information and support line
- Lung disease information resources
- Education seminars and webinars
- Lung cancer support nurse
- Support groups
- Peer-to-peer connections
- Referral to pulmonary rehabilitation and Lungs in Action exercise programs
- Newsletter

External Links

Australian Genomics
australiangenomics.org.au

Consumer Information
genomicsinfo.org.au

US child foundation
child-foundation.org

Lungfoundation.com.au | **Freecall 1800 654 301** | **enquiries@lungfoundation.com.au**