

# What is Interstitial Lung Disease in Children?

Children's Interstitial and Diffuse Lung Disease (chILD) is a group of rare lung diseases found in infants, children and teens. There are many types of chILD. The different types of ChILD can have some of the same symptoms. The symptoms, however, may vary in how severe they are. Adults can have interstitial lung disease too. Some adults and children have the same kind of interstitial lung disease, but there are some forms of chILD that are unique to children and some forms of adult interstitial lung disease that are not seen in children.



ChILD is a problem in the lungs, either in the airways (breathing tubes), alveoli (air sacs), and/or in the interstitium (tissues that surround the alveoli). ChILD can cause low oxygen levels, abnormal lung function, difficulty breathing, and difficulty keeping up with exercise.

ChILD symptoms start at different ages. Some types affect babies while others affect older children. Some types of chILD run in families.

## What are the types of chILD?

Some types of chILD are genetic and passed through families. Some types are caused by an environmental or infectious trigger. Some have an unknown cause. More and more genetic causes for chILD are discovered as we learn more about genes.

Examples of types of chILD:

- Bronchiolitis Obliterans
- Chronic Bronchiolitis
- Connective tissue associated lung disease
- Cryptogenic Organizing Pneumonia (COP)
- Alveolar Capillary Dysplasia with misalignment of the pulmonary veins
- Hypersensitivity Pneumonitis
- Capillaritis
- Neuroendocrine Hyperplasia of Infancy (NEHI), also known as Persistent Tachypnea of Infancy
- Pulmonary Interstitial Glycogenosis (PIG)
- Surfactant dysfunction mutation

## What are common signs and symptoms with chILD?

Since there are many types of chILD, your child may have any of the following:

- Fast breathing (tachypnea)
- Working hard to breathe
- retractions (use of muscles between the ribs or in the neck when breathing)
- Chronic cough
- Abnormal lung sounds heard with a stethoscope, such as wheezing or crackles
- difficulty breathing when exercising
- Coughing up blood (hemoptysis)
- Frequent episodes of bronchiolitis and/or pneumonia
- Respiratory failure
- Poor weight gain or growth (failure to thrive)
- Clubbing (abnormal enlargement of the tips of the fingers/toes)
- Low oxygen levels (hypoxemia) at rest, with sleep, and/or with exercise
- Abnormal findings seen in the lungs on chest x-ray or chest CT (computerized tomography) scan
- Abnormal lung function tests

There are a number of more common lung diseases that can cause these same symptoms. Tests should be done to make sure the symptoms are not from other conditions like cystic fibrosis, heart defects, asthma, or an immune deficiency.

## How is chILD diagnosed?

Interstitial lung disease can be hard to detect and even harder to diagnose. Because of this, if your child has symptoms of interstitial lung disease, they should be seen by a pediatric lung specialist doctor (pulmonologist). There is no single test to diagnose chILD, since each type of chILD is different. The lung specialist will choose which tests to order based on your child's symptoms. Some types have certain patterns on chest CT or lung function tests. Others, like surfactant protein

dysfunction, need blood tests to look for abnormal genes. Often, chILD can be diagnosed with a lung biopsy. Your child will be given general anesthesia so he or she will be asleep and will not feel anything during the lung biopsy. A surgeon makes a small incision between the ribs and removes a small piece of lung tissue. A pathologist examines the tissue and looks for patterns in the lung tissue that might match with a certain type of chILD.

### How is chILD treated?

Since these are rare diseases, research in chILD treatment has been limited. At present, treatment is based mainly on what the lung specialist thinks is best and what has worked in other cases. Treatment also varies based on the type of chILD and can include medications that treat inflammation.

Many children with chILD will need extra oxygen with activity, during sleep, or all of the time. To learn more, see the ATS Patient Information Series fact sheet "Oxygen Therapy in Children". If your child finds it is very hard to breathe, a mechanical ventilator (breathing machine) may be needed.

Some children need airway clearance treatments, like chest clapping or a percussive vest, to help get rid of mucus.

Some children with chILD are given corticosteroid medicines by tablet, liquid, inhaler, or in the vein (IV). Other medicines that suppress the immune system or treat inflammation, like hydroxychloroquine, may also be tried. Inhaled bronchodilators like albuterol can relax airway muscles if there are spasms in the breathing tubes.

In some children, a lung transplant is the best treatment for severe disease.

It is important that children with chILD get all of their vaccines on time, including influenza and pneumococcal pneumonia vaccines.

Growth is also important to monitor in children with chILD. Because their bodies are working harder to breathe, they may need more calories than healthy children.

Always try to protect the lungs of children with chILD. They should not smoke tobacco, marijuana, or other drugs. Avoid exposure to tobacco smoke, vaping and air pollution. Do not use air fresheners or diffusers with essential oils or humidifiers as these may cause harm to the lungs when inhaled.

### What is the likely to happen to my child with chILD?

How and if the condition progresses varies for each type of chILD. Some cases of chILD are very severe and lead to death shortly after birth. Others are chronic (long lasting) and may stay the same or slowly get worse over years. A few types can get better over time.

**Authors:** Marianna Sockrider, MD, DrPH; Leland Fan, MD; Ann Gettys, Robin Deterding, MD; James Hagood, MD; Geoffrey Kurland, MD; Julie McDougal, RRT, MAE

## Rx Action Steps

If your child has unexplained lung problems or does not respond to treatment for common problems, see a pediatric lung specialist doctor.

If your child has chILD,

- ✓ Take action to prevent illness by washing hands, avoiding sick people, and getting the shots your doctor recommends (along with yearly flu shots).
- ✓ Avoid exposure to air pollution and smoke.
- ✓ Give extra oxygen as needed to reduce stress on the lungs and body.
- ✓ Give all medicines as prescribed and keep regular follow-up appointments.

**Healthcare Provider's Contact Number:**

\_\_\_\_\_

## For More Information

**Children's Interstitial Lung Disease Foundation**  
(A member organization of the ATS Public Advisory Roundtable)

- <http://child-foundation.org>
- <http://child-foundation.org/family-support/living-with-child/>

**National Organization for Rare Disorders**

- <http://www.rarediseases.org/>

**National Institutes of Health Office of Rare Diseases Research**

**Genetic and Rare Diseases Information Center (GARD)**

- <https://rarediseases.info.nih.gov/GARD/>

**American Thoracic Society**

- [www.thoracic.org/patients](http://www.thoracic.org/patients) (patients)
- <https://www.thoracic.org/statements/resources/pldd/cp-guideline-080113.pdf> (clinicians)

This information is a public service of the American Thoracic Society. The content is for educational purposes only. It should not be used as a substitute for the medical advice of one's healthcare provider.

