

What is Interstitial Lung Disease in Children?

Children's Interstitial 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, but the types of disease they have differ from children.



Many of the chILD diseases involve the interstitial tissues of the lung. The word "interstitial" refers to the tissues that surround the air sacs (alveoli) in the lung and airways (breathing tubes). Interstitial diseases can make your child's lung function and oxygen levels get low. Some of the conditions that are called chILD diseases may involve other parts of the lung as well.

ChILD symptoms start at different ages. Some types affect babies while others affect older children. Some children appear very healthy until they develop an illness. For some, there is a history of infants in the family having lung disease.

What types of interstitial lung disease are seen in children?

There are many types of interstitial lung diseases, but few affect children. It is not known how many children have chILD, but they are considered rare conditions. Some types are caused by other diseases, and others are unexpected with no known cause. People can inherit certain types of chILD (like surfactant problems) that are passed on in the family through genes. Surfactant is a fluid made in the lungs that helps keep the air sacs open. Some types of interstitial lung diseases are like adult forms of the disease, while some are not seen in adults at all.

Examples of types of chILD:

- Bronchiolitis Obliterans
- Chronic Bronchiolitis
- Connective tissue associated lung disease
- Cryptogenic Organizing Pneumonia (COP)
- Developmental disorders such as Alveolar Capillary Dysplasia
- Hypersensitivity Pneumonitis
- Lung growth abnormalities

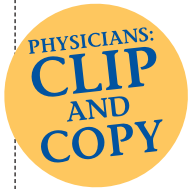
- Neuroendocrine Hyperplasia of Infancy (NEHI), also known as Persistent Tachypnea of Infancy
- Pulmonary Interstitial Glycogenosis (PIG)
- Surfactant dysfunction mutations

What are common signs and symptoms with chILD?

Since there are many types of chILD, your child may have all or some of these signs and symptoms:

- Fast breathing (tachypnea)
- Difficulty moving air in and out with each breath
- Breathing with retractions (use of muscles between the ribs or in the neck when breathing)
- Chronic cough or a cough that comes and goes often
- Abnormal lung sounds heard with a stethoscope, such as wheezing or crackles
- Shortness of breath or 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 or toes
- Low oxygen levels (called hypoxemia at rest, with sleep, or with exercise)
- Abnormal shadows seen in the lungs (usually all through 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



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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 doctor (pulmonologist). There is no single test to diagnose chILD, since each type of chILD is different. The pulmonologist will choose which tests to order based on your child's symptoms. Some types have certain patterns on chest CT ("cat scan") or lung function tests. Others, like surfactant protein deficiencies, need blood tests to look for abnormal genes.

Often, chILD can be diagnosed with a lung biopsy. A lung biopsy requires removal of a small piece of lung tissue for examination in the lab. Your child will be given general anesthesia medication so he or she will be asleep and will not feel anything during the lung biopsy. A surgeon removes a small piece of the lung tissue from one lung through a small incision between the ribs. 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 doctor thinks is best and what has worked in other cases. Treatment also varies based on the type of chILD. There is no cure for any type of chILD at this time. Many children with chILD will need extra oxygen either 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 clear lung mucus.

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

Hypersensitivity pneumonitis is a form of chILD. This condition can be caused by an exposure to certain birds (like pigeons or other birds), or certain molds. If the chILD is from birds or certain molds, removing your child from these exposures will control the chILD. In some children, a lung transplant is the best treatment for severe disease.

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 early death. Others are chronic (long lasting) and may stay the same or slowly get worse over years. A few types

can get better over time. There is no cure for any of the types of chILD at this time.

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Resources:

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

<http://www.childfoundation.us/>

"Get Up and Go with chILD!"—Booklet for families of children with chILD

http://www.childfoundation.us/images/Get_Up_And_Go_With_chILD_-_Jan_2009.pdf

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 Guideline

"Classification, Evaluation, and Management of Childhood Interstitial Lung Disease (chILD) in Infancy."

ChILD Research Cooperative (Am J Respir Crit Care Med Vol 188, Iss. 3, pp 376–394, Aug 1, 2013)

<http://www.thoracic.org/statements/resources/pldd/cp-guideline-080113.pdf>

Rx Action Steps

If your child has unexplained chronic lung problems or does not respond to treatment for common problems, see a pediatric lung 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 tobacco smoke.
- ✓ Give extra oxygen as needed to reduce stress on the lungs and body.
- ✓ Make sure your child exercises (if able) and eats a healthy diet.
- ✓ Give all medicines as prescribed and keep regular follow-up appointments.

Doctor's Office Telephone:
