

# Symptoms of Childhood Interstitial Lung Disease

As there are many types of childhood Interstitial Lung Disease (chILD), the symptoms and severity vary widely. Symptoms can appear at any age during infancy or childhood, and can continue 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.

Symptoms can be triggered by chest infections (colds and flu) and exposure to air pollutants (e.g. dust or environmental tobacco smoke) or allergens. Symptoms may be more marked with an increase in activity or exercise.

It is important to remember every child experiences chILD differently and symptoms and treatments will vary. Early diagnosis and treatment are important, but it is often very difficult to make a diagnosis. If your child is on a path to diagnosis it is important to be referred to a paediatric respiratory specialist with expertise in chILD.

A number of other conditions may have similar symptoms to chILD, such as asthma, primary ciliary dyskinesia, gastro-oesophageal reflux and cystic fibrosis. Your doctor will need to rule out other conditions with similar symptoms as part of the steps in diagnosis.

## Other symptoms may include:

- Reduced exercise tolerance (e.g. in infants this may manifest as difficulties feeding but in older children, frequent pauses with walking or difficulties climbing stairs)
- Loss of weight, failure to gain weight, poor growth (failure to thrive)
- Loss of appetite
- When breathing, sucking in of the skin in between the ribs (rib retractions) and at the base of the throat (tracheal tug)
- Blue tinged lips because of low oxygen levels in the blood (cyanosis)
- Abnormal enlargement of the tips of the fingers or toes (clubbed fingers/toes)
- Recurrent chest infections
- Exacerbations of the underlying condition (as described in Neuroendocrine Cell Hyperplasia of Infancy (NEHI)).

## Common signs and symptoms in children with chILD<sup>1</sup>



Fast breathing (tachypnoea)



Shortness of breath, including difficult and laboured breathing (work of breathing)



Persistent cough



Rattles, crackles or wheeze in chest heard via a stethoscope.

## Severity

The severity of the illness can vary significantly, even in children with the same diagnosis. Some seem barely affected beyond having only a slightly faster breathing rate. Many children require temporary or long-term oxygen therapy during some stage of their disease progress. For cases with severe breathing difficulties, children may require more active assistance with non-invasive ventilation. In the very rare and most severe cases, lung transplantation may be a treatment option.

A severity classification system, from mild through to severe, for children with chILD has been proposed by the American paediatric lung specialist Dr Leland Fan.<sup>1</sup>

## Severity Classification of chILD

### Mildest Form

No Symptoms (asymptomatic)

Symptoms but normal blood oxygen saturation levels

Symptoms with decreased blood oxygen levels

Symptoms with decreased blood oxygen levels at rest

Presence of Pulmonary Arterial Hypertension\*

### Severest Form

\*Pulmonary Arterial Hypertension (PAH) is a type of high blood pressure in the lungs. It occurs when the blood vessels in the lungs become thicker or stiffer. The heart has to work harder to push blood through, increasing the blood pressure in the lungs.

## Signs and symptoms to watch for

It is important to observe your child and their symptoms so you can recognise if your child's condition is deteriorating. Ask your child's healthcare team about warning signs of worsening lung disease and when to seek emergency medical care.

Agree on a plan of action if these warning signs occur including a first response for any specific deterioration. Talk with your child, when possible, and encourage them to identify these signs and ensure they feel empowered to tell you. Trust in yourself and your instincts. When you are uncertain or worried about changes, follow your written care plan, consult your paediatric respiratory specialist or call Triple Zero (000). It can be very stressful during emergency situations and difficult to think clearly, so having a clear prepared emergency plan is invaluable.

Potential signs or symptoms that can help indicate that your child has become unwell include:

- Blue discolouration of the lips or tongue
- Not able to talk in full sentences without taking a breath in between words
- Restless, agitated
- Listless, apathetic Lethargic, drowsy or fatigued
- Increased sweating
- Pale skin
- Fever
- Vomiting
- Tummy ache
- Loss of interest in eating or drinking
- New or worsening cough

Children with chILD will also get normal childhood problems and some of these may also be signs and symptoms of illnesses not related to your child's lung condition. If your child is not their usual self and you are concerned, don't hesitate to seek medical advice and in an emergency call Triple Zero (000).

There are several warning signs or symptoms which should prompt a visit to your child's doctor:<sup>1</sup>

### Respiratory rate

The rate of breathing, or respiratory rate, is a measure of the number of breaths per minute. In children with chILD, it is a simple but effective means to assess their condition. Increased respiratory rate often indicates deterioration. But to recognise this, you first need to know your child's normal breathing rate. The best way to do this is to observe and measure your child's baseline breathing rate when they are relatively well. One method of doing this in children is to lay your hand onto your child's belly, and then count how often it is raised in 60 seconds (young children are "tummy breathers", meaning that they lift their belly as they breathe in). As physical exercise increases this rate and deep sleep reduces it, the optimal time to do this is when your child is awake and well rested (i.e. have not exhausted themselves just prior to measuring it).

## Shortness of breath

Beyond an increase in respiratory rate, worsening shortness of breath is another indicator of deterioration. In infants or toddlers you must look for the typical sign of “indrawing”. This is where there is a sucking in of the skin between the ribs (retractions) or base of the throat (tracheal tug) when breathing in. Their breathing may become more audible. Their posture also may change to find a position where they feel it is easier to breathe. Young children from three to four years of age may be able to report being short of breath themselves.

## Drop in oxygen saturation

If you have an oxygen saturation monitor at home, your healthcare team will explain that oxygen levels fluctuate normally throughout the day in a healthy person (lower during the night than during the day). Likewise, a person’s posture can affect saturations. The effectiveness of the pleth read is also important to consider and can be affected by movement or other artefacts, leading to incorrect saturation reading.

Some children may have lower saturations when they lie on their belly and others when they lie on their side. Similar to respiratory rate, it is important to compare saturations to a known “normal” baseline range for your child. Ask your doctor what level they are happy with and what level indicates that you need to seek medical help.

## References

1. chILD-EU, 2015, Living with chILD: An Information Booklet for Parents and Relatives of affected Children, Available at: [www.klinikum.uni-muenchen.de/Child-EU/download/en/child-register/services/booklet/Living-with-chILD-\\_UK-Version\\_-FINAL.pdf](http://www.klinikum.uni-muenchen.de/Child-EU/download/en/child-register/services/booklet/Living-with-chILD-_UK-Version_-FINAL.pdf).



## FURTHER INFORMATION AND SUPPORT

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

### Lung Foundation Australia Services

- Information and Support Team
- Lung disease information resources
- Education webinars
- Lung Cancer and Respiratory Support Nurses
- Support groups
- Peer-to-peer connections
- Referral to pulmonary rehabilitation and Lungs in Action exercise programs
- E-newsletter

### External Links

- Australian Genomics  
[www.australiangenomics.org.au](http://www.australiangenomics.org.au)
- Consumer Information  
[www.genomicsinfo.org.au](http://www.genomicsinfo.org.au)
- US Child Foundation  
[www.child-foundation.org](http://www.child-foundation.org)

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