Treatment of Childhood Interstitial Lung Disease

There is currently no cure for childhood Interstitial Lung Disease (cHILD), however treatments can lessen symptoms and improve quality of life.

There is currently no cure for chILD, although some children may have certain types of chILD which may slowly improve over time. Early diagnosis and treatment can improve symptoms and lead to better long-term management and health outcomes, including quality of life (how your child feels). In Australia, the chILDRANZ network has enabled training of junior health professionals and sharing of knowledge to improve knowledge about chILD; it has directly impacted positively on individual patient outcomes.

The treatment of chILD is complex and usually involves a variety of approaches.

Standardised treatment guidelines do not currently exist due to the rarity of these diseases, which emphasises the importance of being seen by a paediatric respiratory specialist with experience in managing chILD. Your healthcare team will develop a treatment and management plan specific to the needs of your child. Lifestyle and behavioural changes are also important in treating chILD and keeping well.

Treatment options

A number of treatment options may be used to treat chILD. Furthermore, treating other medical conditions that can contribute to symptoms such as sleep apnoea, gastro-oesophageal reflux, aspiration or heart conditions can reduce the severity of symptoms, particularly breathlessness when active.

Current treatment options include oxygen therapy, exercise, nutrition and medication. Lung transplantation may be a last resort for some.

Oxygen therapy

Providing supplementary oxygen is the most important treatment for all children experiencing low blood oxygen levels (hypoxia).

Oxygen therapy is commonly prescribed with the aim of reducing breathlessness and increasing energy levels by increasing the amount of oxygen passing through the lungs into the blood. Sometimes oxygen therapy may be needed either during activity, sleep, or continuously, depending on the severity of the symptoms. Oxygen therapy may also be used only during periods of acute illness (e.g. when your child is unwell with a respiratory infection).
Oxygen can be supplied in different ways, but usually this is via cylinders or an oxygen concentrator.

Your paediatric respiratory specialist will provide you with the prescription for oxygen. They may refer you to an oxygen specialist nurse who will liaise between yourself and one of the many oxygen supplier companies. You will receive detailed education and training about the safe use of this equipment to enable you to look after your child at home. Learning to use equipment outside of the hospital setting can feel overwhelming initially; it is okay to ask for additional practice and training before you leave the hospital.

If your child is on oxygen therapy, it may be necessary to monitor blood oxygen levels to ensure satisfactory oxygen saturation. Part of the supplied equipment may include a pulse oximeter. Your paediatric respiratory specialist will discuss with you whether you require a monitor at home.

How is the oxygen delivered?

A length of lightweight silicone tubing is connected from the oxygen supply to the patient. The oxygen can then be inhaled through either the nose (nasal cannulas) or the nose and mouth simultaneously (a mask). The longer lengths of tubing provide greater freedom and range of movement for children not able to move the oxygen source easily themselves.

Sometimes, oxygen therapy alone is not sufficient to maintain adequate oxygen saturations and additionally a child may not be able to eliminate enough carbon dioxide from their blood.

In these cases, the child will require assistance with their breathing in the form of ventilatory support (mechanical ventilation). Ventilatory support can be roughly divided into two groups:

- Non-Invasive Ventilation (NIV) – a specialised breathing support provided to patients with severe breathing difficulties via either a specifically designed over the nose and mouth mask or a nasal (nose) mask. These masks are connected to a breathing machine (ventilator) with flexible tubing. The ventilator helps to open the airways and support the breathing muscles, making it easier for the child to breathe.
- Invasive Ventilation – this breathing support is achieved by inserting a breathing tube directly into the wind-pipe (trachea), either via a cut into the throat (tracheotomy) or passing it down through the mouth or nose (intubation). Intubation is primarily used for ventilation in the setting of acute emergencies, when non-invasive ventilation has proved inadequate in correcting low blood oxygen and/or high carbon dioxide levels. Most children requiring ventilation via intubation need to be sedated. Much higher pressures can be achieved with invasive ventilation as compared to non-invasive. When it becomes apparent that a child may require prolonged or long-term ventilation, a tracheotomy, may be the preferred method.

Most children only require mechanical ventilation temporarily or intermittently (e.g. overnight or during acute deteriorations). A few children may require continuous ventilation. Although continuous ventilation does not completely preclude normal daily life, the care of children dependent on ventilation is very demanding for all involved.

Exercise

The diagnosis of chILD does not equate to an end to sport or exercise for the majority, although in extreme cases it may. Exercise is not dangerous for children with chILD; rather it should be encouraged and is an important component of their treatment. Children are excellent at self-regulating themselves with exercise. They stop when they have reached their limit. You do not have to be worried about them over exerting themselves and exacerbating their disease process. Likewise, it is important that adults do not push children past their limits or into doing activities that they do not wish to. Discuss with your paediatric respiratory specialist about the level of participation in physical activity that would be right for your child.

Pulmonary Rehabilitation

Pulmonary rehabilitation is an exercise and education program that helps improve the wellbeing of people who have chronic (ongoing) breathing problems. The program is the most effective evidence-based intervention to manage breathlessness in adults with chronic lung conditions. It improves exercise capacity, quality of life, and teaches people the skills to manage their condition and stay well. Currently, there are limited pulmonary rehabilitation programs designed for children. Similar activity programs can be developed to meet the needs of individual children with a paediatric respiratory specialist. This an area that needs more research in children with chILD as it is not offered in many places.

Feeding and nutrition

One of the common issues in chILD is poor growth, particularly in infants and younger children. Growth and lung function are closely related. Some children may need extra calories because it takes more work for them to breathe. Often this means that they have to eat more than healthy children.

All children with chILD should have regular measurements of weight and length and the results plotted on a growth chart that is regularly reviewed. Inadequate weight gain often indicates insufficient caloric intake for the child’s needs and requires the assistance of a dietitian. A dietitian will help to evaluate your child’s dietary needs and will help determine their nutritional requirements. If your child is not able to eat enough food to compensate for any nutritional deficiencies, then they may need nutritional support. This may include supplemental high caloric drinks or feeding supplements administered using feeding tubes/buttons.

Any child with a significantly weakened immune system has an increased risk of infection. Importantly, this risk includes infection being transmitted by food. You can reduce the risk of your child being exposed to infections from food by following these recommendations:

- Meat, processed meat (e.g. sausages, etc), seafood and dairy products should be stored in the fridge
- Raw meat must be cooked completely through and either eaten on the day of purchase or promptly cooled for later consumption
- Only use fresh eggs and cook them completely (i.e. egg white and yolk must not be ‘runny’ or ‘soft’)
- Do not leave warm food out; eat it freshly cooked or store quickly in the fridge after cooking. Leftovers in the fridge need to be heated completely through before consuming.
- Follow the expiry dates on food products
- Carefully adhere to personal hygiene in the kitchen[1].

Medication

No medication is currently capable of curing any type of chILD. Rather, the medications that are used may lessen symptoms, alter the progression of the disease and improve quality of life.

Severe inflammation often plays a significant role in many types of chILD; hence medications that suppress inflammation are commonly used to treat chILD. Some examples include: anti-inflammatory medications, glucocorticosteroids, hydroxychloroquine, azithromycin and other immune modifying drugs.

Medications for treating chILD often have significant side effects. Therefore, before starting any medications, the doctor will, for each individual case, weigh up the potential benefits of the medication versus the burden of its potential side effects. Discuss with your paediatric respiratory specialist the purpose of each medication prescribed for your child, what the common side effects are and how to manage them.

Medications are not used to treat every case. For some medical conditions, medications prove to be ineffective or are unnecessary. This is especially true in conditions secondary to structural changes or developmental disorders of the lung. Furthermore, children with NEHI typically respond well to oxygen therapy but not to any of the above listed medications.

Your paediatric respiratory specialist will closely evaluate each medical therapy that may be prescribed for your child to determine whether it is required or not, and to ensure the benefits outweigh the burden of any side effects.

Due to the rarity of these diseases, the majority of them do not have any clinical studies about the safe or effective use of the medications in their treatment. Most of the medications are well known and proven treatments in other diseases. Currently, it is not clear whether they are truly beneficial, or under which circumstances they are most beneficial in treating chILD.

Lung transplantation

In very rare, severe cases, a child will progressively deteriorate despite medical therapy. Subsequently, lung transplantation is considered when: exercise tolerance is significantly reduced in everyday life; daily activities are only possible under significant strain (or not at all); and the progressive deterioration of the lung function leads to a concern for survival.

Lung transplantation is major surgery and comes with a substantial risk for major complications and is not suitable for everyone. Therefore, transplantation is only ever considered after all other therapeutic options have been exhausted, and the potential benefits of the surgery outweigh the risks. After transplant there is a need for children to have lifelong immunosuppressive drugs and regular monitoring such as repeat bronchoscopies. The five-year survival following lung transplantation is reported to be around 55% and is improving all the time with better pre and post-transplant management and surgical techniques2. Other non-medical considerations contribute to the likelihood of success of lung transplantation, including the ability to follow the very strict post transplantation treatments.

Treatment Outcomes

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.

The treating team

Throughout your journey, you will meet different healthcare professionals. They can help answer your questions and concerns.

- The **paediatric respiratory specialist** (specialist doctor for children's lung diseases) is responsible for establishing the diagnosis and treatment plan for your child. They will often coordinate your child's healthcare, including the involvement of other types of healthcare professionals.
- Your **paediatrician** and **General Practitioner (GP)** may remain involved in your child's care, providing support in response to your child's medical needs.
- **Nurses** play an important role in the treatment of the children as inpatients as well as outpatients and in the community. They have the closest contact to them and are therefore, in addition to their medical care, an important person of reference.
- The **paediatric cardiologist** (heart specialist for children) will perform echocardiograms at regular intervals to exclude the presence of Pulmonary Arterial Hypertension.
- The **psychologist** is a very important member in the team, supporting the child and family in dealing with (and overcoming) the stresses of the underlying condition. They develop a special trust relationship with the family.
- Situationally required specialists include **rheumatologists, immunologists, intensive care physicians, gastroenterologists** and **genetic counsellors.**
- The **dietitian** provides advice regarding optimal nutrition. Assessing the caloric intake and correcting as required.

- **Physiotherapists** are very important in the treatment of chILD. They teach special breathing techniques to help mobilise lower airway secretions if present (autogenic drainage), help improve physical conditioning and fitness (i.e. exercise tolerance).
- **Speech therapists** support and assist children that require tracheal cannula with alternate speaking techniques.
- **Social workers** work with children and their families to help them adjust to, and cope with the challenges surrounding their child's health condition.
- The **paediatric surgeon** will become involved if a diagnostic lung biopsy is being considered.

Play therapists help children to express themselves through creativity. Some hospitals may also have access to **hospital teachers, sports therapists, and hospital clowns.** These team members help to improve physical health and wellbeing, while also helping them to cope better with their disease, provide education and break up the tedium of their hospital stay[1].

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.

2. Benden, C. 2017, Pediatric lung transplantation. *Journal of Thoracic Disease*, 9(8): 2675-2683. Available at: http://dx.doi.org/10.21037/jtd.2017.07.84.

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
- Consumer Information
  www.genomicsinfo.org.au
- US Child Foundation
  www.child-foundation.org

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