Pediatric Pulmonology Conditions Evaluated and Treated

As a parent, watching a child suffer from a respiratory disorder can be frightening and worrisome. Our respiratory specialists provide compassionate care for a wide-range of pediatric respiratory disorders. We also oversee one of the largest cystic fibrosis programs in the country.

Pulmonary care is caring for and treating the lungs and respiratory (or breathing) system. This can include many kinds of lung conditions, such as asthma, bronchitis, chronic obstructive pulmonary disease, pneumonia, cystic fibrosis, and others. The University of Utah Health Care Pulmonary Services also provides tests and diagnostics for our clinics and hospitals with blood gas analyses and services for athletes and patients with performance evaluations that test aerobic capacity. For the best lung and pulmonary care, visit our fellowship-trained specialists and find high quality, individualized treatment whatever your condition or need.

We Provide Treatment for:

- **Asthma and difficult to treat asthma**: Asthma is a long-term (chronic) lung disease that causes your child's airways to become sensitive to certain things (triggers). Several things happen to the airways when a child is exposed to triggers:
  - The lining around the airways swell.
  - The muscles around the airways tighten.
  - The airways make more thick mucus than normal.

  All of these things will cause the airways to narrow. This makes it difficult for air to go in and out of your child’s lungs and causes the symptoms of asthma.

- **Cystic Fibrosis**: Cystic fibrosis is an inherited chronic disease that affects the lungs and digestive system of about 30,000 children and adults in the United States (70,000 worldwide).

- **Congenital lung problems**: Congenital lung disorders, also known as cystic lung disease or congenital lung malformations, occur while a baby is still in its mother's womb. Most congenital lung disorders are discovered during prenatal ultrasounds. About 10 percent of congenital lung disorders are diagnosed at birth, while another 14 percent show up by age 15.

- **Types of congenital lung disorders**:

  **There are four general types of congenital lung disorders**:

  1. **Bronchogenic cysts**: usually develop on the esophagus or trachea but can also sometimes be found on the lower lobes of the lung. They can compromise airways if they become infected or grow too large.
2. **Congenital cystic adenomatoid malformations (CCAMs):** form when a baby's lung tissue grows more than normal. Babies may have just a single CCAM or several. They can cause cysts that fill with fluid or solid masses in the lungs. CCAMs can also prevent the alveoli (air sacs in the lungs) from developing normally. CCAMs usually form in one lobe of the lung. (CCAMs are sometimes referred to as congenital pulmonary airway malformations or CPAMs.)

3. **Lobar emphysema:** is a rare, serious condition that can interfere with an infant’s airways by trapping airflow during breathing. It can result in over inflation of the lobes of the lung.

4. **Pulmonary sequestrations:** are solid masses of lung tissue. Pulmonary sequestrations don't connect to any of the lungs' airways or blood supply. They can be inside of the lungs (intralobular) or outside (extralobular) in either the chest or stomach.

- **Bronchopulmonary dysplasia:** A chronic lung disease in infants who received mechanical respiratory support with high oxygenation in the neonatal period.

- **Recurrent pneumonia:** more than two episodes of pneumonia in 18 months

- **Persistent pneumonia:** symptoms that do not clear within 14 days and radiograph that do not revert to normal within 4-6 weeks

- **Aspiration lung injury:** Aspiration lung injury is caused by a failure of the normal protective mechanisms that prevent aspiration of gastric contents. Prevention measures are focused on preserving these protective mechanisms. There are three components to the lung injury. The first component is mechanical obstruction to airways from particles in the aspirate. Treatment is aggressive pulmonary toilet to restore airway patency. The second component, seen beginning in the first several hours, is a chemical injury to the airways, leading initially to bronchorrhea, airway constriction, and edema. Later, there is a marked increase in the risk of bacteria due to altered lung defenses. Treatment is supportive care. The third component is lung injury due to the inflammatory response. This process can evolve to an adult respiratory distress syndrome pattern. With modulation of lung inflammation (currently being developed), morbidity and mortality rates will decrease.

- **Noisy breathing:** Stridor is noisy breathing that occurs due to obstructed air flow through a narrowed airway. Stridor is not in and of itself a diagnosis, but rather is a symptom or sign that points to a specific airway disorder.

The timing and the sound of your child's noisy breathing provides clues to the type of airway disorder:

• Inspiratory stridor occurs when your child breathes in and it indicates a collapse of tissue above the vocal cords.
• Expiratory stridor occurs when your child breathes out and it indicates a problem further down the windpipe.
• Biphasic stridor occurs when your child breathes in and out, and it indicates a narrowing of the subglottis, the cartilage right below the vocal cord.

- **Respiratory complications of neuromuscular disease:** Neuromuscular disease compromises both the gas exchange and pump functions of the respiratory system. This can have profound implications for both growth and development of the respiratory system, as well as morbidity and mortality. Aspiration lung disease is common, and leads to increasingly restrictive pulmonary physiology over time. Abnormal lung and chest wall mechanics, and weak respiratory muscles, can combine to cause respiratory failure. Improving the balance between the work of breathing (by decreasing the respiratory load) and the respiratory pump (by improving respiratory muscle strength and decreasing respiratory muscle fatigue) can help prevent the onset of respiratory failure. Airway clearance techniques and non-invasive ventilation are two important tools in this effort. Better ways of assessing the respiratory pump, mechanical function, control and fatigue are needed especially in children.

- **Chronic ventilatory support:** To Achieve and maintain adequate pulmonary gas exchange, minimize the risk of lung injury, reduce patient work of breathing, optimize patient comfort, normalize blood gases and provide comfortable breathing.

- **Interstitial lung disease:** Interstitial lung disease, or ILD, includes more than 100 chronic lung disorders. These diseases are not cancer and are not caused by an infection.

  Interstitial lung diseases are named after the tissue between the air sacs of the lungs called the interstitium. In these diseases the interstitium is affected by scarring (fibrosis).

  The symptoms and course of these diseases may vary from person to person, but the common link between the many forms of ILD is that they all begin with an inflammation.

  • **Bronchiolitis:** This is inflammation of the bronchioles (the small airways).
  • **Alveolitis:** This is inflammation of the alveoli (the air sacs where oxygen and carbon dioxide exchange in the blood takes places).
  • **Vasculitis:** This is inflammation that involves the small blood vessels (capillaries).

  Most ILDs are diagnosed as:
  • A drug-induced disease
  • **Hypersensitivity pneumonitis:** This is an allergic response to breathing certain kinds of dust.
Other, less common types of ILD are:
• **Sarcoidosis**: in which you have small areas of inflammation in your lungs
• **Idiopathic pulmonary fibrosis**: This is scarring of your lungs that has no known cause.
• **Bronchiolitis obliterans organizing pneumonia**: This is a rare form of interstitial pneumonia.
• **Histiocytosis X**: which is an abnormal increase in the number of immune cells called histiocytes.
• **Chronic eosinophilic pneumonia**: This is a buildup of blood cells called eosinophils in your lungs.
• **Collagen vascular disease**: This is a connective tissue disease in which your immune system attacks your body's own tissues. This is an example of an autoimmune disease.
• **Granulomatous vasculitis**: which is inflammation and death of lung tissue
• **Goodpasture's syndrome**: in which immune cells attack lung tissue and cause bleeding in your lungs
• **Pulmonary alveolar proteinosis**: This means that a large amount of protein has built up in the alveoli.

**In ILD, the lung is affected in 3 ways:**
1. Lung tissue is damaged in some known or unknown way.
2. The walls of the air sacs in the lungs become inflamed.
3. Scarring (fibrosis) starts in the interstitium.

Fibrosis leads to permanent loss of your lung tissue's ability to carry oxygen. The air sacs, as well as the lung tissue around the air sacs and the lung capillaries, are destroyed by the formation of scar tissue.

The diseases may run a gradual course or a rapid course. People with ILD may notice variation in symptoms, from very mild to moderate to very severe. The condition may stay the same for a long time or it may change quickly. The course of ILDs is unpredictable. If they progress, the lung tissue thickens and becomes stiff. Breathing becomes more difficult.

- **Sleep disorders**: Sleep disturbances or problems are common during infancy and childhood, including having trouble getting your child to bed, nightmares, and night terrors.

- **Sleep Apnea**: Sleep apnea is a disorder in which a child's breathing is obstructed and interrupted for periods of 5 to 10 seconds or more while she or he is asleep. These interruptions may occur hundreds of times a night, causing your child to gasp for air and disrupting sleep. The condition occurs in an estimated 1-3 percent of children. Children between the ages of two to six years old are most commonly affected, though it can be seen in infants and adolescents.

When a child's breathing is obstructed during sleep, this can affect the body in many ways. For instance, the heart rate slows, the nervous system is stimulated, blood pressure rises, the brain is aroused and sleep is disrupted. Repeated interruptions to sleep and sleep apnea can cause your child to be sleep deprived, cranky and ill behaved.

The leading cause of sleep apnea in children is enlarged tonsils and adenoids. However, some children with enlarged tonsils will not have any symptoms, whereas others with small tonsils will have severe sleep apnea. Children with other disorders, such as obesity, craniofacial anomalies (related to facial structure or appearance), cerebral palsy, muscular dystrophy and Down syndrome are at an increased risk for sleep apnea.

**Specialty Centers & Clinics at Eccles Outpatient Specialty**

- Pediatric Pulmonary and Sleep Medicine
  - General Pulmonary Clinics
  - Sleep Clinics
  - Sleep/Behavior Clinics
  - Trach/Vent Clinics
- Intermountain Cystic Fibrosis Center
  - Cystic Fibrosis Clinics
  - Cystic Fibrosis/Gastrointestinal Clinics