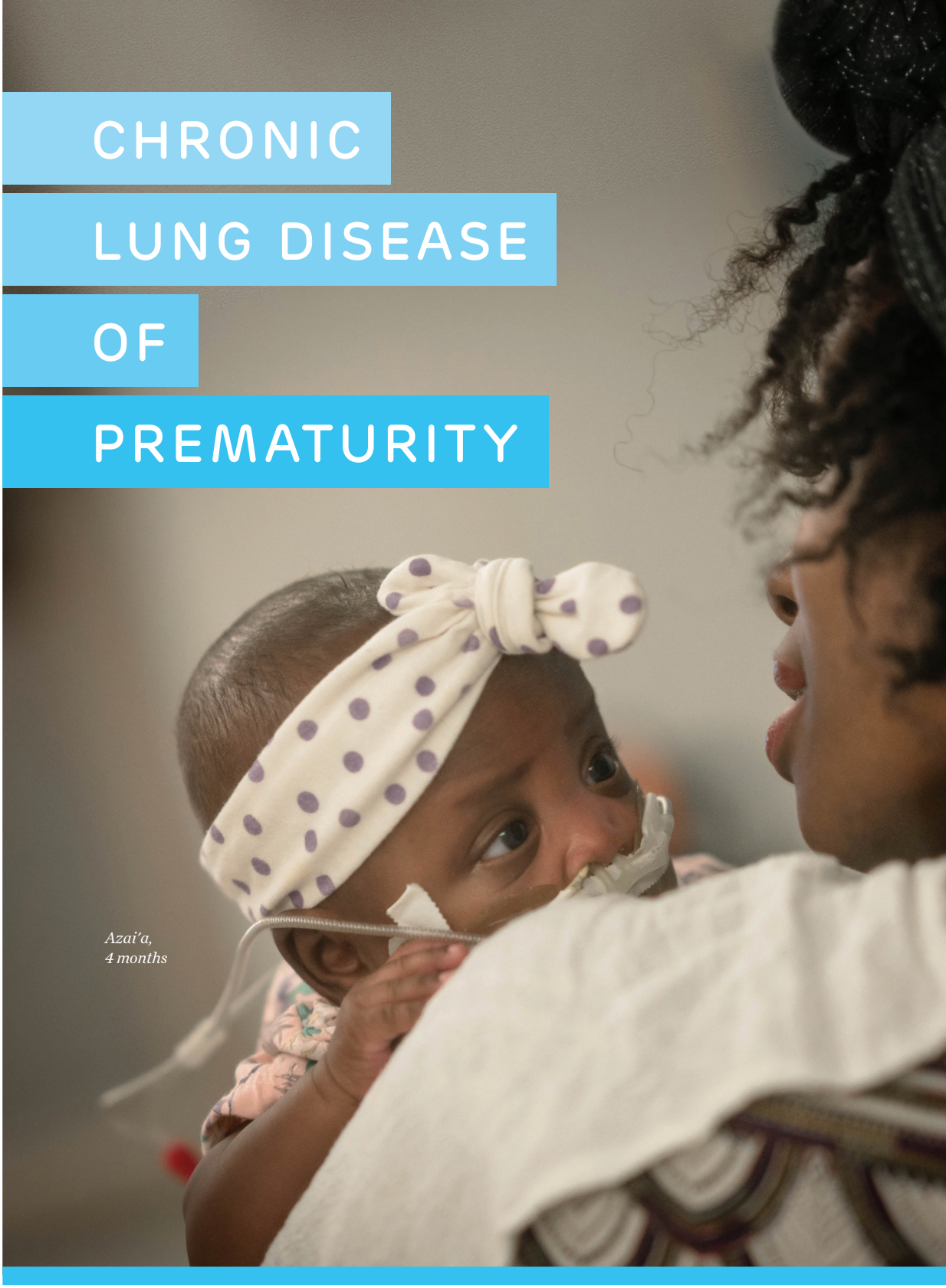
CHRONIC LUNG DISEASE OF PREMATURITY



Azai'a,
4 months

Children's Hospital of Philadelphia®
Division of Pulmonary & Sleep Medicine

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The Division of Pulmonary and Sleep Medicine at Children's Hospital of Philadelphia created the Post-preemie Lung Disease Clinic specifically to care for children with chronic lung disease (CLD) in the critical years after they leave the Neonatal Intensive Care Unit through childhood. The clinic's goal is to keep these children on a trajectory that will maximize their lung function and health while minimizing other illnesses that often occur in children with CLD.

WHAT IS CHRONIC LUNG DISEASE?

Many babies born prematurely will develop chronic lung disease (CLD) while in the neonatal intensive care unit (NICU). The chronic lung disease that preterm infants can develop is called bronchopulmonary dysplasia (BPD). While premature infants with a very low birth weight are the most likely to develop CLD or BPD because their lungs are underdeveloped, even babies born a few weeks early can develop chronic respiratory symptoms. Often, very low birth weight babies with BPD will need to go home on oxygen and medicines until their lungs recover. Since most lung growth occurs during the first two years of life, it is important to address lung development early — before irreparable damage is done — to allow the lungs to grow and heal.

Lily and Ava,
8 months

WHAT CAUSES CHRONIC LUNG DISEASE?

Premature babies have underdeveloped lungs compared to full-term infants. The lungs of premature babies are fragile and easily damaged or inflamed. If your baby's lungs are injured, they may have difficulty breathing and need oxygen. This can affect other areas of your baby's health. It is not clear why some preterm babies develop lung disease and others do not, but some causes are:

- Prematurity: The earlier a baby is born, the more underdeveloped the lungs.
- Low amounts of surfactant: This is a substance in the lungs that helps keep the lungs open.
- High levels of oxygen support in the NICU: This is needed to support growth and survival, but can cause injury to lung cells in preterm infants.
- Ventilator: This is often needed, but the pressure of air delivered by the ventilator can injure the airways.
- Difficulty swallowing or gastroesophageal reflux: Since lungs are underdeveloped, even babies who don't require ventilators can develop chronic lung disease due to formula or stomach acid getting into their lungs.
- Poor weight gain or heart disease: This can interfere with lung growth.

HOW WILL YOU KNOW IF YOUR CHILD HAS CHRONIC LUNG DISEASE?

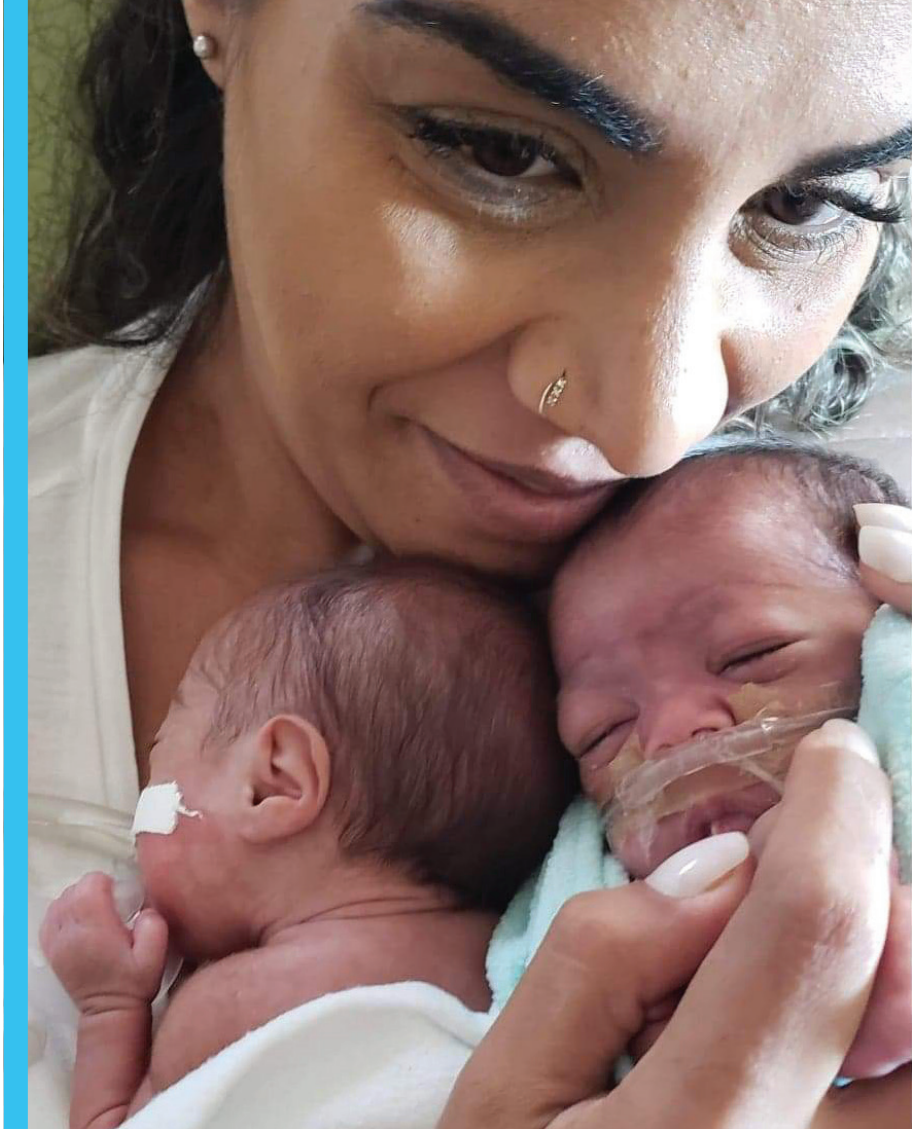
We start to consider chronic lung disease when a baby has breathing problems after birth has abnormal chest X-rays, or the baby needs respiratory medicines, oxygen or a ventilator (breathing machine) to help them breathe normally.

Risk Factors

Besides some of the causes listed above, some other risk factors for chronic lung disease include:

- Birth weight less than 1,500 grams (3 pounds, 5 ounces)
- Patent ductus arteriosus (PDA) – a heart defect where a connection between the blood vessels in the heart and lungs does not close after birth
- Maternal womb infection (chorioamnionitis)
- Development of any infection during or shortly after birth
- A family history of asthma

Barbara (left) and Stefania (right) with their mother Katerina.



WHAT ARE SYMPTOMS OF CHRONIC LUNG DISEASE?

- Breathing faster than normal
- Difficulty breathing
- Retractions (skin sinking in between ribs or neck when breathing)
- Head bobbing
- Nostrils flaring out when breathing
- Noisy breathing (such as wheezing, cough or chest congestion)
- Difficulty gaining weight

WHAT IS THE TREATMENT FOR CHRONIC LUNG DISEASE?

Medication

There are three general types of medicines used to treat babies with CLD. Not all babies will need each type of medicine.

Diuretics

The lungs of babies with chronic lung disease collect fluid easily. Diuretics (or water pills) help remove this fluid. This may help your baby breathe more easily. Diuretics are given by mouth or through a feeding tube (if your child has one). Diuretics can sometimes lower blood levels of sodium, potassium or chloride or cause dehydration. If your child begins to show signs of dehydration (diapers less wet than usual, no tears when they cry, dry lips or mouth), you should call your healthcare provider. Some examples of diuretics include chlorothiazide (Diuril®), hydrochlorothiazide (HydroDiuril®), furosemide (Lasix®) and spironolactone (Aldactone®). Your child may need regular blood tests to check their electrolytes.

Short-acting bronchodilators

Sometimes muscles around airways can tighten with viral infections, stress or exercise. This makes it difficult to breathe (similar to asthma).

This medicine helps to relax these muscles. It can be given with a metered dose inhaler (MDI, often called "the pump"), with a spacer or in a liquid form using a nebulizer (often called "the machine"). At home, these medicines are usually prescribed to be given every four hours when needed. If your child needs these medicines more frequently than every four hours, you should call your healthcare provider or take your child to the Emergency Department. Examples of short-acting bronchodilators include albuterol (Proventil®, Ventolin® and Proair®), levalbuterol (Xopenex®) and iprotropium (Atrovent®).

Steroids

These medicines help control and prevent swelling in the airways. They also decrease the amount of mucus in the airways. When prescribed, inhaled corticosteroids are given by a meter dose inhaler or a nebulizer. They are usually given every day to control chronic respiratory symptoms. Your healthcare provider will tell you when you can stop giving this medicine to your child. The most common side effect is thrush (yeast infection in the mouth). You should always use a spacer (also called an aero-chamber) when giving this medicine since this can help prevent thrush and allow the medicine to be delivered to the lungs. You should also rinse your child's mouth after giving this medicine to prevent thrush. Examples of steroids include beclomethasone (Q-Var®), fluticasone (Flovent®) and budesonide (Pulmicort®). Sometimes there is a need to give a corticosteroid, such as prednisone or dexamethasone (Decadron®) by mouth or feeding tube. These steroids are often prescribed if your child develops significant breathing problems caused by a respiratory viral illness. For illnesses such as these, prednisone or dexamethasone would be used.

Respiratory Support and Equipment

Oxygen: Some children with chronic lung disease need oxygen at home to achieve normal oxygen levels in the blood and to help with growth. Some children who are weaning down on their oxygen may only need to be on oxygen while asleep. It is important to give your child the amount of oxygen prescribed by your healthcare provider. Do not change the amount of oxygen without discussing it with your healthcare provider. If you think your child needs more oxygen than has been prescribed, call your healthcare provider immediately as this could be a medical emergency.

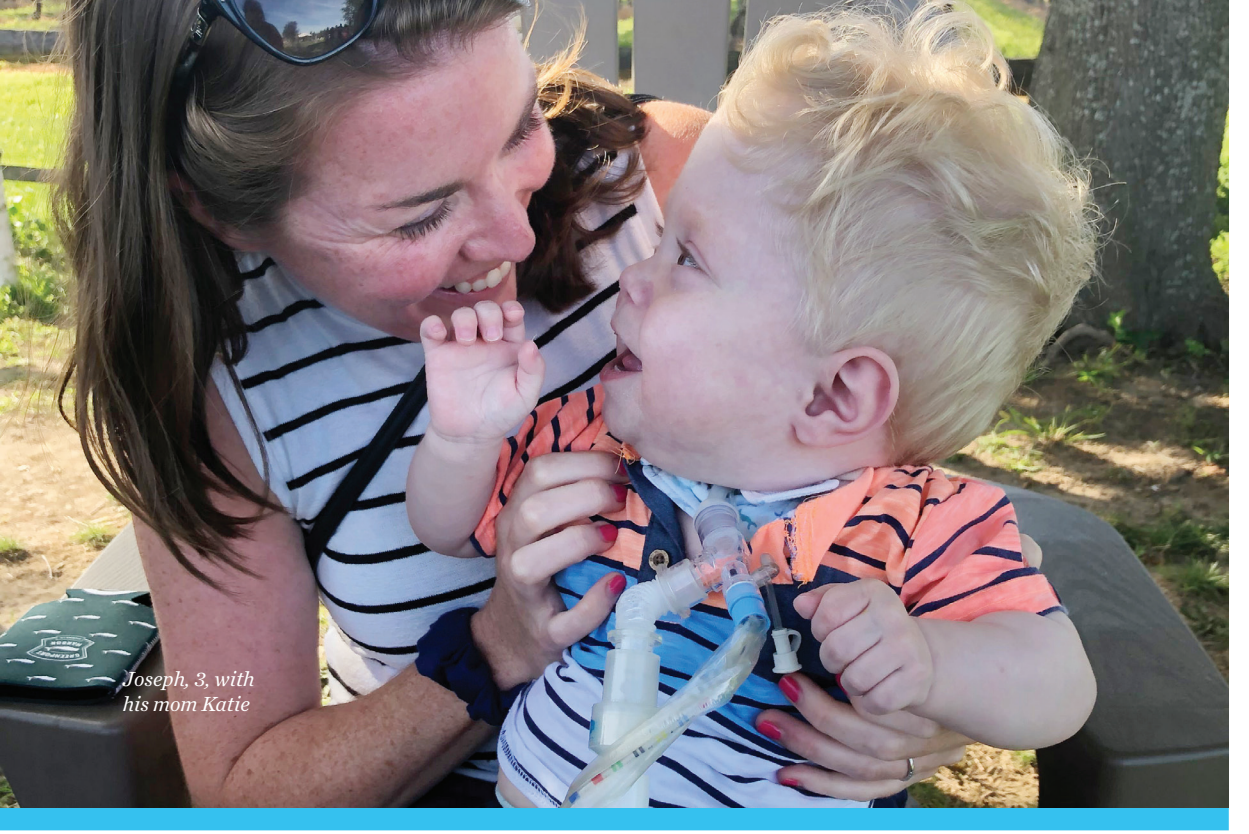
Tracheostomy: Sometimes children with upper airway abnormalities such as subglottic stenosis, vocal cord weakness or chronic respiratory failure will need a tracheostomy. Children who need a tracheostomy should always have an awake caregiver. The awake caregiver must be able to determine if your child is having breathing difficulties and must be able to suction the tracheostomy tube and change the tracheostomy tube if needed. These caregivers should also be trained in cardiopulmonary resuscitation (CPR). Blockage of the tracheostomy tube can prevent your child from receiving oxygen. This is an emergency, and suctioning of the mucus plug and/or changing the tracheostomy tube with bag and masking should be done immediately. You should call 911 for any emergency.

Respiratory therapist Joe Bolton teaches Kemin Sr. how to give his son, Kemin Jr., 1, corticosteroids with a spacer.

Home ventilator: Some children with chronic respiratory failure will have a tracheostomy and be on a home ventilator. These children should always have two working ventilators at home and batteries to support the ventilators if the electricity goes out. These children will usually need skilled nursing care at home and require frequent communication with their Pulmonary healthcare provider and Ear, Nose and Throat surgeon (otolaryngologist) who will help determine when the child can be safely weaned off the ventilator and have the tracheostomy removed.

If your child is on a home ventilator, you should not change ventilator settings at home or take your child off the ventilator for any period of time without speaking with your healthcare provider. Your child may need an overnight sleep study to determine their readiness to be weaned from a home ventilator.

Pulse oximeter: This is a machine that allows you to monitor your baby's oxygen level and heart rate at home. An alarm will go off if your baby has a low heart rate or a low oxygen level (usually below 92%). Always check your baby if the alarm sounds; sometimes there will be a false alarm if your baby is moving or because of poor placement of the probe. Call your healthcare provider if you are having trouble with using the pulse oximeter. Typically, the pulse oximeter will be used until your baby is off supplemental oxygen and respiratory support.



Joseph, 3, with his mom Katie

Nutritional Support and Equipment

Babies with chronic lung disease have more trouble growing than other babies. Some reasons include:

- Babies with CLD may have trouble sucking and swallowing, which makes it harder for them to breathe when they are feeding. Since they use more energy to eat, they have less energy to grow.
- Babies with CLD need extra nutrition to help them repair lung damage.

In order to get the right amount of nutrients, your baby may need a special formula. Usually by the time a baby is ready to leave the hospital, they will be able to take formula by mouth, but some babies need to be fed through a tube that delivers food directly into their stomach.

Tube Feedings

There are two types of feeding tubes.

- Nasogastric tube (NG-tube): inserted through the nose, down the throat and into the stomach
- Gastrostomy tube (G-tube): surgically placed through the abdomen and into the stomach

Feeding tubes are sometimes used for:

- Children who are not able to eat enough by mouth
- Children who aspirate (get food in their lungs when they swallow)

CONDITIONS THAT MAY AFFECT CHRONIC LUNG DISEASE

Gastroesophageal reflux (GER, acid reflux or heartburn) is when acid from the stomach backs up into the esophagus (the tube leading from the mouth to the stomach). A muscle located between the stomach and esophagus normally prevents stomach acid from coming up into the esophagus. All babies may have some reflux until they grow and this muscle gets stronger. Reflux can interfere with normal feeding and growth. Signs of reflux may include painful arching of the back, frequent spitting up of formula, vomiting, wheezing, stuffy nose or coughing.

Your healthcare provider may diagnose your child's reflux based on symptoms, but may also order tests to measure the amount of acid coming up from the stomach. Mild reflux can be treated by thickening the formula or breast milk as instructed by your healthcare provider. It can be helpful to raise the head of the bed, as instructed, burp the baby often and hold the baby upright after feeding. Some children need medicines to decrease acid in their stomach. If your child has severe reflux and none of the treatments above have worked, surgery to control reflux may be indicated.

Pulmonary hypertension, or high blood pressure inside of the lungs, is most commonly seen in infants with severe chronic lung disease. Although poor lung growth can cause pulmonary hypertension, some infants with CLD can develop pulmonary hypertension from pulmonary vein stenosis. Pulmonary hypertension and its causes can be diagnosed with an echocardiogram (an ultrasound of the heart) or a cardiac catheterization (a hospital procedure to measure pressures in your baby's heart). Symptoms may include an ongoing need for supplemental oxygen, edema (swelling) of the lungs or poor weight gain. Untreated pulmonary hypertension can damage the heart. Pulmonary hypertension is sometimes treated with supplemental oxygen and medications that can lower high blood pressure in the lungs. If your child has pulmonary hypertension, they should routinely see a cardiologist. They will often need echocardiograms to determine when to adjust and wean medications.

Respiratory syncytial virus (RSV) and **influenza virus** (flu) are common viruses that can make babies very sick. All children with CLD should receive an annual flu shot once they are 6 months of age and older. Most children under 2 years old with BPD should also receive monthly RSV shots, if recommended by their healthcare provider. In adults and many children, RSV often causes a basic "cold." But for babies who were born prematurely or babies with lung disease, RSV and the flu can cause worsening symptoms very quickly. These symptoms include difficulty breathing, breathing faster than usual and retractions (ribs sinking in). If your child has these symptoms, you should immediately contact your healthcare provider. They will likely ask you to bring your child to the office or the Emergency Department. These viruses are diagnosed by swabbing the inside of the baby's nose and sending the swab to a lab for testing. Test results are usually back the same day. These respiratory viruses can cause breathing problems and cough for several weeks. Your child may require additional treatments during this time.

Other respiratory viruses such as rhinovirus, adenovirus, human metapneumovirus or parainfluenza can cause similar respiratory symptoms in your child. There are no vaccines for these viruses, and testing for them is not generally done. If your child catches one of these respiratory viruses, they may develop significant breathing problems and require respiratory medications to help their breathing. If your child is wheezing, your child may need a short-acting bronchodilator and a steroid given by mouth or feeding tube. Your doctor may decide that your child needs to be admitted to the hospital.

Reducing risk of viral illness

You can decrease your child's risk of illness from viruses by doing a few simple things:

- ALWAYS wash your hands before touching your child.
- Keep your child away from people who have cold symptoms.
- Once your child turns 6 months old, they should receive a flu shot during the fall or winter. Your child will require two flu shots during the first flu season, then one flu shot every fall after that. All household members should receive the annual flu shot as well.
- If recommended by your healthcare provider, your child should receive a monthly RSV immunization (Synagis®) during RSV season (usually November to March). This immunization only protects your child for 30 days, so they will need a shot once a month during RSV season. It is only given to children up to 2 years of age. Some children with severe chronic lung disease also should receive Prevnar 13 immunizations starting at age 2 and then again every five years to help prevent bacterial infections caused by respiratory viruses.

Secondhand Smoke

Smoking around any children, especially premature babies, may cause them to have increased wheezing and difficulty breathing. It can also put them at risk for pneumonia, ear infections and other infections. The best way to protect your child is to not smoke tobacco or other substances (including electronic cigarettes). There is no safe way to smoke around your child. Most smokers wish to quit, but because nicotine is highly addictive, quitting smoking is not easy — but it can be done. There are a lot of resources available, whether online or in person with a healthcare professional, to help you quit.

If you or a family member is having difficulty quitting, please do NOT smoke in a car with your child or inside your home. Consider smoking outside and changing your clothing and washing your hands and face when you come back inside. Do NOT smoke around oxygen as this can cause a serious fire.

NATURAL COURSE OF CHRONIC LUNG DISEASE

Infants and toddlers

Respiratory viruses can very quickly lead to breathing problems in infants and toddlers with chronic lung disease. Many children with CLD require urgent appointments with their pediatrician or visits to the Emergency Department during the first two years of life. Some are admitted to the hospital. If your child goes to a daycare center, that may increase their chance of catching a virus.

Children

About 40% of older children with a history of chronic lung disease will develop asthma-like symptoms later in childhood or shortness of breath with exercise. They may require chronic respiratory medicines to treat these symptoms. These medicines may include an inhaled corticosteroid and/or albuterol. Even older children with a history of chronic lung disease may get sick quickly with respiratory viruses due to their decreased lung function. These children will need to have pulmonary function tests. The results of these tests help us create the best treatment plan.

Young adults

About 25% of young adults with a history of chronic lung disease will have chronic respiratory symptoms, which may appear as asthma, cough or shortness of breath with exercise.

Xander and Katorian, 3

WHEN TO CALL THE DOCTOR

It is very important to seek medical care immediately if your child is breathing differently. Depending on the severity of your child's lung disease, they may already show some symptoms, such as poor feeding or retractions. If your child's symptoms ever get worse or new symptoms appear, it is important to report these to their healthcare provider. You want to avoid further lung injury or life-threatening situations.

Call your healthcare provider if you see:

- Breathing that is too fast or too slow
- Continuous coughing
- Trouble catching their breath
- Nostrils flaring
- Retractions (skin sinking between ribs when breathing or using neck muscles to breathe)
- Noisy breathing (wheezing, grunting, chest congestion)
- Skin or lip color is pale, dusky or blue
- Temperature is too cold or too hot (fever)
- Poor feeding (not wanting to eat, not wanting to wake up to eat)
- Increased vomiting or diarrhea
- A change in activity (very fussy, sleeping more than normal, floppy)
- Urinating less often

CLINICAL RESEARCH

Clinical research is an important part of learning more about CLD and helping to improve outcomes for babies and children with BPD. To learn more about what research study opportunities are available, please contact the BPD Research Team at preemielungresearch@chop.edu.

YOUR CHILD'S CARE TEAM

Provider: _____

Social Worker: _____

Next appointment: _____

NOTES

NOTES

WAYS TO KEEP YOUR CHILD HEALTHY

- Get all immunizations.
- Wash your hands — and your child's hands — often.
- Give all medicines as prescribed.
- Go to all doctor appointments.

Please call **215-590-3749** if you need to contact a member of your treatment team or schedule/reschedule an appointment.

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