

CARING FOR YOUR CHILD

Handbook for School-age
and Adolescent Children Living
with Chronic Lung Disease
Related to Prematurity



Taylor, 6



**Children's Hospital
of Philadelphia®**

Division of Pulmonary & Sleep Medicine

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Most children who have chronic lung disease (CLD) of prematurity or a history of bronchopulmonary dysplasia (BPD) do very well. They can attend school, participate in sports, and lead happy and productive lives. However, it is important to recognize that your child may have ongoing respiratory challenges that may require special care, as well as follow-up visits with pulmonary and subspecialty services.

ROUTINE RESPIRATORY FOLLOW-UP AND MAINTENANCE RESPIRATORY CARE

Children with a history of BPD may have ongoing respiratory problems. Symptoms may be intermittent or chronic. Symptoms that may occur are:

- exercise intolerance
- chronic cough
- shortness of breath
- wheezing

Children with a history of BPD should be seen by a respiratory specialist, such as a pulmonologist, at least once a year through childhood. This evaluation may include pulmonary function tests (PFTs) and clinical examination. Frequency is based on severity of respiratory symptoms.

Some children may require maintenance or intermittent use of respiratory medicines, including different types of inhaled medicines. Some children with BPD may respond to asthma medications. Some children with BPD have airflow obstruction due to small airways caused by their prematurity and may not respond to asthma medicines. Routine pulmonary function tests are important to decide if your child may benefit from an inhaled medicine for their respiratory symptoms. PFTs can help decide if your child has airflow obstruction without symptoms.

Going to follow-up appointments is key. If PFTs are not performed routinely, decreased lung function may be missed and your child may not receive appropriate treatment or treatment could be delayed. Your child may be treated as though they have asthma even though the symptoms may or may not be due to asthma. Due to underlying disabilities, some children may not be able to perform PFTs and may require other testing.

Airway clearance techniques may help some children with BPD. Your healthcare team will discuss all options for your child to determine which suit your child's situation. Airway clearance techniques may include:

- manual or vest chest physiotherapy
- cough assist
- Acapella® device

Routine chest imaging is not done for children with BPD. Note that radiographic imaging (X-rays) may be needed during acute illnesses or if unexplained respiratory symptoms persist.

Children with BPD who have chronic wet coughs that are unresponsive to therapy may require a bronchoscopy to determine the cause of the cough.

RESPIRATORY SUPPORT

Supplemental oxygen

All children who require supplemental oxygen or are on respiratory support should have a pulse oximeter at home to monitor their oxygen levels. For most children, pulse oximetry readings should be 92% or above. If you notice your child's pulse oximeter is dropping below 92%, contact your healthcare provider immediately. Your child may need immediate evaluation from their primary care provider (PCP) or a visit to the emergency room.

As your child's lung function improves, they may be weaned off supplemental oxygen. Weaning off supplemental oxygen will depend on the severity of your child's lung disease. Your healthcare providers will partner with you to decide the best way to wean your child off supplemental oxygen. This will require careful monitoring with a pulse oximeter during weaning.

Usually, your child will first be weaned off supplemental oxygen during the day. Your healthcare provider will decide when support can be weaned at night. This may require an overnight sleep study.

NOTE: Caregivers should not wean oxygen without speaking first with their healthcare provider.

During periods of illness, supplemental oxygen requirements may increase even if your child typically only requires nighttime oxygen use. If this occurs, your child may require hospitalization for further management. Even after your child is completely weaned off supplemental oxygen, they may require oxygen during periods of illness.

Tracheostomy

If your child has a tracheostomy, every caregiver must be trained to perform tracheostomy care, tracheostomy tube changes, suctioning, CPR and manual bagging with oxygen. For safety reasons, an awake trained caregiver or skilled nurse should always be present to monitor your child's status.

Your child will need frequent follow-ups with their pulmonary and ear, nose and throat (ENT) providers to monitor their airway and respiratory status.

Children with tracheostomies may be at increased risk for tracheal infections. Tracheal infections may cause thickening of secretions, which can block a child's airways. Treatment of respiratory infections may require an increase in airway clearance therapies and/or antibiotics.

Thickened tracheal secretions may also occur if the child is not receiving enough hydration. Blockage of the tracheostomy tube can prevent your child from receiving oxygen. This is an emergency. Immediately suction the mucus plug, change the tracheostomy tube if unable to clear the tube and use bag and mask as needed. Call 911 for any emergency.

Home ventilators

Your child may require chronic (ongoing) ventilatory support. This may be done by either a noninvasive ventilation delivered through a nasal mask or a tracheostomy.

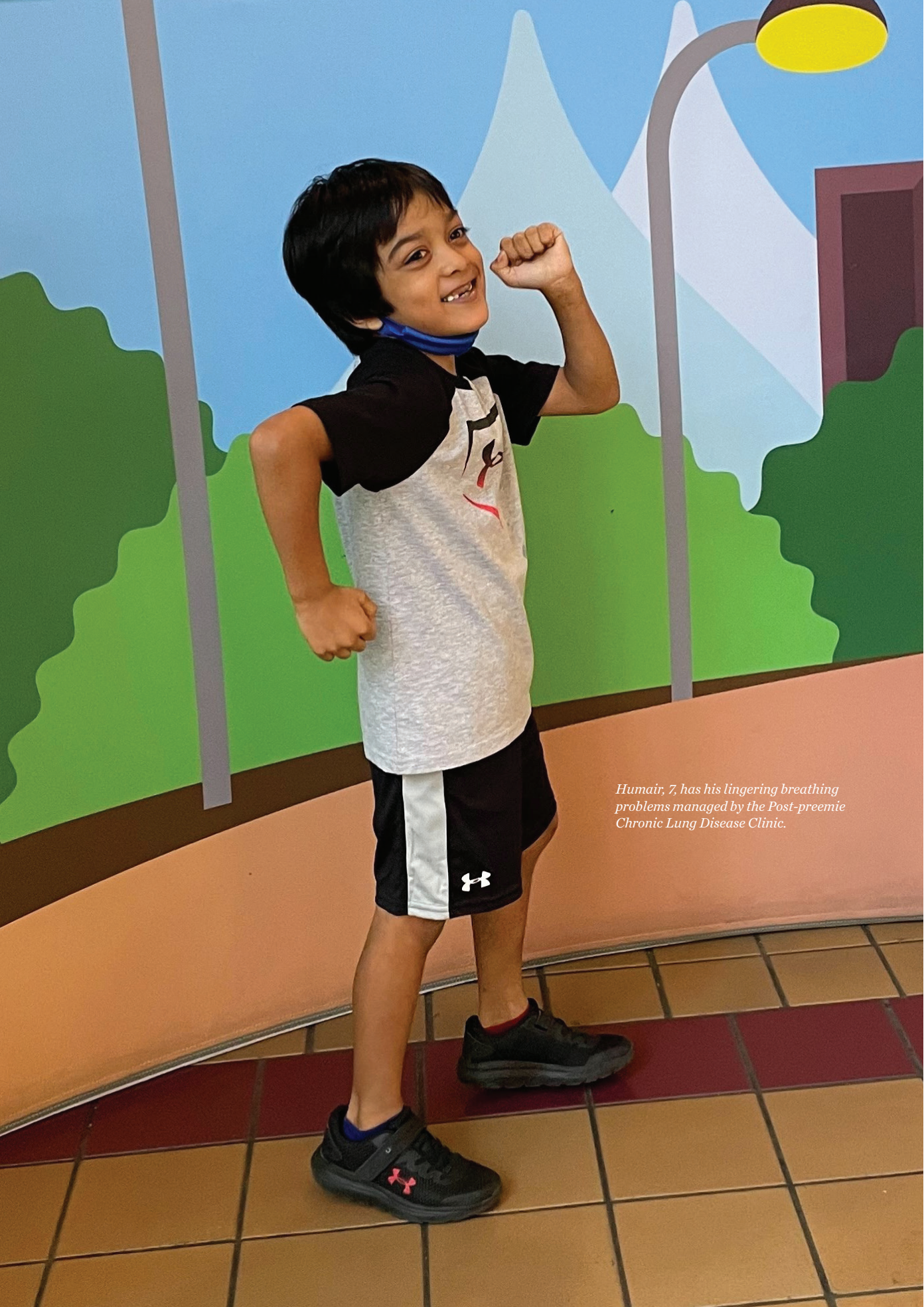
If your child's lung function improves, they may be able to wean off their home ventilator. However, weaning off of the home ventilator will depend on the severity of your child's lung disease and should be discussed with your healthcare provider.

Your provider may decrease settings on the ventilator and start sprints off the ventilator. Sprints are extubation readiness trials when your child uses CPAP for a set amount of time. During these sprints, your child's respiratory status and pulse oximetry must be closely monitored. When your child is off the ventilator during the day, an overnight sleep study may be ordered to decide if your child is ready to come off nighttime ventilation.

If your child has been receiving positive pressure ventilation through a tracheostomy and is weaned off the ventilator, your healthcare provider may recommend decreasing the size of the tracheostomy tube. If this is tolerated, capping the tracheostomy tube may be recommended during the day. If your child tolerates this, an overnight sleep study may be recommended to decide if your child is ready for tracheal decannulation (removing the tracheostomy tube).

Some children may have airway lesions or enlarged adenoid and/or tonsillar tissue. This may require surgery to remove them prior to the tracheostomy tube being removed.

NOTE: The weaning process is different for every child. For more information about the weaning process, please consult with your healthcare provider or specialist.



Humair, 7, has his lingering breathing problems managed by the Post-preemie Chronic Lung Disease Clinic.

MEDICINES

Despite improvements in your child's functioning or well-being, it is extremely important to adhere to your provider's medicine plan. This will help prevent emergency room, urgent care and unplanned primary care visits for your child. To be compliant with medicines, many children will require oversight.

- Some children will require daily medicines, including inhaled corticosteroids (Flovent[®], Asmanex[®], Pulmicort[®]) or inhaled corticosteroids with long-acting beta agonists (Advair[®], Symbicort[®], Dulera[®]).
- Other children will benefit from intermittent, or as needed, use of respiratory medications such as short-acting beta agonists (Albuterol[®]), inhaled corticosteroids (Flovent, Asmanex, Pulmicort), or inhaled corticosteroids with long-acting beta agonists (Symbicort).
- All children and adolescents should use spacers with their inhaled medicines, regardless of age.
- Some children may have intermittent or chronic nasal congestion due to allergies or respiratory illnesses. These children may benefit from antihistamines (Claritin[®] or Zyrtec[®]) and nasal steroids such as Flonase[®].

Vaccinations

Children with BPD should be fully vaccinated according to the recommended immunization schedule for children and adolescents. These vaccinations will be administered by your PCP. Vaccines may include:

- Annual flu vaccines should be given to children with BPD since they are at increased risk for more severe complications due to the flu.
- Children with a history of BPD may benefit from receiving Prevnar-13[®] boosters every five years to protect against certain kinds of bacterial pneumonia that can occur after a viral illness.
- Vaccination against COVID-19 is encouraged. If your child is COVID-19 positive, please contact your pulmonologist or PCP.

Children who do not make antibody responses to vaccines will need to be followed by an immunologist.

All eligible caregivers and family members should be vaccinated against the flu and COVID-19 as well. This will provide added protection for the child with a history of BPD.

ACUTE PULMONARY ILLNESSES

Children with BPD may have more severe respiratory viral infections and may get sicker as a result. These viral respiratory illnesses may require additional care that may include starting short-acting beta agonists, increasing inhaled corticosteroids or using short courses of oral steroids.

Pulmonary illnesses may be more severe in children with a history of BPD, and may require emergency room visits, urgent care visits and hospitalizations for treatment during acute illnesses. Consistent routine follow-up may decrease the need for acute care use.

If your child is seen at an outside hospital, it is critical to give those clinicians a detailed medical history, so your child receives appropriate care. Do not hesitate to ask the outside hospital to contact your pediatrician, pulmonologist and other subspecialists.

Severity of illness can increase quickly in children with BPD. Please contact your pulmonary provider or PCP as soon as you notice symptoms. Children who have an asthma care plan or acute illness plan should initiate their plan. Call your pulmonary provider or PCP.

NUTRITION

It is important to have adequate calories to promote healthy growth. To promote ongoing lung health, it is beneficial for your child to maintain a healthy diet. If your child is having trouble maintaining a healthy weight or difficulty with eating or feeding, your healthcare team may recommend seeing a dietician.

Some children will require supplemental calories to maintain a healthy weight during their childhood and adolescent years. Supplemental calories can be given through oral supplements or a feeding tube. Some supplements that can be used include Pediasure[®], Boost[®] or Kate Farms[®].

Children with cerebral palsy or scoliosis may be at increased risk for aspirating food into their lungs because of difficulty with swallowing. Lung damage can occur when food or liquid gets into the airways and lungs. Children who aspirate may require thickening of their liquids or a gastric or nasogastric (NG) tube for safe feeding.

SLEEP

Extremely low birth weight infants (less than 2.7 pounds) and children diagnosed with BPD are at higher risk for developing obstructive sleep apnea (OSA). OSA can cause sleep disruption, snoring, pauses in breathing, coughing, gasping and low oxygen saturations. This may be detrimental to your child's health.

Although many children snore, your healthcare provider may recommend an overnight sleep study to rule out OSA. OSA can interfere with a restful night sleep, leading to behavioral problems, irritability and fatigue in your child.

If your child has OSA, they should be seen by an ENT who specializes in the care of pediatric patients for further evaluation.

Treatment may include removal of the tonsils and adenoids (T&A) or noninvasive ventilation (CPAP or BiPAP) at night. If your child requires a T&A, they may need to spend the night in the hospital to monitor their breathing after the procedure. If your child is less than 2 years old, has severe OSA and has underlying breathing problems, they will need to spend the night in the hospital.

If your child intermittently snores with allergies or colds, they may benefit from nasal steroids to help with their nasal congestion.

BEHAVIORAL HEALTH

Pre-term children may be seen as being vulnerable to experiencing behavioral health problems because of early life events. It is important to allow your child to participate in age-appropriate activities and events as tolerated.

Children who are born premature may be at higher risk for developmental and behavioral problems such as attention deficit disorder (ADD) or attention deficit hyperactivity disorder (ADHD). If this is interfering with their daily activities and school life, your PCP or healthcare team may recommend behavioral programs or other treatments options.

Some children born pre term may experience periods of anxiety and/or depression. If your child is experiencing these symptoms or you have other concerns about your child's behavior, please contact your PCP or healthcare team, who can recommend a referral to a behavioral health provider.

Taking care of a child with complex medical needs may make it difficult for a caregiver to cope. Sometimes siblings are also affected by having a brother or sister who requires special care. It is important to speak to your own healthcare team, or the siblings' care team, which can assist with a referral to a behavioral health provider as needed.

SCHOOL

Most children born premature should be able to attend school in age and developmentally appropriate classrooms. Some children with preexisting conditions may need additional education or behavioral supports.

For young children who require respiratory and technology support, medical daycare facilities may be available. Children who have tracheostomy tubes, ventilators, supplemental oxygen or require feeding tubes will need to be monitored closely.

All children with tracheostomies and ventilators who attend daycare or are in the school environment will require a nurse skilled in trach and ventilator care.

Some children will benefit from Individualized Education Plans (IEPs) that will be developed with their school. This may include additional support for speech and occupational therapy or emotional support, among other services. Services will depend on your child's needs.

Some children may benefit from a 504 plan, which serves as a blueprint for how the school will provide supports and remove barriers for a student with a disability. A 504 plan provides services and changes to the learning environment to enable students to learn alongside their peers.

SPORTS AND PHYSICAL ACTIVITIES

Many children born premature with a history of BPD can participate in sports and other physical activities.

It is not unusual for them to have some activity limitations with sports. If they have exercise-induced shortness of breath, they may benefit from medicines such as short-acting bronchodilators prior to activities. If you start to notice other symptoms of exercise intolerance such as coughing, wheezing and shortness of breath, please contact your PCP.

Limiting outside activities may be recommended when air quality is poor.



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ENVIRONMENTAL EXPOSURES AND RESPIRATORY HEALTH

Children born premature with a history of BPD can have ongoing respiratory concerns, which can worsen under certain environmental conditions. These unintentional exposures may lead to more frequent acute care visits to your PCP, urgent care or the emergency room.

- Secondhand smoke or other substances that can be inhaled into the lungs can exacerbate or cause worsening of underlying lung problems. Secondhand smoke from tobacco or e-cigarettes can cause increased cough, wheezing, difficulty breathing and worsening of upper-respiratory tract infections. Children exposed to secondhand smoke may also have more shortness of breath with exercise.
- Thirdhand smoke exposure — when your child is in places where people smoke or have smoked — can also be detrimental to lung health. This can occur when chemicals are released into the air from couches and hard surfaces like walls and windows. This will occur even if someone is not actively smoking in the room and can happen many months and even years after someone has smoked in the house or car.
- If you are a smoker, it is more likely that your child will be a smoker. If you or your child are smokers, you can access the Pennsylvania quit line at 1-800-QUIT-NOW, and your PCP can prescribe nicotine replacement therapy to help you quit.

Adolescent smoking

It is common for adolescents to experiment with smoking when they are exposed to other adolescents who smoke. Substances that are inhaled directly into the lungs such as cigarettes, marijuana, e-cigarettes and hookah can be particularly harmful to a child or adolescent who was born premature.

Although e-cigarettes may be considered less harmful than conventional cigarettes, they can cause just as much harm to the lungs. Flavors such as mango, cinnamon, cool mint and Virginia tobacco in e-cigarettes can irritate the lungs and cause lasting harm. Many e-cigarettes contain nicotine, which can lead to life-long addiction.

Adolescents who smoke may have more respiratory illnesses and shortness of breath with exercise, and may require more emergency room or hospital visits. Over time this can decrease quality of life and worsen overall lung function.

Environmental triggers

Children with asthma-like symptoms may need to avoid environmental triggers, such as pollen, dust mites, rodents, cockroaches and other allergies that may trigger their respiratory symptoms. Other environmental exposures that can cause lung inflammation may include work exposures, where an individual is exposed to fine particles in the air.

Airflow obstruction

Some children with BPD are at increased risk for ongoing airflow obstruction as they age. This may put them at risk for early-onset chronic obstructive pulmonary disease (COPD). Minimizing exposure to respiratory infections and harmful environmental factors such as secondhand smoke may decrease the risk of further lung injury.

MEDICAL CONDITIONS THAT MAY BE ASSOCIATED WITH BPD

Children born premature may have other medical conditions that require long-term follow-up. The most common ones are highlighted here.

Cardiovascular care

Your child may have been diagnosed with a cardiac condition early in life due to their prematurity. Most of the time, these conditions resolve with age.

Pulmonary hypertension (PH) can occur in a child with underdeveloped lungs due to their prematurity. This often resolves during the first two years of life. PH is when the blood pressure in the vessels of the lungs is elevated. This can cause the right side of the heart to work harder than expected and can put stress on heart function.

Treatment of PH may require medicines, monitoring of heart function with imaging by echocardiogram and frequent visits to the cardiologist. Some children with a prior history of PH may be at increased risk for PH recurrence in adult life and should be periodically followed by their cardiologist.

Neurology

Intraventricular hemorrhage (IVH) can occur in some children who were born premature. IVH is due to abnormal bleeding in the brain that typically occurs immediately after birth in some pre-term infants.

Children with severe IVH may require ventricular peritoneal (VP) shunts to prevent hydrocephalous, which is increased pressure caused by fluid in the brain. These shunts are usually lifelong and may need to be replaced as the child grows or if the shunt clogs or malfunctions.

Children born prematurely with a history of severe IVH may be at increased risk of developing seizures during their preschool years. These seizures are usually managed by a pediatric neurologist. Sometimes your child may require daily (or as needed) medicine to control seizure severity and frequency.

Some children who are born premature may be at increased risk for cerebral palsy (CP). Children with CP can live normal lives. It may be helpful for your child to be seen by physical medicine and rehabilitation specialists.

Kidney Problems and Prematurity

A small percentage of children with BPD may develop chronic kidney disease, which can be associated with high blood pressure. These children should be followed by a kidney specialist (nephrologist) to monitor kidney function.



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INSURANCE

Some children with BPD may be eligible for Medicaid insurance. This insurance could serve as a child's primary insurance or secondary insurance. Medicaid as a secondary insurance may help to cover medical costs that are not covered by primary insurance.

In some states, children will qualify for Medicaid insurance only if the family's income is at a certain level. However, in other states, some children with BPD will qualify for Medicaid insurance regardless of parental income. Parents can typically apply online, by phone or by a paper application. The application process will vary by state.

FINANCES

When families have a child with complex medical needs such as BPD, it can take a toll on their finances; parents may have to take off from work for appointments and hospitalizations. They need to pay for transportation to and from the hospital. To offset these costs, some children with BPD may qualify for financial assistance through foundational support.

Financial assistance may include aid with rent, mortgage, food, medicines, equipment or utilities. It is important to speak to your social worker to decide if your family qualifies for this type of support.

Some children with BPD may qualify for Supplemental Security Income (SSI). SSI provides monthly cash payments to help meet the basic needs of children who have a physical or mental disability. Children under the age of 18 can receive SSI if they meet Social Security's definition of disability for children and there are limited income or resources in the household.

TRANSITIONING TO ADULT CARE

Adolescents with ongoing respiratory needs will need to transition to adult care. Your pediatric pulmonologist will recommend an adult pulmonologist with expertise in respiratory conditions caused by developmental lung disease such as BPD.

Adolescents with BPD can be seen by adult care providers beginning between 18 and 22 years old. The process of transitioning to adult care occurs much younger. Starting around age 13, children will need to start partnering with their parents to become independent in their own healthcare.

Prior to transitioning to an adult care provider, adolescents with BPD need to understand how to recognize respiratory symptoms that need treatment or intervention. They need to be able to understand their daily medicines, when to take these medicines and how these medicines work. If they require daily medicines, they need to understand the importance of taking these controller medicines even when they feel well. Faithfully taking controller medicines will help prevent pulmonary exacerbations or sick doctor visits. Adherence to their medication routine will allow an individual to be active and partake in normal daily school, work and recreational activities.

Before transitioning to an adult provider, it is important for an adolescent to know their medical history. This should be provided by their pediatric healthcare provider, as knowledge of early life events and prior respiratory interventions may help the adult provider with optimizing care.

Taking personal responsibility for one's own care is important and will help make this transition smooth.

Adolescents with developmental delay will still need to be transitioned to adult providers. However, parents may continue to be their children's guardians and the decision-makers in the care of their children. Parents will partner with the adult providers.

NOTE: It is important for the adult healthcare provider to know your child's complete medical history so that your child continues to be followed by appropriate specialists.

CLINICAL RESEARCH

Clinical research is an important part of learning more about chronic lung disease and helping to improve outcomes for children, adolescents and teens with BPD. To learn more about what research study opportunities are available through Children’s Hospital of Philadelphia’s robust research program, please contact the BPD Research Team at premielungresearch@chop.edu.

Your Child’s Care Team

- Provider: _____
- Social Worker: _____
- Next appointment: _____

LIST OF ABBREVIATIONS

ADD	Attention deficit disorder	IVH	Intraventricular hemorrhage
ADHD	Attention deficit hyperactivity disorder	NG	Nasogastric
BiPAP	Bi-level positive airway pressure	OSA	Obstructive sleep apnea
BPD	Bronchopulmonary dysplasia	PCP	Primary care provider
CLD	Chronic lung disease	PFT	Pulmonary function testing
COPD	Chronic obstructive pulmonary disease	PH	Pulmonary hypertension
CPAP	Continuous positive airway pressure	SSI	Supplemental Security Income
ENT	Ear, nose and throat	T&A	Tonsils and adenoids
IEP	Individualized education plan	VP	Ventricular peritoneal

NOTES

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