

What is Pulmonary Arterial Hypertension?

Pulmonary arterial hypertension (PAH) is characterized by high blood pressure in the lungs. It is not the same as high blood pressure in the body, which is measured by a blood pressure cuff on the arm. For people with PAH, the right side of the heart works harder to push blood through narrowed vessels in the lungs. Eventually, the extra stress causes the heart to enlarge and lose flexibility. That decreases the heart's ability to push blood out of the heart, through the lungs, and to the rest of the body.

Symptoms include:

- Fatigue
- Exercise intolerance
- Shortness of breath with activity
- Dizziness
- Nausea
- Loss of appetite
- Fluid retention (swelling of face, ankles, hands)

Symptoms that require emergency services (911) include:

- Severe chest pain
- Difficulty breathing
 - Fainting
 - Seizures
- Extreme pallor or cyanosis (bluishpurple discoloration) of the lips/nailbeds

Important Considerations

Always notify parents/guardians about your concerns.

Some children with PAH may not tolerate being outside when the weather is cold, hot, or humid, as it may affect their breathing. They may need to stay indoors during lunch and/or recess times.

Children with PAH must drink lots of liquids. While at school, they should keep a water bottle on their desk so they can drink throughout the day.

Special washroom privileges may be needed for children taking water pills (diuretics) or those who experience diarrhea as a side effect of their medication. They must be allowed to go to the washroom as often as necessary.

Attending school full-time can be exhausting and stressful so the student may need an Individualized Education Plan. A student with PAH can alternate between good and bad days. For some, a simple cold can cause significant illness that requires close medical attention. They may also have many medical appointments and may need to travel far to see specialists.

Children with PAH may be unable to participate in all physical activities and may require modifications or alternate activities during Physical Education classes. It may be necessary to have a meeting at the school with the student, parent(s), and/or other caregiver(s) to discuss appropriate alternate activities during gym time.

Children with PAH may need extra time and/or elevator passes to move between classrooms.

Children with PH who don't feel well should be evaluated at the nurse's office. The nurse should always check the student's temperature and refer to the child's Safety Plan to determine the course of action.

This resource was developed by the Pediatric Committee of PHA Canada. Some of the information has been adapted, with permission, from materials created by PHA USA