

Living with your Pulmonary Arterial Hypertension



1st edition



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This content does not replace the recommendations, diagnoses or treatments given by your health care professional.

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Introduction

You have just been diagnosed with pulmonary arterial hypertension (PAH). It is a rare disease that can have serious consequences if left untreated. However, new treatments are helping to better control the progression of the disease.

Here are a few questions that may interest you:

- Why am I out of breath?
- How will my disease progress?
- Are there medications to treat this disease?
- Will I be able to continue working?

This guide addresses these various questions. It contains a lot of information that is useful for you to better understand your condition.

“ To help control PAH symptoms, you must first understand your disease. ”

Some statistics

- It can take about two years for PAH to be diagnosed after the first symptoms appear.
- PAH affects 2 to 3 times more women than men.
- The average age at the time of diagnosis is 50 years.
- PAH affects more than 100,000 people around the world. This represents 15 to 50 people per million.
- In Canada, it is estimated that between 2,000 et 10,000 people suffer from PAH. The number increases by about 250 to 500 cases per year.

Description of PAH

What is PAH?

PAH is a disease that thickens the lining of the small arteries in the lungs. This lining hardens over time.

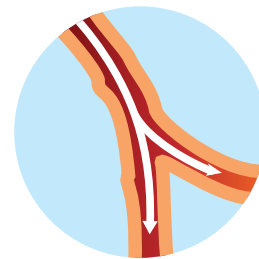
The thickening of this lining decreases the space in which blood can circulate in the arteries and increases resistance to its passage.



Healthy
blood vessel

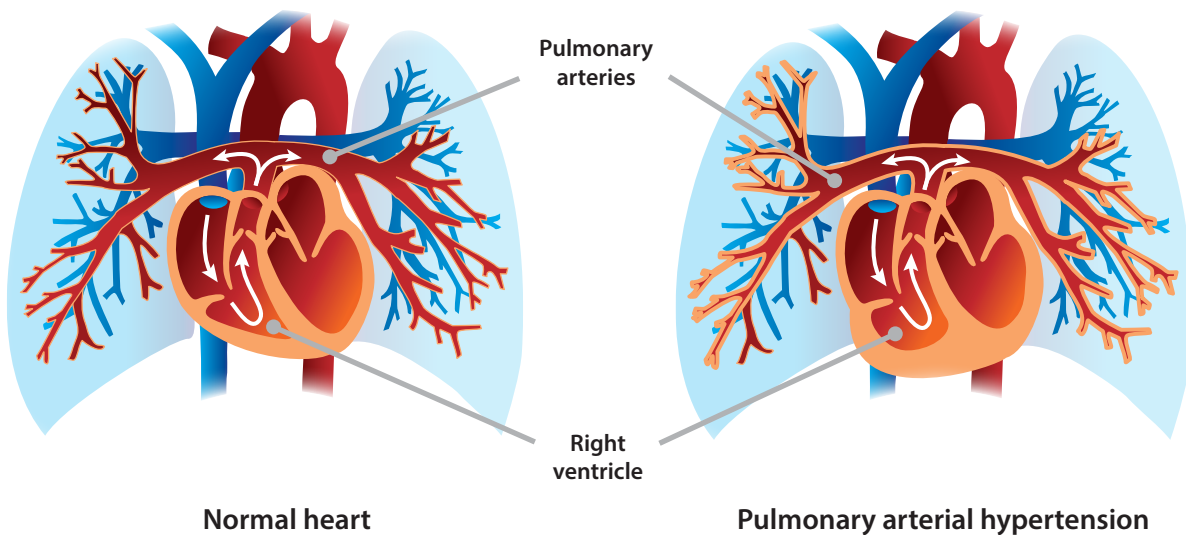


Blood vessel
affected by PAH



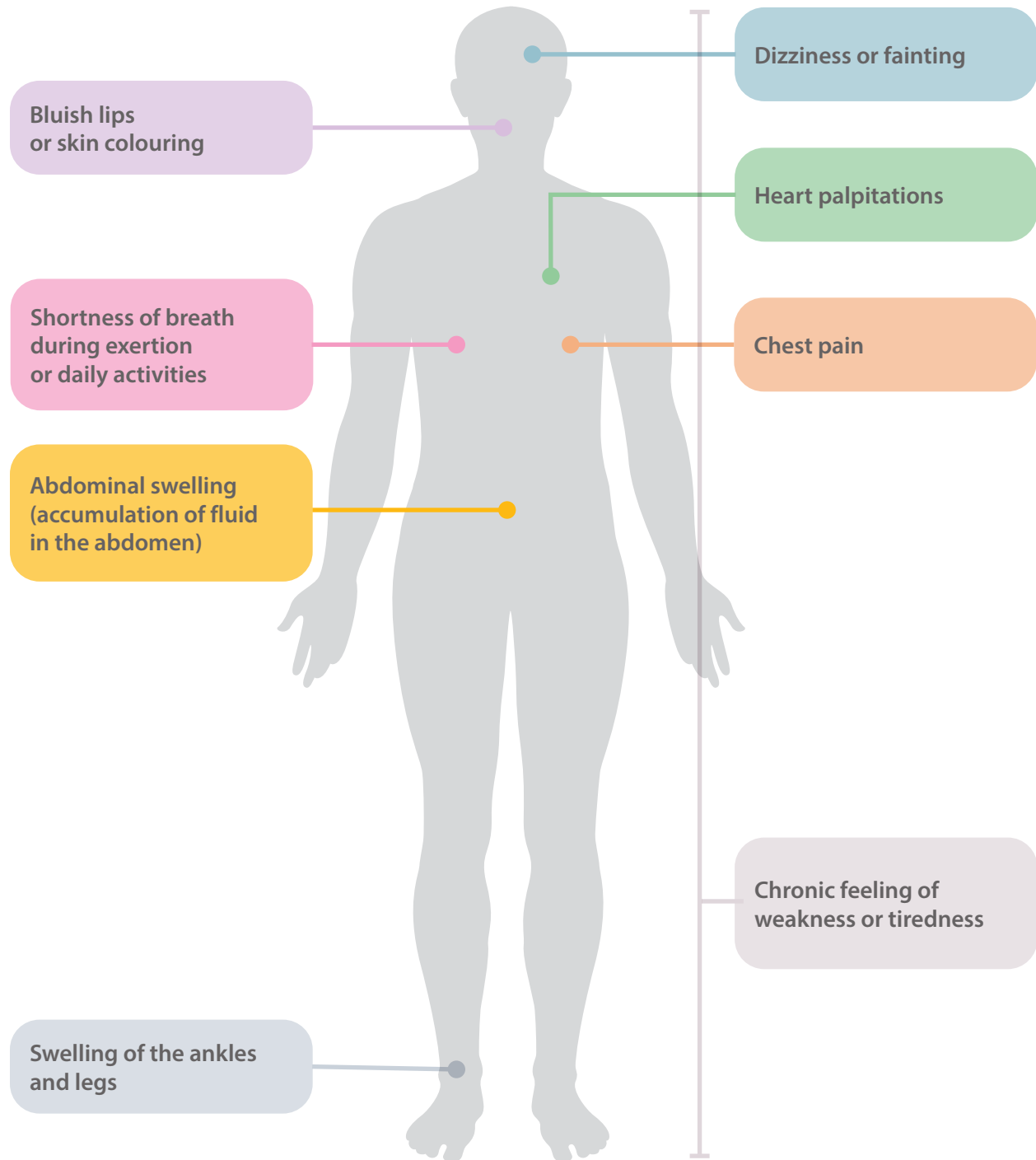
Disease
progression

Pressure in the pulmonary arteries rises. This high pressure tires the heart. In the long term, it can cause the right ventricle of the heart to be unable to efficiently eject blood toward the lungs (right heart failure).



Description of PAH

What are the symptoms?



Description of PAH

What is functional classification?

After assessing your symptoms and test results, your specialist will determine your functional classification. Functional classification is a tool that helps assess the **severity of your symptoms**. It helps your doctor to determine the appropriate treatment and follow-up for you. Functional classification is not a tool to determine the stage or progression of the disease.

The tool used is the New York Heart Association (NYHA) functional classification.

Description of functional classes

- Class 1** PAH that does not limit ordinary physical activities. These activities do not cause shortness of breath, excessive fatigue, thoracic pain or malaise (chest pain or fainting).
- Class 2** PAH that causes a minor limitation in physical activities. There is no discomfort at rest, but ordinary physical activity causes shortness of breath, excessive fatigue, thoracic pain or malaise (chest pain or fainting).
- Class 3** PAH that causes significant limitation in physical activities. There is no discomfort at rest, but doing minor physical activity leads to shortness of shortness of breath, excessive fatigue, thoracic pain or malaise (chest pain or fainting).
- Class 4** PAH that causes an inability to perform any physical activity. Signs of right heart failure such as shortness of breath and fatigue can occur at rest and are worsened by any physical activity.

Description of PAH

What are the causes?

Several diseases can cause high blood pressure in the arteries of the lungs. However, PAH is in a class of its own.

The causes of PAH are as follows:

- Unknown cause (idiopathic PAH)
- Faulty gene (familial or hereditary PAH)
- Autoimmune disease such as scleroderma or lupus
- Liver disease (portal hypertension)
- Acquired immunodeficiency virus infection (HIV)
- Exposure to certain toxins or drugs (e.g. methamphetamines)
- Taking appetite suppressants (anorexigenics)
- Heart malformation at birth (congenital heart disease)

There are also other causes of Pulmonary hypertension (besides PAH). Some respiratory or heart diseases can also be associated with Pulmonary hypertension (e.g. pulmonary fibrosis, left-sided heart failure).



Chronic thromboembolic pulmonary hypertension (CTEPH)

CTEPH is a type of pulmonary hypertension caused by the formation of blood clots that remain in the arteries of the lungs. This is the only form of pulmonary hypertension that, in some cases, can be cured with surgery.

Treatments

Not all treatments are necessarily appropriate for all people with PAH. Your doctor will decide which one will benefit you.

What are the specific treatments for PAH?

Depending on your type of PAH and the severity of your disease, your specialist will offer you treatment adapted to your condition using one or a combination of medications. There is still no cure for the disease. The goal of medication is to stabilize the progression of the disease and control your symptoms.

Here are the medications currently available to treat PAH. All these medications help relax (dilate) the pulmonary arteries. They allow the heart to pump blood more efficiently to the lungs and make it easier for blood to pass through the pulmonary vessels.

These medications are:

- Bosentan (Tracleer)
- Macitentan (Opsumit)
- Ambrisentan (Volibris)
- Sildenafil (Revatio)
- Tadalafil (Adcirca)
- Riociguat (Adempas)
- Selexipag (Uptravi)
- Epoprostenol (Caripul) continuous infusion
- Treprostinil (Remodulin) continuous infusion

“ There is active research on pulmonary hypertension.
Your specialist may suggest that you participate in a clinical study
to try a new treatment that may benefit you. ”

Treatments



Reimbursement and access to medication program

PAH treatments are expensive and require a process to be reimbursed by your group insurance or government drug insurance. Support programs can assist you in this process. Also, your treatment team must coordinate certain medications with your pharmacy.

The clinic nurse will inform you if there is a support program for the medication chosen by your specialist.

- Consult your pharmacist before starting any new prescribed or over-the-counter treatment.
- Inform your pharmacist and your family doctor of any changes in your medication made by your PAH specialist.
- Always carry an up-to-date list of your medications.

Treatments

What are the general treatments?

Diuretics

Many people with PAH have to take diuretics. Diuretics are medications that make you urinate to decrease swelling in your ankles, legs, hands, face and abdomen.

Examples of diuretics:

- Furosemide (Lasix)
- Spironolactone (Aldactone)
- Metolazone (Zaroxolyn)
- Amiloride (Midamor)

You should continue to monitor the amount of fluid you take in per day, even if you take diuretics. Your doctor will often recommend that you limit your fluid intake to 1.5 to 2 litres per day. They will also recommend that you limit your salt consumption.

Calcium channel blockers

Calcium channel blockers are medications that help dilate the blood vessels. They increase the supply of blood and oxygen to the heart and reduce its workload. These medications are prescribed to a minority of people with the disease (about 5%). For the majority, their effectiveness is considered limited and may even cause their condition to deteriorate.

Examples of calcium channel blockers:

- Amlodipine (Norvasc)
- Nifedipine (Adalat)
- Diltiazem (Cardizem)

Treatments

Anticoagulants

Anticoagulants are medications that thin the blood. They prevent the formation of clots in a vein or artery. Your doctor will assess whether it is necessary to prescribe you anticoagulants or not depending on your type of PAH.

Examples of anticoagulants:

- Warfarin (Coumadin)
- Nicoumalone (Sintrom)
- Apixaban (Eliquis)
- Dabigatran (Pradaxa)
- Rivaroxaban (Xarelto)
- Edoxaban (Lixiana)

You should take certain precautions when taking anticoagulants:

- Wear a bracelet indicating that you are taking an anticoagulant.
- Inform all health professionals that you visit (e.g. dentist, family doctor) that you are taking an anticoagulant.
- Use a toothbrush with soft bristles.
- Be careful when using sharp objects.
- Avoid aspirin-based medications and anti-inflammatories like ibuprofen (Advil, Motrin) or naproxen (Naprosyn), unless prescribed by a doctor.
- Avoid contact sports and those that may cause injuries (e.g. hockey, martial arts).
- If you have signs of bleeding, see your doctor.

Pay particular attention to your diet when taking anticoagulants:

- If your doctor has prescribed Coumadin for you, plan a stable and moderate intake of some green vegetables rich in vitamin K (e.g. asparagus, broccoli, spinach, parsley).
- Moderate your alcohol consumption. You should not drink more than one glass per day.
- Avoid over-the-counter vitamin supplements and natural products (e.g. vitamin E, glucosamine, ginseng, algae tablets, St. John's wort). They can interact with your medication. Talk to your pharmacist if necessary.

Treatments

Oxygen

For some people, oxygen at home is sometimes necessary to reduce exertion of the heart. It is generally prescribed when the blood oxygen level is lower than 90% after several minutes at rest.

Your doctor will evaluate your need for oxygen based on:

- The blood gases analysis (blood drawn from the artery in your wrist)
- The results obtained from the 6-minute walk test

Oxygen therapy monitoring will be provided by the local community health centre (CLSC) in your region or the community home specialized respiratory care service, if applicable.

Vaccines

Vaccines strengthen the immune system. They help the body defend itself against viruses and bacteria.

- You and your loved ones should get the flu shot every year.
- Discuss with your doctor whether other vaccines are relevant for you (e.g. COVID-19, pneumococcus, zona).



Treatments

What medications should I take if I have a cold, nasal congestion or allergies?

Colds

To relieve cold symptoms, you should first rest and stay well hydrated. Drinking warm liquids can help relieve congestion. You can take acetaminophen (Tylenol) to decrease the pain and discomfort associated with a cold.

It is not advised to take anti-inflammatories such as ibuprofen (Advil) or naproxen (Naprosyn) because of the water and salt retention that is associated with them.

Nasal congestion

To relieve nasal congestion, you can use saline nasal sprays such as Salinex or Sinus Rinse. If the nasal congestion is severe and annoying despite using a saline solution, talk to your doctor.

Allergies

To treat allergies, check with your pharmacist to see what medications you can take.



Decongestants and expectorant or cough suppressant syrups

These medications should not be used by people with PAH. They often contain pseudoephedrine or phenylephrine. These substances elevate blood pressure and pressure in pulmonary arteries. They also make the heart work harder. You can find them in most cold and allergy tablets and syrups.

Treatments

Is a lung transplant possible?

Lung transplantation involves removing a person's diseased lungs and implanting healthy lungs from a compatible donor. You may be offered a transplant if your disease progresses severely despite optimal treatment and if you are eligible for this intervention.

Do I need to plan for my end-of-life care?

PAH is a chronic and progressive disease. Despite treatment advances, they may not be successful in stabilizing the disease and death may occur. Your doctor may suggest medications to reduce or relieve symptoms such as shortness of breath. Nevertheless, often difficult discussions about the progression of the disease and wishes for end-of-life care become necessary.

Discussing the future helps reduce stress and anxiety. By bringing up this subject with your loved ones, you can think about possible options and make decisions sooner and without pressure. Feel free to also talk with your treatment team. Some information documents on this subject may help you.



Living with PAH

What are some strategies for living with PAH?

There are several strategies to help you live better with your PAH. Maintaining your health and the support of your loved ones are important allies in the fight against the disease.

Choose good life habits.

Adopt a healthy diet and proper sleep hygiene. Moderate your alcohol consumption.

Avoid smoking tobacco or cannabis, or vaping (e-cigarette). Also avoid any drug use.

The I QUIT NOW program is available to you if you want help quitting smoking. Call 1 866 527-7383 or visit tobaccofreequebec.ca/iqitnow. You can also speak with your pharmacist to start nicotine replacement therapy.

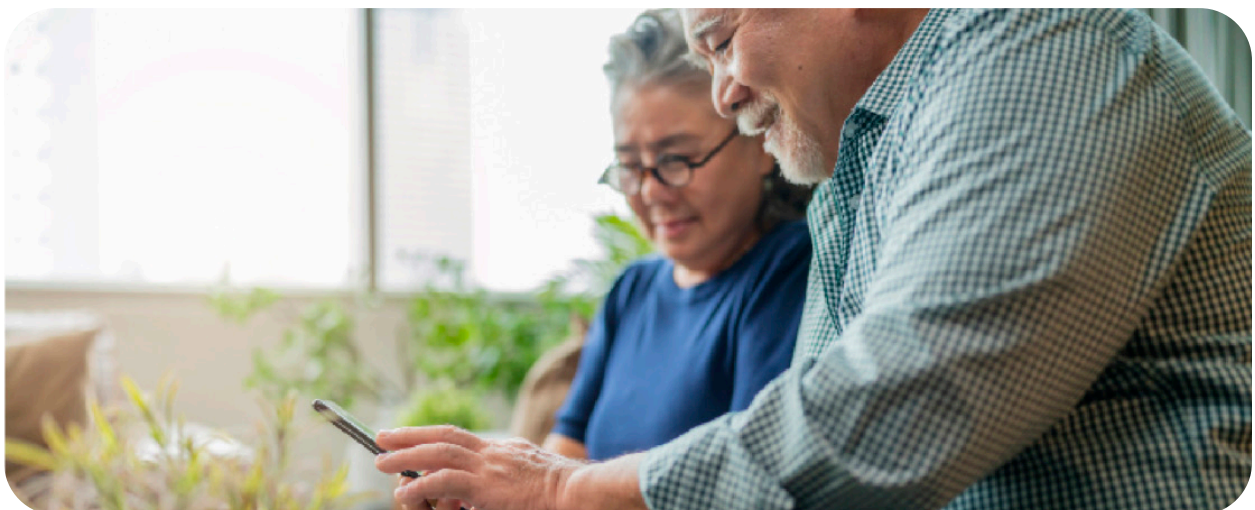
Take care of yourself.

You are the priority. Plan for time to rest during the day. As the disease progresses, you may need to reduce your activities. You have to deal with the limitations and grief related to the disease and learn to live with it. Surround yourself with optimistic people!

Prevent infections.

Here are a few practical tips to prevent infections:

- Wash your hands often. Use an antibacterial gel if soap and water are not available.
- Avoid contact with people who have a cold, flu, gastroenteritis or any other contagious disease.
- Use good dental hygiene.



Living with PAH

Reach out to your family and loved ones to get them involved.

It's important to talk about the effect the disease has on your life, your frustrations, and your disappointments, and the difficulties associated with them. Talk to them about your needs and emotions. Give them documentation on the disease so that they'll be able to understand you better.

If you have young children or grandchildren, explain the disease to them using simple words. Children need knowledge to adapt to change. There are books that explain PAH to children. Talk to your PAH clinic nurse; she can advise you.

Ask for help.

Call on your social network, community organizations or self-help groups. Check with your CLSC to find out what resources are available for your situation.

Participate in self-help groups.

Associations for people with PAH offer information on the disease and psychological support. They allow people with PAH to share their experiences. Their goal is to break the isolation of people with PAH.

Talk about your concerns with the PAH clinic nurse. She will be able to give you contact information for various associations. You will find this information at the end of the document (see [Useful resources](#)).



Important

Feel free to contact Info-Santé at 811 if you are experiencing psychological distress or if you need to talk to professionals quickly. Call 211 or visit qc.211.ca to find community resources available in your area.

Living with PAH

Can I continue working?

If you feel like you're able to continue working, do it! It's the best way to maintain your social ties. You can also work part-time or from home (remote work). Talk to your employer about the possibility of adapting your work to suit your abilities.

Contact your employer's human resources department to find out what disability or long-term illness benefits you may be entitled to.

Meeting with a social worker and an accountant can help you find out about what services you are entitled to (parking sticker for disabled people, tax credits for disabled people, reimbursement of expenses for medical care not provided in your area, etc.).

Some forms must be completed by the doctor. Bring them with you to your visit.

Don't feel guilty about being sick.

“ Accept that you can't do things the way you did before. ”

Lean to listen to yourself and adjust yourself according to your limits.



Living with PAH

Can I travel despite my PAH?

If you are planning a trip, make sure that your condition is stable. Discuss it with your doctor. Prepare everything you will need before your trip.

- Get insurance to protect you in case of a health problem.
- Keep an updated list of your allergies and medications with you at all times.
- Take extra medication in case of unexpected circumstances.
- Use bags or suitcases with wheels (never carry very heavy bags or suitcases).

Air travel

The air we breathe contains 21% oxygen. The higher in altitude we climb, the less oxygen there is. Aircraft cabins are pressurized to maintain this level of oxygen. However, above 2,400 m in altitude, the amount of oxygen available in the air and in the aircraft decreases. You may be affected by this decrease in oxygen during and after the flight.

If you're planning a trip by plane, tell your doctor. They may be able to prescribe an altitude test (hypoxia test) to check whether you will need oxygen during the flight. It's better to get a medical note justifying the need to bring certain medical equipment on board the plane (if you are receiving continuous infusion treatment, for example).

Sun exposure

Your skin may become more sensitive to the sun because of your medications.

- Use a sunblock with a sun protection factor of SPF 30 or higher.
- Wear a hat.

Sexuality and pregnancy

How can I handle my sexuality?

Learning that you have PAH can be an emotional shock. Other factors such as fatigue, shortness of breath, anxiety, anger, depression and changes in your body perception can have an impact on your life, intimate relationships and libido. This is often the case with people who have chronic diseases.

Preserving your love life helps maintain your quality of life. Talk openly with your partner. They may be afraid of making you sicker. Good communication is essential.

It's important to come to terms and accept the physical and emotional changes caused by the disease. Try to find enjoyable ways for you and your partner to nurture your intimacy, such as cuddling, massages and positions that allow you to be less out of breath. If you respect your exertion tolerance, sexual relations should not cause any symptoms or be dangerous to your health.

An evaluation of your physical condition may be necessary before engaging in sexual relations. Don't hesitate to approach the subject with your health care team, who will be able to reassure you and give you advice if needed.

“ Don't push away happiness, friendship or relationships with others. Don't let others make decisions for you. Enjoy the good times in life. ”

Sexuality and pregnancy

Is pregnancy risky when you have PAH?

Women with PAH are strongly advised not to become pregnant. Pregnancy involves a significant risk of mortality for the mother and the baby. Deaths in pregnant women with PAH can occur in 1 to 2 women out of 4.

During pregnancy, blood volume increases and can almost double. This increases the heart rate and decreases the ability of the heart to push the blood throughout the body and the lungs. For a person with PAH, these changes during pregnancy are generally poorly tolerated. They can lead to increased pressure in the lungs and severe right heart failure.

Several medications used to treat PAH can cause fetal malformations. In addition, numerous problems can occur during labour and delivery:

- Blood loss
- Uterine contractions
- Reaction of the body to pain
- High risk of blood clot formation (emboli)

These problems may harm the health of the mother and the baby.

Although it is strongly discouraged, if you're planning a pregnancy, discuss it with your doctor first.

Sexuality and pregnancy

Which contraceptive methods can I use?

It's necessary to use a safe and effective contraceptive method like an intrauterine device (IUD). Condoms or diaphragms should only be used temporarily, while waiting for a safer contraceptive method.

It is recommended to avoid using oral contraceptives due to the higher risk of blood clots. Certain PAH treatments can also interact with oral contraceptives and make them less effective. If necessary, discuss it with your doctor. They will determine the best oral contraceptive for you or refer you to a contraceptive specialist.

For some women, giving up the idea of being a mother can raise many emotions. It's important to approach the subject openly with your life partner. Also, the fear of becoming pregnant can affect your intimate relationship and decrease a couple's desire. If necessary, talk to the clinic nurse. They can refer you to an appropriate resource.



Diet

Do I have to limit my salt intake?

Salt (sodium) is an essential mineral found in the human body and in several foods. It is necessary to the proper functioning of the body. However, when too much is consumed, the body retains too much water. This fluid retention increases the workload of the heart, which can lead to a deterioration in your PAH. People with PAH should reduce their salt intake by controlling their diet.

Here is some information to help you limit your salt intake:

- Salt is found in greater quantities in pre-packaged or prepared foods.
- Salt can also be hidden in sweetened foods (e.g. cereals, breads, desserts).
- Even foods that indicate they have a reduced salt content may contain a lot of salt.

Nutrition labels

Read the nutrition information labels when you're grocery shopping. Look for foods that contain **140 mg of sodium or less per serving**, or 6% of the recommended daily intake. Pay attention to serving sizes indicated on the nutrition label. If you eat more than the suggested serving, the amount of salt you ingest also increases.

Nutrition Facts	
Serving of 1 cup (200 g)	
Amount per Serving	% Daily Value
Calories 260	
Fat 13 g	20 %
Saturated 3 g + Trans 2 g	25 %
Cholesterol 30 mg	10 %
Sodium 660 mg	28 %
Carbohydrates 31 g	10 %
Dietary Fibre 0 g	0 %
Sugars 5 g	
Protein 5 g	
Vitamin A 4 %	• Vitamin C 2 %
Calcium 15 %	• Iron 4 %

Suggested serving. Sodium values are calculated for this serving size. Caution: if you eat more of it, the amount of sodium consumed also increases.

Look at the number of mg of sodium (salt) for one serving. You should aim for 140 mg of sodium or less per serving.

Percentage of the recommended daily salt intake. For example, if you eat one serving of this food, you have consumed more than one quarter of your daily salt allowance.

Tips for a low-salt diet

- Do not salt your food while cooking.
- Do not add table salt to your meals.
- Avoid particularly salty foods.
- Get store-bought seasonings with no salt added.
- Choose fresh or frozen vegetables over canned ones.
- Choose canned goods with no salt added.
- Cook homemade sauces made with fresh or canned tomatoes with no salt added for your pasta.

Tips for enhancing the flavour of foods

- Steam or bake foods. These cooking methods preserve the flavour of foods.
- Season your meals with lemon juice, garlic, shallots or herbs (e.g. basil, parsley, thyme).
- Use low-sodium recipes to prepare your meals. Check out the Heart and Stroke Foundation of Canada's recipe section!

Diet

Foods to favour

- Unsalted peanuts and nuts
- Homemade or “no salt added” broths
- Fresh fruit, dried fruit
- Low-sodium vegetable or tomato juice
- Fresh or frozen vegetables
- Canned legumes with no salt added
- Eggs
- Low-sodium breads and crackers
- Plain pasta, rice, flour, couscous
- Baked or boiled potatoes with no salt added
- Tapioca
- Fresh meats, poultry, fish
- Yogurts

Foods to avoid

- Salted canned foods
- Breaded foods
- Salted peanuts and nuts
- Salted crackers
- Regular store-bought bouillon broths, liquid or cubes
- Cold cuts (e.g. ham, sausages)
- Chips and snacks
- Salted cheeses
- Vegetable or tomato juice
- Pickles
- Store-bought soups and sauces
- Store-bought dips or spreads
- Smoked or cured meats



Do I have to limit my fluid intake?

Your PAH specialist may ask you to limit your fluid intake (fluid restriction). Taking in too much fluid can lead to an accumulation of water in your body. This requires additional effort from your heart and causes fatigue. This fluid retention can lead to swelling in your feet, ankles, legs or belly. You may also notice weight gain, increased shortness of breath or dizziness.

It is generally recommended to keep daily fluid intake at 1.5 to 2 litres. **Your doctor will let you know the amount of fluid you are allowed per day, depending on your condition.**

Fluids include:

- water
- hot or cold beverages (tea, coffee, milk, juice, soft drinks, alcohol)
- soup broth
- soup
- Jello-type jelly and frozen desserts
- ice

It is important to drink a minimum of 1 litre of fluid per day. This protects your kidneys and prevents you from getting dehydrated.

“ Your recommended daily fluid intake
is _____ litre(s) or _____
8-ounce (250 ml) glasses. ”

How can I calculate the amount of fluid I drink per day?

Use a pitcher or measuring cup that can hold at least 2 litres of liquid. For each amount of liquid you consume, transfer the equivalent amount of water into your pitcher. It will make it easier for you to see what you are consuming.

Record your amount of liquid consumed in a notebook.

Equivalent liquid measurements

Ounces (oz)	Millilitres (ml)	Cups (c.)
1 oz (1 ice cube)	30 ml	2 tbsp.
8 oz	250 ml	1 c.
50 oz	1,500 ml (1.5 L)	6 c.
67 oz	2,000 ml (2 L)	8 c.

Some tips to relieve thirst

- Add lemon to your water to quench your thirst.
- Brush your teeth after meals and before going to bed.
- Freeze grapes or berries and suck on one when you are thirsty.
- Control the temperature in your home to avoid overheating it and maintain adequate humidity (30% to 50%).
- Avoid irritating beverages or liquids like coffee or mouthwash containing alcohol.
- Chew gum or suck on a lemon-flavoured sugarless candy, which will help you produce saliva.
- Take 2 to 3 drops of olive oil as needed to lubricate your mouth.
- Rinse your mouth with water when it's dry or suck on an ice cube.
- Sip your liquids slowly.

If you're still bothered by a dry mouth, medication can be prescribed for you. Consult your pharmacist.

Should I watch my weight?

People living with PAH are at greater risk of water retention. This fluid retention can alter your weight. A change in your weight can therefore indicate water retention caused by the disease. You should monitor your weight regularly.

- If you tend to gain weight, weigh yourself once a day when you wake up. Do this after urinating. Do not wear shoes. Note your weight on your calendar.
- If your weight is stable, weigh yourself once a week. Note your weight on your calendar.

Sudden weight gain may be caused by a deterioration in your health.



Caution

If your weight increases by 1.5 kg (3 lb) in less than 48 hours or by 2.5 kg (5 lb) in one week, notify the PAH clinic.

Here are a few tips to help you maintain a healthy weight:

- Eat three balanced meals per day.
- Have a source of protein at each meal.
- Eat fresh fruit and vegetables.
- Choose whole-grain carbohydrates.
- Decrease your meal portion size.
- Eat slowly.
- Eat when you really feel hungry.
- Limit snacks in the late evening or when you're not hungry.
- Avoid eating when you're stressed or bored.
- Find fun activities to do.
- Choose products low in fat.
- Avoid juices and sweetened beverages.
- Consult a nutritionist.

Diet

To control your weight, it is important to combine a healthy diet with physical activity.

- Cook a variety of dishes.
- If your condition allows, walk instead of using a car.
- Climb the stairs at your own pace rather than taking the elevator.
- Do your household tasks according to your ability.
- Exercise as much as you can tolerate.

“ My current weight _____ ”
“ My target weight _____ ”

Tips for stimulating your appetite

Many people with PAH experience loss of appetite or stomach pain. A few reasons may explain this condition: nausea, diarrhea, shortness of breath, fatigue, progression of PAH and certain medications.

Here are a few tips to help you if you experience loss of appetite:

- Relax before meals.
- Create a relaxing and pleasant atmosphere at mealtimes.
- Eat small meals at the same time each day.
- Take small bites.
- Eat the biggest meal of the day when your appetite is strongest.
- Add healthy, protein-rich snacks (e.g. yogurt, peanut butter, nuts).
- Choose colourful foods and foods you like.
- Choose foods with a soft texture. They require less effort to chew.
- Avoid drinking while eating. Instead, drink after the meal.

Don't forget to watch your daily allowed fluid limit. Discuss meal supplements such as Boost or Ensure with your nutritionist. To consult a nutritionist, ask your health care team or CLSC.

Exercices

Can I exercise despite my PAH?

It is recommended to do regular physical exercise. Exercise improves your quality of life as well as your psychological and physical health. A physical health assessment and the advice of your doctor are necessary before starting an exercise program.

Combining exercise and specific treatments improves tolerance to exertion. Low-intensity activities like walking, shopping, gardening, golf and walking your dog are beneficial. Adapt the exercise to your abilities and follow the advice of a health professional.

Respiratory rehabilitation

Respiratory rehabilitation is a program aimed at improving your quality of life through education and training. The exercise program is adapted to your abilities and goals. It is done under professional supervision. Participation in such a program requires a referral from a health professional.

Your treatment team will decide if respiratory rehabilitation is right for you. Feel free to talk to them if this interests you.

What are the risks associated with exercising?

For some people, exercise can cause:

- A heart rhythm disorder
- Dizziness
- Worsening of shortness of breath
- Loss of consciousness

If such symptoms appear, stop the physical activity and quickly discuss the situation with your health care team.

Exercices



Some practical advice

- Choose an activity that you enjoy.
- Start your activity slowly and gradually increase intensity.
- Do your physical activity at least 3 to 4 times per week.
- Plan to be accompanied when swimming or hiking in the mountains.
- Schedule breaks.
- Respect your abilities: stop before you are short of breath or exhausted.



Environments and activities to avoid

- Avoid altitude.
- Avoid exercising during hot and humid periods. Instead, do your activity in an air-conditioned place (e.g. walk in an air-conditioned shopping mall).
- Avoid doing activities that require intense physical exertion (e.g. running, tennis, hockey).
- Avoid exercises that require holding your breath (e.g. lifting heavy loads, digging, shovelling).
- Avoid contact sports due to the risk of injury and bleeding.



Medical follow-up

What will my follow-up involve at the PAH clinic?

Your doctor will decide on the frequency of your visits according to the severity and stability of your disease. At each visit, tell us about any medical change or follow-up (medications, upcoming surgery, etc.).

Tests and examinations

A 6 minute walk test and a pulmonary X-ray are generally done at each visit. This helps your PAH specialist evaluate the progression of the disease and check the effectiveness of the treatment. They may then decide to add certain additional tests, for example:

- Right heart catheterization (Swan-Ganz)
- respiratory function tests
- cardiac ultrasound
- blood tests

Blood tests

Blood tests allow your doctor to monitor your tolerance of the medication and your heart's functioning (e.g. NT-proBNP test).



Reminder

The NT-proBNP test (N-terminal pro-Brain Natriuretic Peptide) is the measurement of a hormone secreted by the heart which is used to diagnose and measure the severity of heart failure. This test helps monitor the response to treatment and progression of the disease.

Medical follow-up

Meeting with the nurse

During your visits or telephone calls to the PAH clinic, you will speak with a nurse. Together with your PAH specialist, the nurse will assess your physical and psychological health. They will educate you and answer your questions to help you better understand your disease and treatment. The nurse coordinates your care with the hospital department and communicates with pharmacies.

The nurse can also refer you to various health professionals or relevant community resources, for example:

- home assistance needs assessment
- oxygen therapy equipment
- nursing care at home
- social worker
- psychologist

“ Feel free to contact the clinic nurse if you have questions about your health. ”



Consultation at another hospital

If you have to go to a hospital other than the one where your PAH is being monitored, the doctor you see may have little knowledge about your disease.

If this occurs, this doctor should contact one of the specialists at the PAH clinic.

Medical follow-up

When should I contact the PAH clinic?

Contact the PAH clinic if you experience:

- unusual shortness of breath (during your usual activities)
- shortness of breath when lying down or which prevents you from lying down.
- persistent fatigue for more than one week with no other symptom.
- more frequent shortness of breath.
- loss of consciousness.
- fever with a temperature higher than 38°C (100.4°F)
- appearance of or increase in swelling of the ankles, legs, abdomen or face
- coughing up blood
- a weight gain of more than 1.5 kg (3 lb) in less than 48 hours or 2.5 kg (5 lb) in one week



Do you have a central or subcutaneous catheter?

Quickly contact the PAH clinic if the insertion site is:

- abnormally red
- painful
- warm
- oozing, with or without odour

Call **911** immediately if your central catheter is damaged (Caripul infusion).

Medical follow-up



When should I go to the emergency department?

Go immediately to the **emergency department** of the hospital closest to you if you experience any of these signs or symptoms:

- rapid deterioration in your shortness of breath
- chest pain
- coughing up large amounts of blood

Your condition requires immediate attention. Ask a loved one to accompany you or call the emergency department by dialing **911**.

Help and resources

Whom can I contact if I have questions?

Always have the telephone numbers of your treatment team at hand.

PAH program nurse clinician

Questions about your health status, your disease or your treatment.

Voicemail: _____

Fax: _____

Email: _____

Pager or cell phone: _____

PAH clinic

Questions about your appointment.

Telephone: _____

Community pharmacy

Telephone: _____

Fax: _____

Specialized pharmacy

Telephone: _____

Fax: _____

Support program nursing staff

Telephone: _____

Help and resources



Useful resources

Associations

Quebec

Association pulmonaire du Québec [Quebec Lung Association] poumonquebec.ca/en

Fondation HTAPQ [Quebec Pulmonary Arterial Hypertension Association] htapquebec.ca

Regroupement québécois des maladies orphelines [Quebec Coalition for Rare Disorders] rqmo.org/en

Canada

Pulmonary Hypertension Association of Canada phacanada.ca

Heart and Stroke Foundation of Canada heartandstroke.ca

Worldwide

European pulmonary hypertension association (Europe) phaeurope.org

Pulmonary Hypertension Association (United States) phassociation.org

Association des malades, aidants et transplantés hypertension pulmonaire [Association of patients, caregivers and transplant recipients with pulmonary hypertension] (France) htapfrance.com

Diet

Heart and Stroke Foundation of Canada heartandstroke.ca/healthy-living/recipes
Low-sodium recipes

Contraception and adoption

Action Canada for Sexual Health & Rights actioncanadashr.org

Ministère de la Santé et des Services Sociaux: Secrétariat à l'adoption internationale [Ministry of Health and Social Services: International Adoption Office] adoption.gouv.qc.ca/en

Official site on international adoption in Quebec

Help and resources



Useful resources

Reading for children (comic book)

Qu'arrive-t-il à la maman de Pierre ? MEDIKIDZ expliquent l'hypertension artérielle pulmonaire (in French only).

Dr Kim Chilman-Blair et Dr Columba Quigley, 2016, Londres : Medikidz limited.LTD.

IUCPQ-ULaval

There are other educational documents produced by the Institute. You can view them on our website:

bibliotheque-patients.iucpq.qc.ca.



The following documents may interest you (in French only):

- [*Mieux connaître le médicament bosentan \(Tracleer\) : hypertension artérielle pulmonaire*](#)
- [*Mieux connaître le médicament macitentan \(Opsumit\) : hypertension artérielle pulmonaire*](#)
- [*Mieux connaître le médicament ambrisentan \(Volibris\) : hypertension artérielle pulmonaire*](#)
- [*Mieux connaître les médicaments tadalafil \(Adcirca\) et sildénafil \(Revatio\) : hypertension artérielle pulmonaire*](#)
- [*Mieux connaître le médicament riociguat \(Adempas\) : hypertension artérielle pulmonaire*](#)
- [*Mieux connaître le médicament selexipag \(Uptravi\) : hypertension artérielle pulmonaire*](#)
- [*Mieux connaître le médicament époprosténol \(Flolan ou Caripul\) : hypertension artérielle pulmonaire*](#)
- [*Mieux connaître le médicament tréprostinil \(Remodulin\) : hypertension artérielle pulmonaire*](#)

Enter the title of the document you want to consult into the search field.

Glossary

6-minute walk test: involves walking the longest distance possible for a period of 6 minutes. Helps measure functional capacity and exertion tolerance.

Autoimmune disease: immune system dysfunction. Certain cells of the body attack the organs as though they were foreign bodies.

Cardiac ultrasound: medical imaging technique that uses ultrasound to visualize all of the heart structures (atria, ventricles, valves, muscle).

CIUSSS: *Centre intégré universitaire de santé et de services sociaux* [Integrated University Health and Social Services Centre].

CLSC: *Centre local de services communautaires* [Local Community Services Centre].

HIV: human immunodeficiency virus. A virus that attacks the immune system of the human body. The person's immune system becomes weaker and they become more at risk of contracting other diseases. When the person is no longer able to defend themselves from infections, it is called acquired immunodeficiency syndrome (AIDS).

Left heart diseases: heart diseases affecting the left side of the heart (e.g. valve problems, stenosis).

Lupus: chronic autoimmune disease that can attack the skin of the face (wolf mask or butterfly rash), but also the joints (arthritis) and other organs (e.g. lungs, heart, kidneys).

New York Heart Association Functional Classification: Classification of the severity of pulmonary hypertension symptoms.

NT-proBNP blood test (N-terminal pro-Brain Natriuretic Peptide): screening for a hormone secreted by the heart that is used to diagnose and measure the severity of heart failure. This test makes it possible to monitor the response to treatment and progression of the disease.

PAH: pulmonary arterial hypertension.

Glossary

Pulmonary radiography: helps obtain images of the heart and lungs using X-rays.

RAMQ: Quebec health insurance plan.

Respiratory function test: helps measure the amount of air the lungs can hold and the ability to expel air from the lungs.

Right heart catheterization (Swan-Ganz): introduction of a catheter into a blood vessel to explore the right heart chambers and measure the pressure in the right atrium, right ventricle, pulmonary artery and pulmonary capillaries. The oxygen saturation of the blood is also measured.

Right heart failure: inability of the right side of the heart to efficiently pump blood to the lungs to meet the body's needs. It is characterized by a decrease in the heart's capacity for exertion.

Scleroderma: chronic autoimmune disease characterized by excessive collagen production and damage to the small blood vessels. It causes excessive scarring within various organs. This imbalance causes the skin to harden and the affected organs to change.

Veno-occlusive disease: rare form of pulmonary hypertension that affects the small veins and capillaries of the pulmonary circulation system.

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