

PH • Pulmonary Hypertension

A plain-English guide for you and your family

WHAT IS HAPPENING

Pulmonary high blood pressure (hypertension) means the pressure inside the arteries of your lungs is too high. Normally those arteries are wide and soft, so blood flows easily from the right side of your heart through your lungs to pick up oxygen. In PH, something narrows or blocks those lung arteries — it might be thick, stiff artery walls (type 1, called PAH), backup from a failing left heart (type 2), damage from lung diseases like COPD or sleep pauses in breathing (apnea) (type 3), clots stuck in the vessels (type 4), or a mix of causes (type 5). Whatever the cause, the right side of your heart now has to squeeze much harder to push blood through that high-pressure pipe. At first you feel short of breath when you walk or climb stairs, you tire easily, and you might feel dizzy or faint because not enough oxygen-rich blood reaches your muscles and brain. Over months or years, the overworked right heart muscle stretches and weakens (right heart failure), so blood backs up into your body's veins: your neck veins bulge, your belly swells and hurts (because your liver fills with backed-up blood), and your ankles and legs puff up with fluid. None of these signs is random — each one is your body's attempt to keep blood moving and oxygen flowing. The key is to find out which of the five types you have, because the treatment is completely different for each: type 1 gets lung-artery medicines, type 2 needs better left-heart control, type 3 needs oxygen and lung care, type 4 may need clot removal, and type 5 tackles the mix. Caught early and treated right, many people live longer and feel much better.

What you might feel

- Shortness of breath during activity
- Feeling tired or weak all the time
- Chest discomfort or pressure
- Swelling in your ankles, legs, or belly
- Your neck veins bulge or stand out
- Bluish lips or skin (cyanosis)
- Feeling dizzy or fainting (syncope)

- A loud second heart sound (P2) when the doctor listens with a stethoscope
- The right side of your heart gets bigger (right ventricular enlargement)

Call the doctor right away if...

- Sudden fainting or near-fainting during exercise—this can mean your heart is at its limit and cannot send enough blood to your brain.
- New or worsening leg swelling, bulging neck veins, or a swollen belly—these are signs that the right heart is failing and blood is backing up into your body.
- Blue lips, fingernails, or skin—this means your blood oxygen is dangerously low.
- Chest pain that feels like pressure or squeezing, especially with activity—the struggling right heart may not be getting enough oxygen itself.
- Coughing up blood (hemoptysis)—high lung pressure can burst small blood vessels in the airways.

What the care team will do

- Monitor oxygen saturation continuously and keep SpO2 above 90 percent with supplemental oxygen as ordered
- Watch for signs of right heart failure: measure jugular venous distension (how high neck veins sit), check for new or worsening leg swelling, listen for extra heart sounds, and report immediately if patient becomes confused or blood pressure drops
- Give diuretics (water pills like furosemide) exactly on schedule and track daily weights, intake and output, and electrolytes (sodium, potassium)
- Avoid giving too much intravenous fluid; give fluid boluses only when ordered and monitor closely for worsening shortness of breath or dropping oxygen levels
- Teach the patient to pace activities, rest between tasks, and call for help instead of straining (like during bowel movements), and give stool softeners as ordered
- Administer pulmonary vasodilator medications (sildenafil, tadalafil, bosentan, ambrisentan, macitentan, epoprostenol) on time and through the correct route, and never stop them suddenly
- Monitor blood pressure closely, especially after giving vasodilator medications, and report systolic BP below 90 or symptoms of dizziness and lightheadedness
- Coordinate care so the patient gets an echocardiogram (heart ultrasound), right heart catheterization if not done yet, and specialty referral to a pulmonary high blood pressure (hypertension) expert center

Medicines you may be given — and why

Sildenafil (Revatio)

Sildenafil blocks an enzyme (PDE5) that breaks down a helpful molecule called cGMP. When cGMP stays around longer, the smooth muscle inside the lung arteries relaxes and the vessels open wider. That lowers the pressure and makes it easier for the heart to push blood through the lungs.

Tadalafil (Adcirca)

Tadalafil blocks the same enzyme (PDE5) that sildenafil does, but it lasts longer in your body—about 24 hours instead of 4 to 6. It keeps cGMP levels up so the lung arteries stay relaxed and open all day. That lowers the pressure the right side of your heart has to push against.

Bosentan (Tracleer)

Bosentan blocks receptors for endothelin, a powerful chemical that tightens lung arteries and makes their walls thicker. By blocking both endothelin receptors (ET-A and ET-B), bosentan stops that squeezing and thickening, so the arteries stay more open and the pressure drops.

Ambrisentan (Letairis)

Ambrisentan blocks only the ET-A receptor, the one that squeezes lung arteries and tells the wall to thicken. It leaves the ET-B receptor alone, which may help your body clear extra endothelin. By stopping the squeeze signal, ambrisentan lowers lung artery pressure.

Macitentan (Opsumit)

Macitentan blocks both ET-A and ET-B receptors for endothelin, stopping the signal that makes lung arteries squeeze and thicken. It binds very tightly to the receptors and gets into the tissue well, so it keeps working even as your body tries to make more endothelin.

Epoprostenol (Flolan, Veletri)

Epoprostenol is a man-made version of prostacyclin, a natural chemical your body makes to open blood vessels and stop platelets from clumping. In PAH, your lungs do not make enough prostacyclin. Epoprostenol replaces it, relaxing the lung arteries and lowering pressure. It also stops the vessel walls from getting thicker.

Treprostinil (Remodulin, Tyvaso, Orenitram)

Treprostinil works just like epoprostenol—it mimics prostacyclin to relax lung arteries, lower pressure, and stop platelets from clumping. It lasts longer in the body, so you can give it under the skin (subcutaneous), through an IV, by breathing it in (inhaled), or as a pill, depending on the form.

Iloprost (Ventavis)

Iloprost is a prostacyclin mimic that you breathe in as a mist six to nine times a day. It goes straight to the lung arteries, relaxes the muscle, and opens the vessels. By delivering the medicine directly to the lungs, it lowers pressure there with less effect on the rest of the body.

Selexipag (Uptravi)

Selexipag is a pill that you take twice a day. It is NOT a prostacyclin itself, but it turns into a molecule that locks onto prostacyclin receptors and mimics what prostacyclin does—relax lung arteries and lower pressure. Because it is not an actual prostacyclin, it lasts longer and is more stable.

Riociguat (Adempas)

Riociguat boosts an enzyme called soluble guanylate cyclase, which makes cGMP—the same relaxing molecule that PDE5 inhibitors protect. Riociguat works in two ways: it makes the enzyme more sensitive to nitric oxide AND it directly turns on the enzyme even when nitric oxide is low. That raises cGMP, relaxes lung arteries, and lowers pressure.

Calcium-channel blockers (CCBs): Nifedipine, Diltiazem, Amlodipine

CCBs block calcium from entering the muscle cells in artery walls. Without calcium, the muscle cannot squeeze. That keeps the lung arteries relaxed and open. CCBs work ONLY in a small group of PAH patients whose arteries open up during a vasodilator test (acute vasoreactivity test) at heart catheterization. For those few, CCBs can lower pressure safely and long-term.

Oxygen (supplemental O₂)

Oxygen is NOT a drug, but breathing extra oxygen helps when your blood oxygen level is low (hypoxemia). Low oxygen makes lung arteries squeeze tight (hypoxic vasoconstriction), which raises pressure even more. By keeping oxygen levels normal, you prevent that extra squeezing and take some load off the right heart. Oxygen is especially important in Group 3 pulmonary high blood pressure (hypertension) (lung disease) and at high altitude.

Diuretics (water pills): Furosemide (Lasix), Bumetanide, Torsemide, Spironolactone

Diuretics help your kidneys dump extra salt and water into the urine. In pulmonary high blood pressure (hypertension), the right heart can get weak and blood backs up, causing fluid to leak into the lungs, belly, and legs (edema, fluid buildup in the belly (ascites)). Diuretics pull that extra fluid out, which makes breathing easier and takes pressure off the heart. They do NOT lower pulmonary artery pressure directly.

Anticoagulation (blood thinners): Warfarin, Apixaban, Rivaroxaban

Blood thinners do NOT dissolve clots, but they stop new clots from forming. In CTEPH (Group 4), clots are stuck in the lung arteries. In PAH (Group 1), slow blood flow and thick vessel walls raise clot risk. Anticoagulants keep the blood from clotting as easily, which may prevent more clots from piling on and making things worse. Warfarin has been used traditionally; newer agents (apixaban, rivaroxaban) are being studied.

Where this information comes from

- ESC/ERS Guidelines for Pulmonary Hypertension 2022
- AHA/ACC Pulmonary Hypertension Scientific Statement
- Chest (Journal of the American College of Chest Physicians)
- World Health Organization Pulmonary Hypertension Classification

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