

PH · Pulmonary Hypertension

PH diagnosis needs right heart catheter to measure lung artery pressure directly; type 1 (PAH) gets special medicines and rarely calcium-channel blockers, but only if a vasoreactivity test proves they work; every other type treats the root cause first.

THE ONE-LINE TRUTH

Five types by cause: blood vessel wall thickening (type 1), left heart backup (type 2), lung disease (type 3), clot blockage (type 4), or mixed (type 5) — all raise pressure in the lung arteries and wear out the right heart.

The chain — how it unfolds

1. High pressure in the lung arteries forces the right ventricle to squeeze much harder with every beat. — why: Whether from thick artery walls (type 1), backup from left heart failure (type 2), oxygen-starved vessel clamping (type 3), clot blockage (type 4), or mixed causes (type 5), the right heart now faces a brick wall instead of an open road, so it has to work like a marathon runner every single minute.
2. The right ventricle muscle thickens at first, then stretches and weakens, causing right heart failure. — why: Constant overwork makes the muscle bulk up (right ventricular hypertrophy), but eventually the wall thins out, the chamber balloons (dilation), and the pump loses strength — blood can no longer move forward efficiently.
3. Blood backs up into the body's veins, making neck veins bulge, the liver swell and hurt, and legs and belly puff up with fluid. — why: The failing right heart cannot empty itself, so pressure rises backward through the right atrium into the superior and inferior vena cava (the big veins draining your body); high vein pressure pushes fluid out of blood vessels into tissues (edema) and engorges the liver (hepatomegaly, right-upper-quadrant pain).
4. You feel short of breath, especially when walking or climbing stairs, because your muscles and brain are starved for oxygen. — why: The weakened right heart pumps less blood through the lungs, so less oxygen gets picked up and delivered to your body; when you exercise, demand spikes but supply cannot keep up, triggering air hunger (dyspnea) and fatigue.

5. You may feel dizzy, lightheaded, or faint (syncope), especially during effort or standing up quickly. — why: Low cardiac output means less oxygen-rich blood reaches your brain; the brain is extremely sensitive to oxygen drops, so even a brief dip causes dizziness or fainting — this is a red flag for severe PH and high risk of sudden death.
6. Chest discomfort or pressure can occur, mimicking angina, because the thickened right ventricle outgrows its own blood supply. — why: The right coronary artery feeds the right ventricle, but when the muscle wall thickens dramatically, the existing blood vessels cannot deliver enough oxygen to the deeper layers, causing starved blood flow (ischemia) (oxygen debt) and chest pain, especially during exertion.
7. Without treatment, the right heart fails completely, organs shut down, and sudden death from arrhythmia or shock is common. — why: End-stage right heart failure means almost no blood moves forward; the brain, kidneys, and liver fail from low oxygen and buildup of waste; the overstretched heart muscle becomes electrically unstable, triggering fatal rhythms like ventricular a fast heart rate (tachycardia) or fibrillation.
8. Every visible sign — loud P2 heart sound, swollen neck veins, enlarged liver, leg swelling (edema), low oxygen on pulse ox — traces back to the same root: high lung artery pressure breaking the right heart. — why: The loud P2 is the pulmonary valve slamming shut under high pressure; swollen veins and swelling (edema) are backup from right heart failure; low oxygen is poor forward flow through the lungs — none are separate diseases, all are dominoes falling from the same upstream break.

Spot it — what you see, and why

- Shortness of breath during activity: High pressure in the lung arteries makes it harder for blood to pick up oxygen, so you feel out of breath when your body needs more oxygen—like when walking up stairs or carrying groceries.
- Feeling tired or weak all the time: Your heart has to pump extra hard against the stiff, tight arteries in your lungs, so less oxygen-rich blood reaches your muscles and organs, leaving you feeling drained even after rest.
- Chest discomfort or pressure: The right side of your heart is working much harder than usual to push blood through high-pressure lung vessels, and that extra strain can cause a heavy or tight feeling in your chest.
- Swelling in your ankles, legs, or belly: When the right heart cannot pump blood forward efficiently, blood backs up into the veins; fluid then leaks out into the tissues, causing puffiness (edema) in the legs, ankles, and sometimes the abdomen (ascites).
- Your neck veins bulge or stand out: Blood is backing up from the struggling right heart into the large veins that drain into it, raising the pressure in the jugular veins (the big veins in your neck) so they look swollen or raised.

- Bluish lips or skin (cyanosis): When the lungs cannot load enough oxygen into the blood because of the high pressure and poor flow, the blood stays darker and gives a blue tint to lips, fingernails, or skin.

Red flags — act now

- 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.

Treat it — in this order

1. Right heart catheterization to confirm the diagnosis and measure exact pressures → Gives you the real mean pulmonary artery pressure (mPAP) and pulmonary artery wedge pressure (PAWP) so you know which of the five groups the patient has and how severe it is. This number decides whether the patient gets pulmonary vasodilators (drugs that open lung blood vessels) or needs a different treatment.
2. PDE5 inhibitor (sildenafil 20 mg three times daily or tadalafil 40 mg once daily) or endothelin receptor antagonist (ambrisentan 5–10 mg daily, bosentan 125 mg twice daily, or macitentan 10 mg daily) for Group 1 pulmonary arterial high blood pressure (hypertension) → Relaxes and opens the thick, stiff blood vessels in the lungs by blocking signals that make them squeeze tight (endothelin) or by boosting signals that make them relax (nitric oxide and cGMP). This lowers pulmonary artery pressure and lets the right heart pump more easily.
3. Treat the upstream cause: diuretics and GDMT for left heart failure (Group 2), oxygen and inhalers for COPD or interstitial lung disease (Group 3), anticoagulation and surgery for chronic clots (Group 4) → Stops blood from backing up into the lungs (Group 2 left heart failure), restores oxygen and opens airways (Group 3 lung disease), or removes the clot dam blocking blood flow (Group 4 CTEPH). Fixing the root cause lowers pulmonary pressure naturally.

4. Loop diuretic (furosemide 20–80 mg daily, titrated to symptoms) for fluid overload and right heart failure → Removes extra fluid that backs up when the right heart fails, so the patient can breathe easier, neck veins go down, and swelling improves. This takes load off the right heart.
5. Supplemental oxygen (target oxygen saturation 90 percent or higher) for patients with low oxygen in the blood (hypoxemia) → Low oxygen makes pulmonary blood vessels squeeze tight (hypoxic vasoconstriction). Giving oxygen relaxes them a little and lowers pulmonary pressure. This helps the right heart work less hard.

Your top priorities at the bedside

1. Monitor oxygen saturation continuously and keep SpO₂ above 90 percent with supplemental oxygen as ordered — why: Low oxygen (hypoxemia) makes pulmonary arteries squeeze tighter (hypoxic vasoconstriction), which raises pressure in the lungs and forces the already-weak right heart to work even harder.
2. 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 — why: Right ventricular failure happens when the right heart cannot pump against high lung pressures anymore; blood backs up into the body causing organ damage, and forward flow drops causing shock and death.
3. Give diuretics (water pills like furosemide) exactly on schedule and track daily weights, intake and output, and electrolytes (sodium, potassium) — why: Fluid overload from a failing right heart causes dangerous swelling in the liver, belly, and legs; diuretics pull off extra fluid but can cause low potassium (hypokalemia) and kidney injury if we give too much.