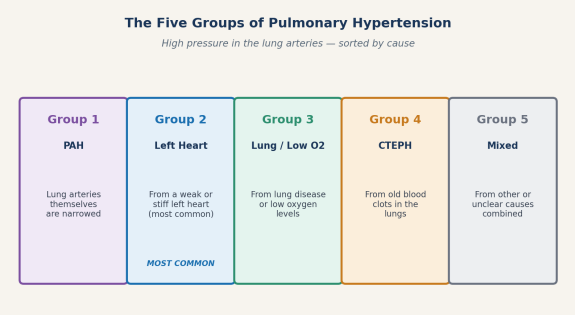
Understanding Pulmonary Hypertension

High Blood Pressure in the Arteries of Your Lungs

Five different causes, five different paths — finding yours is the key.



Online: https://go.riasalimd.com/ph-guide

Rias KS Ali, MD FACC

Board Certified in Interventional Cardiology & Cardiovascular Disease

4740 Mile Stretch Drive, Holiday FL 34690 | Office: (727) 943-5200 | Fax: (727) 943-5201 | RiasAliMD.com

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Names & Terms You Will Hear

Term	Meaning
Pulmonary hypertension (PH)	High blood pressure in the arteries that carry blood from the heart to the lungs.
Pulmonary arterial hypertension (PAH)	Group 1 PH. The small lung arteries themselves narrow and stiffen. Rare, but it has its own medicines.
Mean pulmonary artery pressure (mPAP)	The average pressure in the lung arteries. PH is a value over 20 mmHg, measured by a right heart catheter.
Right heart catheterization (RHC)	The test that confirms PH and measures the true pressure. The gold standard.
Cor pulmonale	Right-heart strain or failure caused by lung problems, including long-standing PH.
CTEPH	Chronic thromboembolic PH (Group 4) — PH caused by old blood clots in the lungs. Often curable.
Pulmonary vascular resistance (PVR)	How hard the heart must push to move blood through the lungs. Higher numbers mean stiffer lung arteries.
Idiopathic PAH (IPAH)	Group 1 PH with no known cause found. 'Idiopathic' means the cause is unknown.

Call 911 if you have severe shortness of breath, chest pain, fainting, or you cough up blood. PH can strain the heart quickly. **Do not stop** your PH medicines or blood thinners on your own — stopping suddenly can be dangerous.

What Is Pulmonary Hypertension?

- Pulmonary hypertension (PH) is high blood pressure in the arteries that carry blood from the heart to the lungs. It is different from the usual blood pressure measured on your arm.
- PH is defined as a mean pulmonary artery pressure over 20 mmHg at rest. A normal value is about 14 mmHg.
- These lung arteries are normally a low-pressure system. When they narrow, stiffen, or get blocked, the pressure rises.
- The right side of the heart must then push harder to move blood through the lungs. Over time this strains and weakens the right heart — this is the main danger.
- PH is sorted into five groups by its cause. The group decides the treatment, so finding the right group is the key first step.
- **Group 2** — PH from left-heart disease — is by far the most common cause. **Group 1 (PAH)** is rare but has special medicines.
- PH can be mild or severe. Some causes can be treated or even cured. Others are managed to slow the disease and ease symptoms.

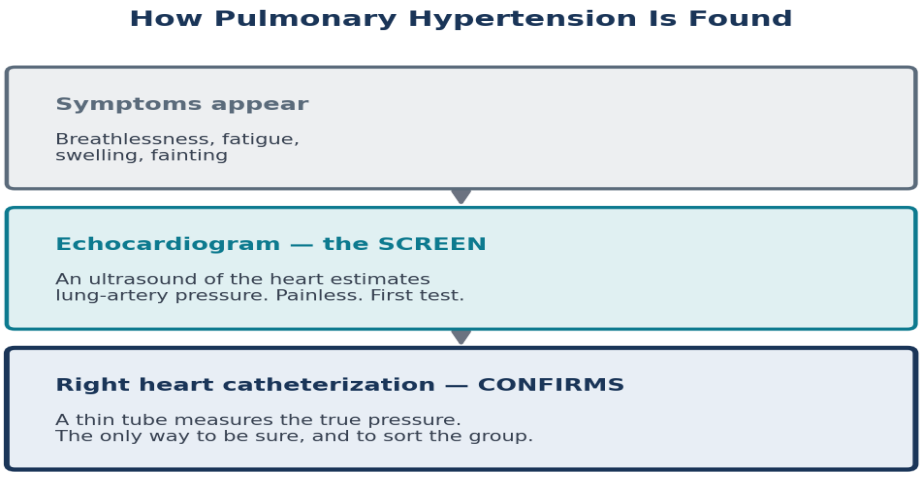
The Five Groups of Pulmonary Hypertension — Cause and Main Treatment

Group	Main Cause	Main Treatment
1 — PAH	The small lung arteries themselves narrow and stiffen (idiopathic, genetic, drugs, connective-tissue disease).	PAH-specific medicines that open the lung arteries; transplant if severe.
2 — Left Heart	A weak or stiff left heart, or a heart-valve problem, backs pressure into the lungs. The most common cause.	Treat the left-heart disease or valve. PAH drugs are not used.
3 — Lung / Low O2	Lung disease (COPD, fibrosis) or low oxygen (including sleep apnea) tightens lung arteries.	Treat the lung disease; oxygen; treat sleep apnea.
4 — CTEPH	Old blood clots scar and block the lung arteries.	Surgery or balloon treatment — can often be cured; lifelong blood thinners.
5 — Mixed	Several or unclear causes combined (blood, metabolic, or other disorders).	Treat each underlying cause that is found.

The group decides the treatment. The same medicine can help one group and harm another. That is why a right heart catheterization confirms the diagnosis and the group before any specific therapy begins.

Why It Matters

- PH makes the right side of the heart work much harder. Untreated, this can lead to right-heart failure — the most serious complication.
- Symptoms often start slowly and are easy to blame on aging or being out of shape. This delays diagnosis, sometimes by years.
- The group matters enormously. Group 1 (PAH) needs special medicines. Group 2 needs the left heart treated. Group 4 (CTEPH) may be cured with surgery.
- An echocardiogram can screen for PH, but only a right heart catheterization can confirm it and measure the true pressure. See our Right Heart Catheterization guide.
- Group 4 (CTEPH) is special — it comes from old blood clots and can often be cured. Many cases are missed. Anyone with PH should be checked for it with a special lung scan.
- Early diagnosis and care at a PH center improve outcomes. PAH medicines and CTEPH surgery have changed what is possible.
- Pregnancy is very high risk with PH, especially PAH. Reliable birth control is important. Discuss any pregnancy plans with your team.



How PH is found: symptoms prompt an echocardiogram (the painless screening test), and a right heart catheterization confirms the diagnosis and measures the true lung-artery pressure. The catheter is also what sorts PH into its group.

How Serious Is It? — A Risk Snapshot

Lower Risk	Intermediate Risk	Higher Risk
• Few or no symptoms with daily activity	• Breathless with moderate effort	• Symptoms at rest or with light effort
• Right heart still pumping well	• Some signs of right-heart strain	• Fainting, or right-heart failure signs
• Good walking distance on testing	• Shorter walking distance	• Right heart enlarged and struggling
• Goal: keep it here with treatment	• Treatment is often stepped up	• Needs intensive, specialist-led care

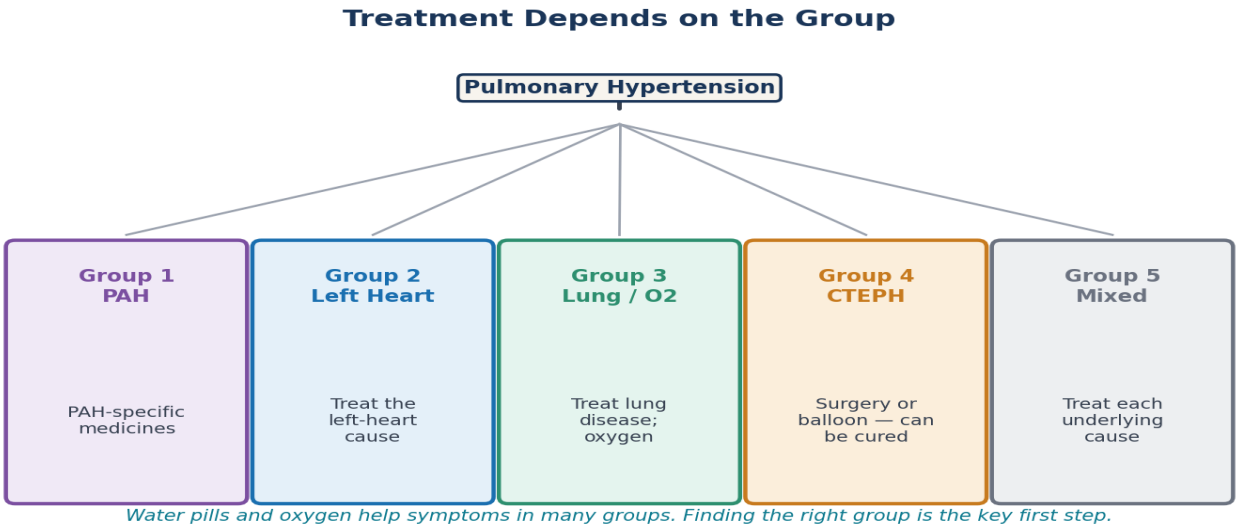
Risk Factors

Some patients are more likely to develop this condition. Knowing your risk helps your care team take extra precautions.

Risk Factor	Why It Increases Risk
Left-heart disease (Group 2)	A weak or stiff left heart, or a leaky/tight heart valve, backs pressure up into the lungs. This is the most common cause of PH.
Lung disease or low oxygen (Group 3)	COPD, pulmonary fibrosis, and sleep apnea lower oxygen and tighten lung arteries over time. See our Sleep Apnea guide.
Past blood clots in the lungs (Group 4 / CTEPH)	Clots that do not fully dissolve can scar and block lung arteries. See our Pulmonary Embolism guide.
Connective tissue disease (Group 1)	Scleroderma and lupus raise the risk of PAH. People with these conditions should be screened.
Certain drugs and toxins (Group 1)	Some diet pills (like fen-phen), methamphetamine, and a few other drugs can trigger PAH.
Congenital heart disease (Group 1)	A heart defect present from birth can push extra blood and pressure into the lungs over many years.
Liver disease and HIV (Group 1)	Advanced liver disease (portal hypertension) and HIV infection both raise the risk of PAH.
Family history or gene changes (Group 1)	Some PAH runs in families. A gene change (BMPR2) can be passed down. Relatives may be offered screening.

Treatment Options

- **Step 1 — Find the group.** Treatment depends entirely on the cause. A right heart catheter, lung scans, and blood tests sort PH into its group before any specific therapy starts.
- **Group 1 (PAH) — special medicines.** PAH has its own drugs that relax and open the lung arteries. The three main families are endothelin blockers (ambrisentan, macitentan), PDE5 inhibitors (sildenafil, tadalafil), and prostacyclins (epoprostenol, treprostinil). Many people take two or three together.
- **Group 2 (left heart) — treat the heart.** The best treatment is to treat the left-heart problem itself — heart failure or a valve issue. PAH-specific drugs are *not* used here and can be harmful. See our HFpEF guide.
- **Group 3 (lung / low oxygen) — treat the lungs.** Manage the lung disease, treat sleep apnea, and use oxygen if levels are low. Oxygen helps relax the lung arteries.
- **Group 4 (CTEPH) — can be cured.** If old clots are in reachable spots, surgery (pulmonary endarterectomy) can remove them and potentially cure the PH. If surgery is not possible, balloon pulmonary angioplasty opens the arteries instead. Lifelong blood thinners are also used.
- **Group 5 (mixed) — treat each cause.** This group has several or unclear causes. Care is tailored to each problem found.
- **Support treatments for many groups.** Water pills (diuretics) ease swelling, oxygen helps when levels are low, and gentle supervised exercise can improve stamina.
- **When PH is severe.** If the right heart is failing despite treatment, options include advanced PAH drug combinations or, in select Group 1 patients, lung or heart-lung transplant. See our Advanced Heart Failure guide.



Treatment branches by group. Group 1 (PAH) gets special lung-artery medicines; Groups 2 and 3 are treated by fixing the underlying heart or lung cause; Group 4 (CTEPH) can often be cured with surgery or balloon treatment; Group 5 is tailored to each cause.

Group 1 — Pulmonary Arterial Hypertension (PAH)

- **What it is:** the small arteries in the lungs themselves narrow, stiffen, and thicken. This is the rare type that has its own special medicines.
- **Causes:** often no cause is found (idiopathic). It can also come from genes, connective-tissue disease (scleroderma, lupus), HIV, liver disease, congenital heart defects, or certain drugs.
- **Special medicines:** three main families open the lung arteries — endothelin blockers (ambrisentan, macitentan), PDE5 inhibitors (sildenafil, tadalafil), and prostacyclins (epoprostenol, treprostinil). Two or three are often combined.
- **How they are given:** some are simple pills. The strongest prostacyclins are given through a small pump under the skin or into a vein, or breathed in.
- **Severe disease:** when medicines are not enough and the right heart is failing, a lung transplant or heart-lung transplant may be considered in select patients.
- **Why the group matters:** these medicines help Group 1 but can be harmful in Group 2 (left heart). That is why a right heart catheter confirms the group first.

Group 2 — From Left-Heart Disease (Most Common)

- **What it is:** the most common cause of PH. A weak or stiff left heart, or a leaky or tight heart valve, backs pressure up into the lungs.
- **Common sources:** heart failure with reduced or preserved pumping (HFrEF or HFpEF), and mitral or aortic valve disease.
- **The key treatment:** treat the left-heart problem itself. Better-controlled heart failure or a fixed valve lowers the lung pressure too.
- **What is NOT used:** PAH-specific medicines are generally avoided here. They can push more fluid into the lungs and cause harm.
- **Support measures:** water pills for fluid, salt limits, and treating high blood pressure, atrial fibrillation, and sleep apnea all help.
- See our HFpEF guide for stiff-heart failure, a very common driver of Group 2 PH.

Group 3 — From Lung Disease or Low Oxygen

- **What it is:** long-term lung disease or low oxygen makes the lung arteries tighten and the pressure rise.
- **Common sources:** COPD (emphysema), pulmonary fibrosis (scarred lungs), and obstructive sleep apnea, which lowers oxygen during sleep.
- **The key treatment:** treat the underlying lung disease as well as possible, and treat sleep apnea with CPAP if present.
- **Oxygen therapy:** if oxygen levels are low, supplemental oxygen relaxes the lung arteries and lowers strain on the right heart. Use it as prescribed, including at night.
- **Lifestyle:** quitting smoking, vaccines, and pulmonary rehab all protect the lungs and slow the disease.
- See our Sleep Apnea guide — treating apnea can improve Group 3 PH.

Group 4 — CTEPH (Old Blood Clots) — Often Curable

- **What it is:** chronic thromboembolic PH (CTEPH). Old blood clots in the lungs did not fully dissolve. They scar and block the lung arteries, raising the pressure.
- **Why it is special:** this is the one type of PH that can often be **cured**. Everyone diagnosed with PH should be screened for it, usually with a special lung scan (V/Q scan).
- **Surgery (pulmonary endarterectomy):** when the clots are in reachable spots, surgeons remove the old clot and scar. This can return lung pressure toward normal — a potential cure. It is done at expert centers.
- **Balloon treatment:** when surgery is not possible, balloon pulmonary angioplasty opens the narrowed arteries from the inside, over several sessions.
- **Medicine:** a drug called riociguat can help when CTEPH is inoperable or comes back after surgery.
- **Blood thinners for life:** people with CTEPH stay on anticoagulation to prevent new clots. See our Pulmonary Embolism guide.

Group 5 — Mixed or Unclear Causes

- **What it is:** PH from causes that do not fit the other four groups, or from several causes at once.
- **Examples:** blood disorders (such as sickle cell disease), sarcoidosis, certain metabolic and thyroid disorders, and kidney disease on dialysis.
- **The key treatment:** there is no single drug for Group 5. Care is aimed at each underlying condition that is found.
- **Why diagnosis is careful:** sorting out exactly which problems are driving the PH guides the treatment, so a full work-up matters.
- **Support measures:** oxygen, water pills, and treating any contributing heart or lung problem all help with symptoms.

Comfort Measures at Home (No Medication Needed)

These simple steps support healing and ease symptoms. Use them alongside any medication your doctor prescribes.

- Keep all follow-up visits and tests. PH is a long-term condition that needs regular checks to catch changes early.
- Get your flu, pneumonia, and COVID shots. A chest infection is much more dangerous when the lung arteries are under strain.
- Limit salt to ease swelling and fluid buildup. Read labels — packaged and restaurant foods hide the most salt.
- Weigh yourself a few times a week. A quick gain can mean fluid is building up. Tell us about a rise of 3 to 5 pounds.
- Stay gently active within your limits. Ask about a supervised pulmonary rehab or exercise program — pushing too hard can be harmful in PH, so get guidance first.
- If you use oxygen, use it as prescribed — including at night and with activity. It helps your heart as well as your breathing.
- Avoid high altitude and unpressurized flights without advice. Thin mountain air lowers oxygen and can worsen PH.
- Do not get pregnant without talking to your PH team first. Pregnancy is very high risk in PH, especially PAH. Use reliable birth control.
- Quit smoking and avoid secondhand smoke. Ask us for help — it is one of the most powerful steps for your lungs and heart.
- Tell every doctor and pharmacist you have PH. Some common drugs (certain decongestants and others) can be risky.

Risks, Benefits, and Alternatives

Every choice has trade-offs. Use this table to start the shared decision conversation.

Option	Risks	Benefits	Alternatives
PAH-Specific Medicines (Group 1)	Side effects: headache, flushing, leg swelling, low blood pressure. Some are pills; some need a pump or breathing device. Liver checks may be needed. They do not cure PAH.	Relax and open the lung arteries. Improve symptoms, walking distance, and survival. Combining two or three works better than one. The main treatment for Group 1.	Treating an underlying cause (Groups 2-4). Oxygen and water pills for symptoms. Transplant for severe, drug-resistant Group 1.
Pulmonary Endarterectomy (CTEPH Surgery)	Major open-chest surgery using a heart-lung machine. Done only at expert centers. Risks of bleeding, stroke, and lung injury. Not everyone's clots are reachable.	Can **cure** Group 4 (CTEPH) by removing old clot material. Often returns lung pressure toward normal. The treatment of choice when clots are surgically reachable.	Balloon pulmonary angioplasty if surgery is not possible. Lifelong blood thinners. PAH-type medicines for residual disease.
Balloon Pulmonary Angioplasty (CTEPH)	Done in several sessions. Risks include lung-vessel injury and bleeding. Does not remove clots — it opens narrowed spots. Needs an experienced team.	Opens blocked lung arteries with a balloon for CTEPH that surgery cannot reach. Lowers pressure and improves symptoms. A good option for inoperable or leftover disease.	Pulmonary endarterectomy surgery if clots are reachable. Lifelong blood thinners. Medicines (riociguat) for inoperable CTEPH.

Option	Risks	Benefits	Alternatives
Long-Term Oxygen Therapy	Tanks or a concentrator at home. Tubing limits movement somewhat. Does not treat the root cause of most PH.	Raises blood oxygen and helps relax the lung arteries. Eases breathlessness and supports the heart. Especially useful in Group 3 (lung disease).	Treat the underlying lung disease or sleep apnea. PAH medicines for Group 1. Water pills for fluid buildup.

Common Misconceptions

Myth	Reality
Pulmonary hypertension is just the same as regular high blood pressure.	FALSE. PH is high pressure in the lung arteries, a separate low-pressure system. Regular blood pressure pills do not treat it, and some can even be harmful.
All pulmonary hypertension is treated the same way.	FALSE. Treatment depends entirely on the group. PAH medicines help Group 1 but can harm Group 2. Finding the right group is essential.
Nothing can be done for pulmonary hypertension.	FALSE. Group 4 (CTEPH) can often be cured with surgery or balloon treatment. Group 1 has medicines that improve survival. Groups 2 and 3 improve when the cause is treated.
PAH medicines work for every type of PH.	FALSE. PAH-specific drugs are for Group 1 (and some Group 4). In Group 2 (left-heart) they can cause harm. The group must be confirmed first.
An echocardiogram alone is enough to diagnose PH.	Not quite. An echo is the best screening test, but it only estimates pressure. A right heart catheter is needed to confirm PH and measure the true pressure.
Once my old blood clots were treated, they cannot cause PH.	FALSE. Clots that do not fully dissolve can scar the lung arteries and cause CTEPH months or years later. This is why anyone with PH should be screened for it.
Feeling short of breath is just part of getting older.	Be careful. New or worsening breathlessness, fatigue, swelling, or fainting deserves a check. PH is often missed because symptoms are blamed on aging or being out of shape.
Pregnancy is safe if my PH is mild.	FALSE. Pregnancy is very high risk in PH, especially PAH, even when mild. Always plan with your PH team and use reliable birth control.

Cross-links to companion guides: Right Heart Catheterization — HFpEF — Pulmonary Embolism — Sleep Apnea — Advanced Heart Failure

Possible Complications

Knowing what can go wrong helps you spot problems early. Most complications are uncommon, especially with treatment.

Where / What	What Can Happen
Right-heart failure (cor pulmonale)	The main danger of PH. The strained right heart weakens and cannot keep up. It causes leg and belly swelling, fatigue, and breathlessness.
Low output and fainting	When the right heart cannot push enough blood through the lungs, the body and brain get less. This causes dizziness, near-fainting, or fainting — especially with effort.
Irregular heart rhythms	The stretched, strained heart can develop fast or irregular rhythms, such as atrial flutter or fibrillation. These can worsen symptoms quickly.
Blood clots	Slow, sluggish blood flow in widened lung arteries raises the risk of clots. Blood thinners are used in some types, especially CTEPH.
Coughing up blood (hemoptysis)	Rarely, high lung-artery pressure can cause bleeding in the airways. Any coughing up of blood needs prompt medical attention.
Fluid buildup (edema and ascites)	Right-heart strain backs fluid up into the legs, belly, and sometimes the liver. Water pills and a low-salt diet help control it.
Low blood oxygen	PH and its causes can drop oxygen levels, especially with activity or sleep. This adds strain to the heart and may need oxygen therapy.
Sudden worsening	An infection, missed medicine, or new clot can cause a fast decline. This is a medical emergency and may need hospital care.

Points to Know

If you remember nothing else, remember these key points.

- PH is high pressure in the lung arteries — a mean pressure over 20 mmHg. It strains the right side of the heart.
- There are five groups, sorted by cause. Group 2 (left heart) is the most common; Group 1 (PAH) is rare but has special medicines.
- An echocardiogram screens for PH; a right heart catheter confirms it and measures the true pressure. Both steps matter.
- Treatment depends on the group. PAH drugs help Group 1 but can harm Group 2. The right diagnosis comes first.
- Group 4 (CTEPH) comes from old blood clots and can often be cured with surgery or balloon treatment. Everyone with PH should be checked for it.
- Oxygen, water pills, and supervised exercise help symptoms in many groups. Push activity only with guidance.
- Pregnancy is very high risk with PH. Plan with your team and use reliable birth control.
- Care at a specialized PH center improves outcomes. Ask for a referral if you have not been to one.

When to Call Us - and When to Call 911

If you are not sure, call. We would rather hear from you twice than miss a real problem.

- New or worse shortness of breath — at rest, lying flat, or with light activity.
- Fainting, near-fainting, or repeated dizziness — call us promptly; fainting with PH can be serious.
- Chest pain or pressure — call 911 if it is severe or does not ease.
- Fast, pounding, or irregular heartbeat that is new for you.
- Coughing up blood — seek medical care right away.
- New or fast-worsening swelling of the legs, ankles, or belly.
- Weight gain of 3 to 5 pounds in a few days — it can mean fluid is building up.
- Bluish lips or fingertips, or oxygen levels lower than your usual target.
- Fever, cough, or signs of a chest infection — these are more dangerous with PH.
- If you think you may be pregnant — contact your PH team the same day.

Office: (727) 943-5200

Trusted Resources

The links below are the trusted resources Dr. Ali used to write this guide and that he recommends you visit for more information. Each is a clickable link in the digital version and a Short.io tracked URL on the printed version.

- Pulmonary Hypertension Association — Patients - Patient support, accredited PH care centers, and disease-management resources.
- AHA — Pulmonary Hypertension - American Heart Association overview of PH, its types, and symptoms.
- Cleveland Clinic — Pulmonary Hypertension - Patient guide to the five groups, diagnosis, and treatment.
- Cleveland Clinic — CTEPH (Group 4) - Patient guide to chronic clot-related PH and its potentially curative surgery.
- Mayo Clinic — Pulmonary Hypertension - Comprehensive patient guide to causes, symptoms, and management.
- NIH / MedlinePlus — Pulmonary Hypertension - Official NIH patient page with plain-language information and links.
- American Lung Association — Pulmonary Hypertension - Patient education on PAH and lung-related PH from the American Lung Association.
- ACC CardioSmart — Pulmonary Hypertension - ACC patient-education platform covering PH diagnosis and care.

Sources Used to Build This Guide

Every recommendation in this guide is grounded in one or more of the studies, guidelines, or trusted resources below. Short links are tracked so Dr. Ali can see which references patients are reading; nothing personally identifying is captured.

- ESC/ERS Pulmonary Hypertension Guidelines (2022) *[Guideline]*
  Current definition (mean PA pressure over 20 mmHg), 5-group classification, diagnostic algorithm (echo screen, right heart cath confirms), and group-specific treatment standards
- ACC — 2022 ESC/ERS PH Guidelines: Key Points *[Guideline]*
  Clinician summary of hemodynamic definitions (PVR over 2 Wood Units, wedge 15 mmHg or less for PAH) and risk-stratified treatment
- AHA — Pulmonary Hypertension *[Patient-Education]*
  Plain-language framing of what PH is, types, and symptoms
- Cleveland Clinic — Pulmonary Hypertension *[Patient-Education]*
  Five-group classification, diagnosis, and treatment overview in patient language
- Mayo Clinic — Pulmonary Hypertension *[Patient-Education]*
  Causes, symptoms, complications (right heart strain), and management
- CHEST — Pharmacologic Therapy for PAH in Adults (CHEST Guideline) *[Guideline]*
  Group 1 PAH-specific drug classes (endothelin receptor antagonists, PDE5 inhibitors, prostacyclins) and combination therapy
- Cleveland Clinic — Chronic Thromboembolic Pulmonary Hypertension (CTEPH) *[Patient-Education]*
  Group 4 CTEPH: pulmonary endarterectomy is potentially curative; balloon angioplasty for inoperable disease
- Annals of Thoracic Surgery — Pulmonary Thromboendarterectomy for CTEPH *[Journal]*
  Pulmonary endarterectomy (PEA) as the potentially curative treatment of choice for operable CTEPH
- European Heart Journal — Balloon Pulmonary Angioplasty Consensus (ESC Working Group, 2023) *[Guideline]*
  Balloon pulmonary angioplasty (BPA) for technically inoperable CTEPH or residual PH after PEA
- NIH/MedlinePlus — Pulmonary Hypertension *[Patient-Education]*
  Official NIH patient page with plain-language symptoms and links
- Pulmonary Hypertension Association — Patient Resources *[Patient-Education]*
  Patient support, accredited PH care centers, and disease-management resources

About This Guide

Rias KS Ali, MD FACC | Board Certified in Interventional Cardiology & Cardiovascular Disease

4740 Mile Stretch Drive, Holiday FL 34690 | Office: (727) 943-5200 | Fax: (727) 943-5201 | RiasAliMD.com

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Reviewer note: Reviewed against Cleveland Clinic, Mayo Clinic, and AHA pulmonary hypertension pages. This guide adds a five-group cover map, an echo-to-right-heart-cath diagnostic path diagram, a treatment-by-group table, per-group deep-dive sections, and a clear note that Group 4 (CTEPH) can often be cured with surgery or balloon treatment.

Disclaimer

This guide is for educational purposes only and does not replace medical advice from your physician. Recommendations may not apply to every patient. Always follow the specific instructions of Dr. Ali and your care team. In an emergency call 911 or go to the nearest emergency department.

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