Pulmonary hypertension medication list

Every FDA-approved PAH therapy by class and pathway, with functional class, combinations and monitoring

Pulmonary arterial hypertension is treated by widening the pulmonary arteries and slowing the vessel remodeling behind the pressure. Three pathways do that work: endothelin, nitric oxide and sGC, and prostacyclin. Almost every approved drug acts on one of them.

That is why an alphabetical drug list is hard to use at the bedside. Grouping by pathway shows what a patient already has covered, and which column is still open. Escalation means adding a column you have not covered yet.

1. EVERY FDA-APPROVED PAH MEDICATION, BY CLASS

DRUG CLASS	GENERIC (BRAND)	ROUTE	KEY MONITORING
Endothelin receptor antagonists Endothelin pathway	**Bosentan (Tracleer)**	Oral	Monthly liver function tests. REMS for hepatotoxicity and pregnancy risk
	Ambrisentan (Letairis)	Oral	REMS for pregnancy risk, monthly pregnancy testing
	Macitentan (Opsumit)	Oral	REMS for pregnancy risk. No mandated monthly LFTs
PDE-5 inhibitors Nitric oxide pathway	**Sildenafil (Revatio)**	Oral, IV	Systemic hypotension, vision changes
	Tadalafil (Adcirca)	Oral	Blood pressure. Contraindicated with nitrates
Soluble guanylate cyclase stimulator Nitric oxide and sGC pathway	**Riociguat (Adempas)**	Oral	Never with a PDE-5 inhibitor or a nitrate
Prostacyclin analogues Prostacyclin pathway	**Epoprostenol (Flolan, Veletri)**	IV infusion	Continuous pump operation, line infections
	Treprostinil (Remodulin, Tyvaso)	Subcutaneous, inhaled, oral	Infusion site reactions, jaw pain
	Iloprost (Ventavis)	Inhaled	Cough, throat irritation
IP receptor agonist Prostacyclin pathway	**Selexipag (Uptravi)**	Oral	Headache, jaw pain, fluid retention
Calcium channel blockers	**Nifedipine, diltiazem**	Oral	Only after a positive vasoreactivity test. Blood pressure and edema
Fixed-dose combination	**Macitentan and tadalafil (Opsynvi)**	Oral	Same REMS as macitentan, plus blood pressure
Activin signaling inhibitor Outside all three pathways	**Sotatercept (Winrevair)**	Subcutaneous, every 3 weeks	Hemoglobin and platelets before each of the first 5 doses, then periodically

Opsynvi, approved in March 2024, puts macitentan and tadalafil in one daily tablet and removes a pill from the regimen. Sotatercept arrived the same month with a mechanism none of the older classes share, given on top of background therapy.

2. WHO FUNCTIONAL CLASS SETS THE STARTING REGIMEN

WHO FC	CLINICAL SYMPTOMS	RECOMMENDED INITIAL STRATEGY
I	No dyspnea with ordinary activity	Observation, or monotherapy where treatment is indicated
II	Mild exertional dyspnea and fatigue with ordinary activity	Upfront dual therapy, usually an ERA plus a PDE-5 inhibitor
III	Marked exertional dyspnea, unable to perform usual activities	Dual therapy, moving to triple therapy when risk assessment is high
IV	Dyspnea or syncope at rest, unable to perform any activity	Triple therapy including an IV or subcutaneous prostacyclin

Read this as a starting point, not a rule. A patient in class III with comorbidities may be managed differently from a young patient with idiopathic disease. What stays constant is writing the class down at every visit.

3. COMBINATION THERAPY IS THE DEFAULT AT DIAGNOSIS

Sequential monotherapy has largely gone. Targeting two or three pathways at once produces faster hemodynamic improvement and fewer episodes of clinical worsening. Approved pairings include:

- Bosentan (ERA) with sildenafil (PDE-5 inhibitor).
- Ambrisentan (ERA) with tadalafil (PDE-5 inhibitor).
- Macitentan (ERA) with tadalafil (PDE-5 inhibitor), also sold as the single tablet Opsynvi.
- Riociguat (sGC stimulator) alone, or alongside a prostacyclin.
- Triple therapy: an ERA, a PDE-5 inhibitor, and a prostacyclin or IP agonist.

> **One combination is never allowed**
>
> Riociguat and a PDE-5 inhibitor both act on the nitric oxide pathway. Together they cause severe hypotension and syncope. The same warning applies to riociguat with any nitrate.

4. WHAT EACH CLASS NEEDS YOU TO MONITOR

- **Endothelin receptor antagonists.** Bosentan is the only one that still requires monthly liver function tests. The FDA removed that for ambrisentan in 2011, and it was never mandated for macitentan. All three stay under a REMS for teratogenicity, so patients who can conceive need contraception counseling and monthly pregnancy tests.
- **PDE-5 inhibitors.** Blood pressure at each review, and a question about vision changes on sildenafil.
- **Continuous prostacyclin infusions.** Daily pump management and a look at the infusion site. A line infection is the complication that puts these patients in the hospital.
- **Sotatercept.** Hemoglobin and platelet counts before each of the first five doses, then periodically. Erythrocytosis and thrombocytopenia are why those counts matter.

5. WORKING THE REFERENCE INTO A PAH VISIT

1. **Record the baseline**
   At diagnosis, note WHO functional class and every current medication, diuretics and anticoagulants included.

2. **Map the pathways**
   Mark which of the three pathways each drug already covers. The open column is your escalation candidate.

3. **Plan the combination**
   Decide between monotherapy, dual therapy and triple therapy, then choose the classes to combine.

4. **Set the monitoring**
   Write down baseline labs, blood pressure, echocardiography and catheterization dates, with the review interval for each drug.

5. **Track the escalation**
   At each follow-up, usually 4 to 12 weeks after a dose change, reassess functional class, hemodynamics and side effects.

6. BEFORE YOU CLOSE THE CONSULTATION

	CHECK
	Functional class recorded today, not carried over
	Every targeted agent listed with its class and route
	Pathways covered, and the one still open, written down
	Next monitoring date set, with the parameter named
	REMS and contraception status current for anyone who can conceive

7. SUPPORTIVE DRUGS, AND WHERE LISTS GO WRONG

Targeted therapy is only part of the regimen. Diuretics manage fluid overload from right heart failure. Anticoagulants are used in selected patients to reduce thrombosis risk. Supplemental oxygen corrects hypoxia, and digoxin is sometimes added for right ventricular support.

Four failures that turn up again and again

- The record holds brand names only, so nobody can see which pathway is already covered.
- A drug stopped for side effects stays on the list, because the change was noted in free text.
- REMS dates and pregnancy test results live in a separate spreadsheet that one nurse maintains.
- A route change goes unrecorded, so a patient moved to inhaled treprostinil still reads as subcutaneous.

This reference summarizes FDA-approved labeling and published guideline recommendations. It does not replace the prescribing information, the current ESC/ERS guidelines, or your own clinical judgment.