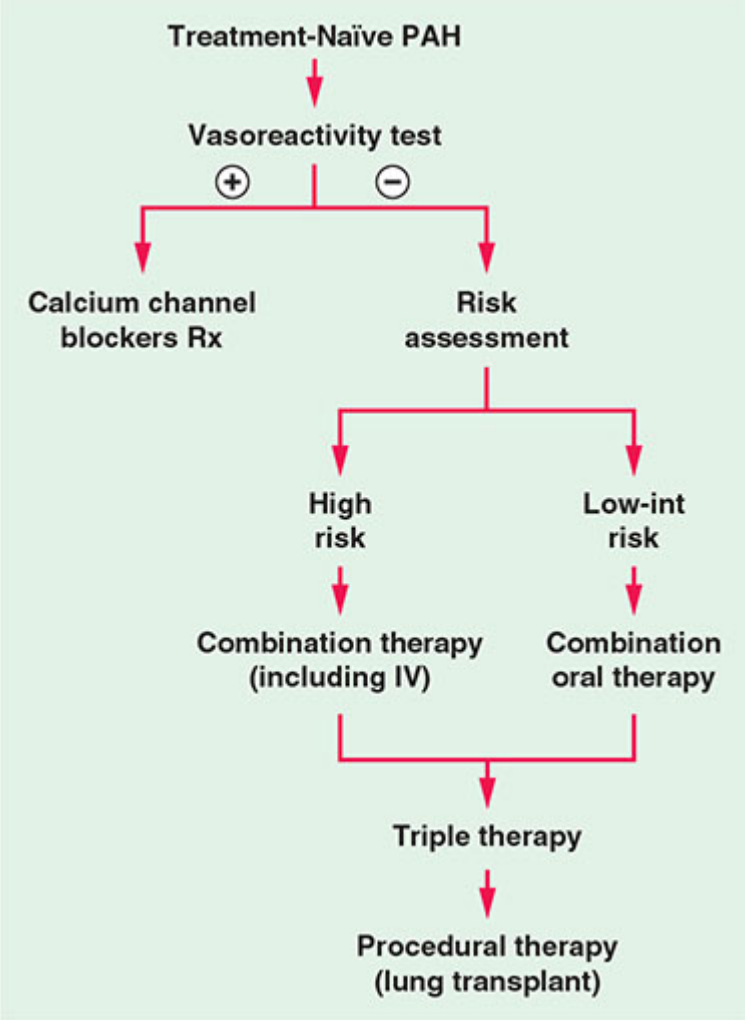

FIGURE 283-8 Treatment strategy overview for patients with newly diagnosed pulmonary arterial hypertension (PAH). Incident, treatment-naïve patients diagnosed with PAH should be assessed with vasoreactivity testing at the time of right heart catheterization. Patients with a positive vasoreactivity test indicating an acute and robust vasodilatory response following administration of nitric oxide (or other approved pulmonary vasodilator) are treated with high-dose oral calcium channel blocker therapy (Rx) and have a favorable prognosis, but compose a minority (<5%) of all PAH patients. Among patients with a negative vasoreactivity study, treatment selection is determined by clinical risk: high-risk patients, such as those with advanced heart failure or syncope, are treated with combination drug therapy, including intravenous prostacyclin therapy. Low- or intermediate (Int)-risk patients are initiated on combination oral therapy, which generally includes an endothelin receptor antagonist and phosphodiesterase type 5 inhibitor. Subsequent add-on therapy (i.e., triple therapy) is considered in patients who deteriorate clinically or fail to improve. Lung transplantation or other