



The 6th World Symposium of PH: Risk Stratification and Medical Treatment in PH (Part 3)

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Expert Analysis

Note: This is **Part 3** of a 6-part series on the 6th World Symposium on PH.

[Part 1](#) | [Part 2](#) | [Part 3](#) | [Part 4](#) | [Part 5](#) | [Part 6 \(Coming Soon!\)](#)

1. The progress observed in medical therapy of pulmonary arterial hypertension (PAH) over the last decade is not related to the discovery of new pathways but to the development of new strategies for combination therapy and on escalation of treatments based on systematic assessment of clinical response.
2. The current treatment strategy is based on the severity of PAH as assessed by a multiparametric risk stratification approach. Goals of therapy and factors associated with better prognosis include functional capacity Class I-II, 6-minute walk distance >400 meters, and normal right ventricular function per echocardiography and hemodynamic parameters.
3. Current treatment recommendations call for upfront oral combination therapy for low- and intermediate-risk patients with PAH and upfront combination therapy that should include parenteral prostacyclin therapy for patients with high-risk features.
4. Patients should be re-evaluated 3-6 months from the start of combination therapy. If goals of therapy are not met, sequential triple therapy or escalation of therapy from oral to parenteral prostacyclin is to be considered. Current recommendations are for evaluations every 3-6 months, particularly for the high-risk patients who may need lung transplant referral if they are refractory to maximal medical therapy.
5. Regarding vasoreactivity testing, it is recommended to evaluate response to calcium channel blockers only for patients with idiopathic PAH, heritable PAH, and PAH associated with drugs and toxins. If positive, very high doses of

calcium channel blockers should be used (i.e., amlodipine 20 or 30 mg daily). If goals of therapy not achieved after 3-6 months, it is recommended to start specific PAH therapy.

6. Supportive therapy should include supervised exercise training.
7. Anticoagulation is not recommended for associated PAH but may be considered for idiopathic, heritable, or drug-induced PAH because data in those groups are less controversial.
8. Occasionally, patients have an extraordinary response to therapy, and transition to a less-invasive therapy is considered. Because much of the literature on this topic is retrospective, prospective but observational, or prospective randomised but open label, this approach is not recommended except in rare circumstances and under close expert care. There have been conflicting outcomes in the transition from parenteral prostacyclins to inhaled or oral prostacyclins. When discontinuation of bosentan is necessary due to liver function test elevations, transitioning to ambrisentan or macitentan is safe. At the time of the 6th World Symposium of PH, there was insufficient evidence to recommend transition from sildenafil or tadalafil to riociguat for improving efficacy, although studies are underway.

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