Background: Pulmonary arterial hypertension (PAH) is a progressive disease that can result in morbidity and mortality. Several new PAH medications are available since 2012, which are intended to delay progression and improve functional measures. More information is needed regarding medication treatment patterns and adherence.

Methods: Patients with a diagnosis code for pulmonary hypertension and treated with an approved medication for PAH (ERAs, PDE-5is, prostacyclins/IP receptor agonists, sGCs) identified by pharmacy claims between January 2012 and March 2017 were included. Patients were ≥18 years old with continuous enrollment with medical and pharmacy coverage for 6 months before (with no PAH pharmacy claim) and ≥1 year after initiating a PAH-related medication. Patients were followed until disenrollment from the plan or end of study (March 2018). Medication treatment patterns were examined during ≥1 year of follow-up.

Results: The study included 1878 patients, mostly female (64.4%) with mean (+SD) age 66.4±13.3 years. Common comorbidities included hypertension (81.7%), heart failure (58.8%), coronary artery disease (52.2%), type 2 diabetes mellitus (40.9%), sleep apnea (38.3%), and obesity (28.8%). Only 9.2% of patients initiated treatment with combination therapy. PDE-5is and ERAs were used in 71.9% and 23.9%, respectively, of initial treatment regimens. Only 37.8% of patients remained on their initial medication regimen until study end. Of patients who discontinued medication therapy (32.0%) or modified (30.2%) their initial regimen, 78.4% did so within one year and the median time to therapy discontinuation or modification was 4.1 months (IQR 1.6-10.4 months). Combination therapies comprised 50.4% (286/567) of second regimens, most often with ERA+PDE-Si alone (153/286, 53.5%) or with prostacyclin/IP receptor agonist (14.0%). Of patients later switching to combination therapy, most (60.3%) did so within nine months of starting the initial medication regimen. The mean proportion of days covered (PDC) was 86%, and was higher for patients with initial combination therapies (95%) than monotherapies (85%). ERAs as initial treatment were associated with higher mean 12-month PDC (82% vs. 67%) and persistence (median 4.9 vs. 4.5 months) than PDE-5is.

Conclusions: Patients with PAH most often initiated medication treatment with monotherapies, usually PDE-5is in spite of lower adherence and persistence than ERAs. Adjustments to initial medication regimens occurred early and in the majority of patients. Most patients modified or discontinued their initial treatment regimen, although most patients remained treated with monotherapies despite 2015 ESC/ERS guidelines following the results from the AMBITION treatment strategy trial with initial combination therapy.