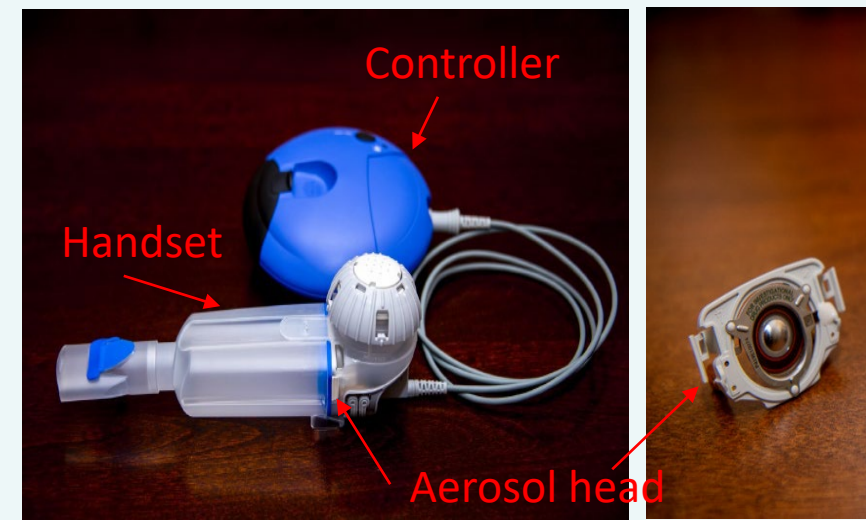


BACKGROUND

- AP01 is a novel formulation of pirfenidone optimized for inhalation using the Pari eFlow nebulizer.
- The AP01-002 (ATLAS) study of AP01 in Idiopathic Pulmonary Fibrosis (IPF)¹ began in June 2019 and was followed by the AP01-005 open label extension (OLE) study.
- Additional subjects with IPF and with Progressive Pulmonary Fibrosis (PPF) were enrolled in the OLE study (naïve cohort).
- We report AP01 safety and efficacy in subjects out to 180 weeks

NEBULIZER

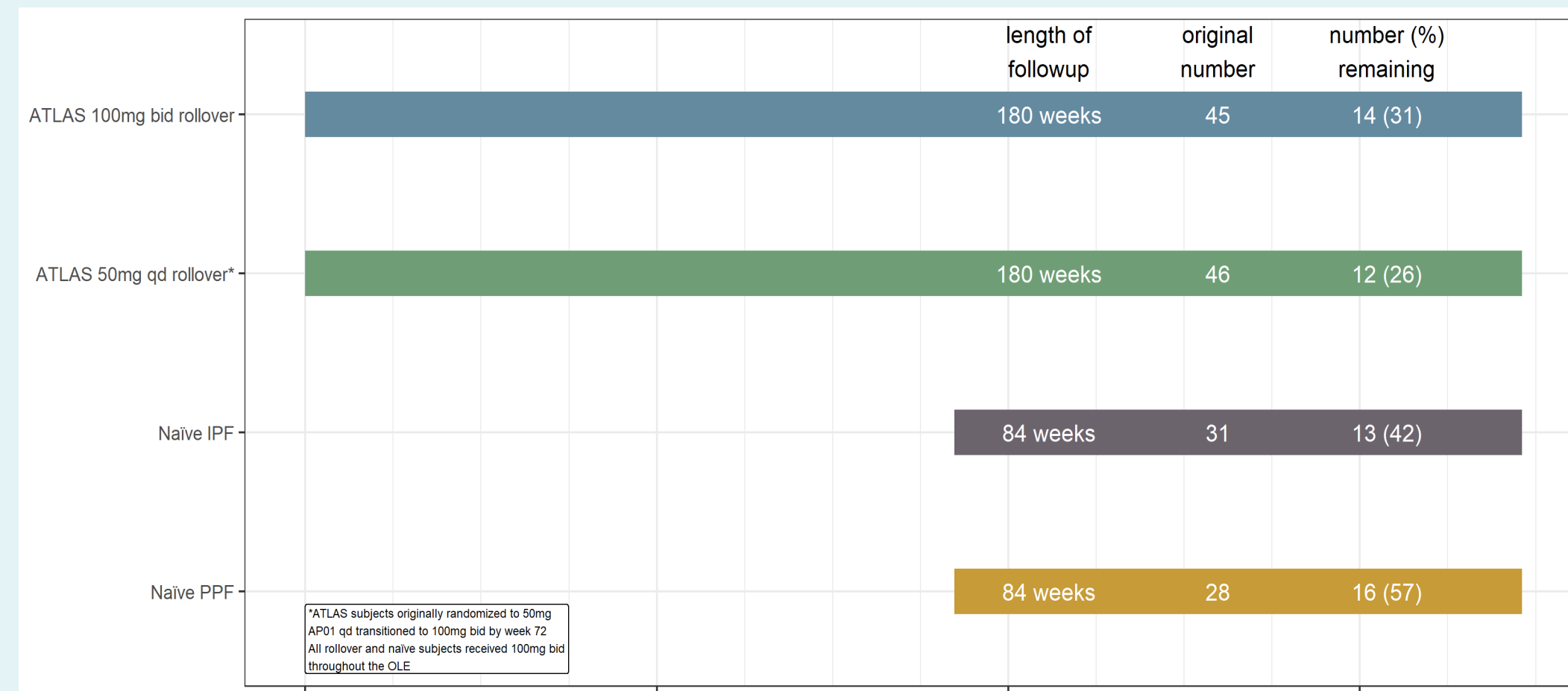
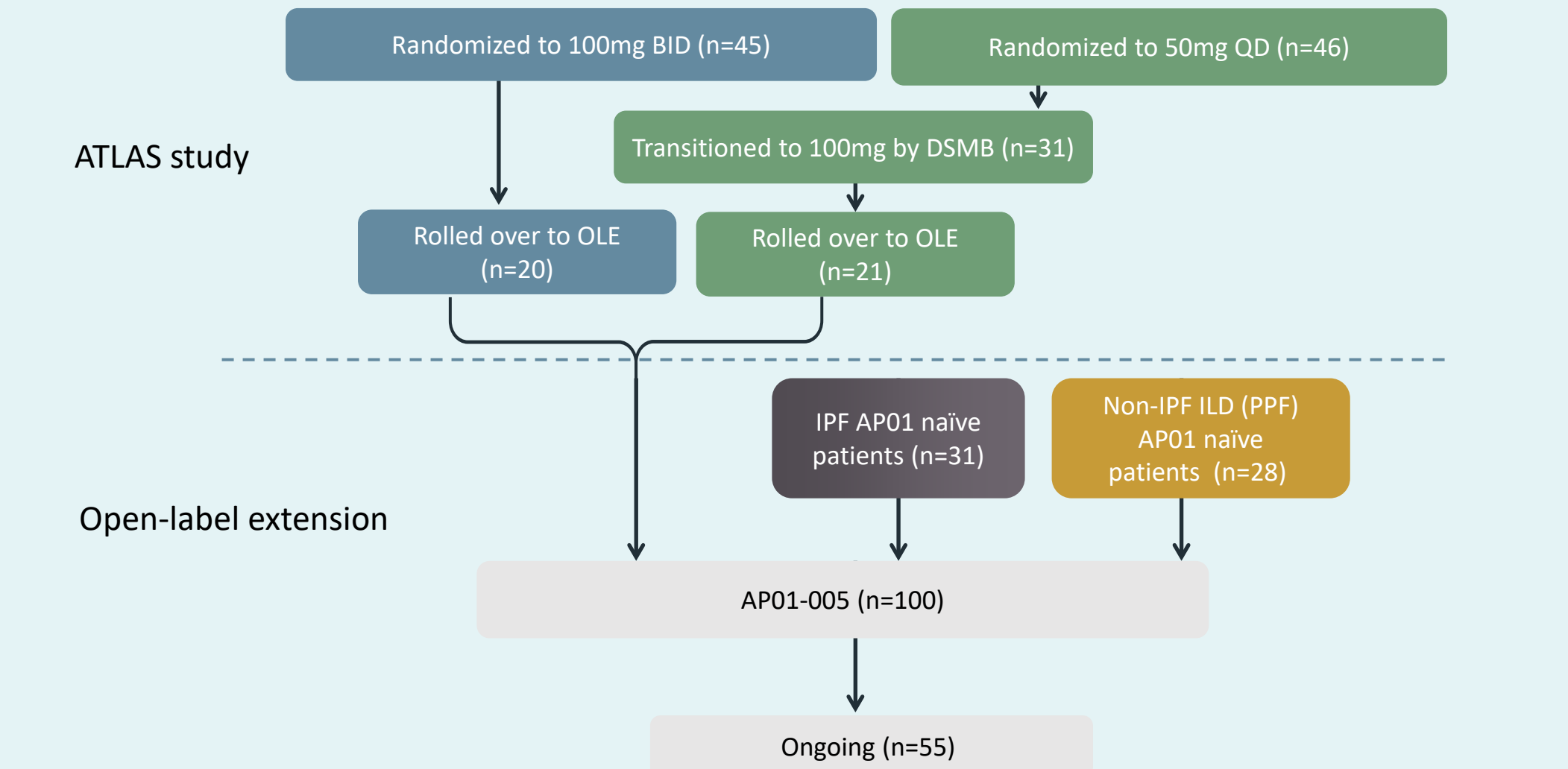
- AP01 is administered by the modified eFlow Nebulizer
- Vibrating mesh membrane delivers 8 mL over a median time of 8-9 minutes
- Comparing 100mg nebulized AP01 with 801mg oral pirfenidone
 - Alveolar C_{max} is 35x higher
 - Plasma exposure is 15x lower



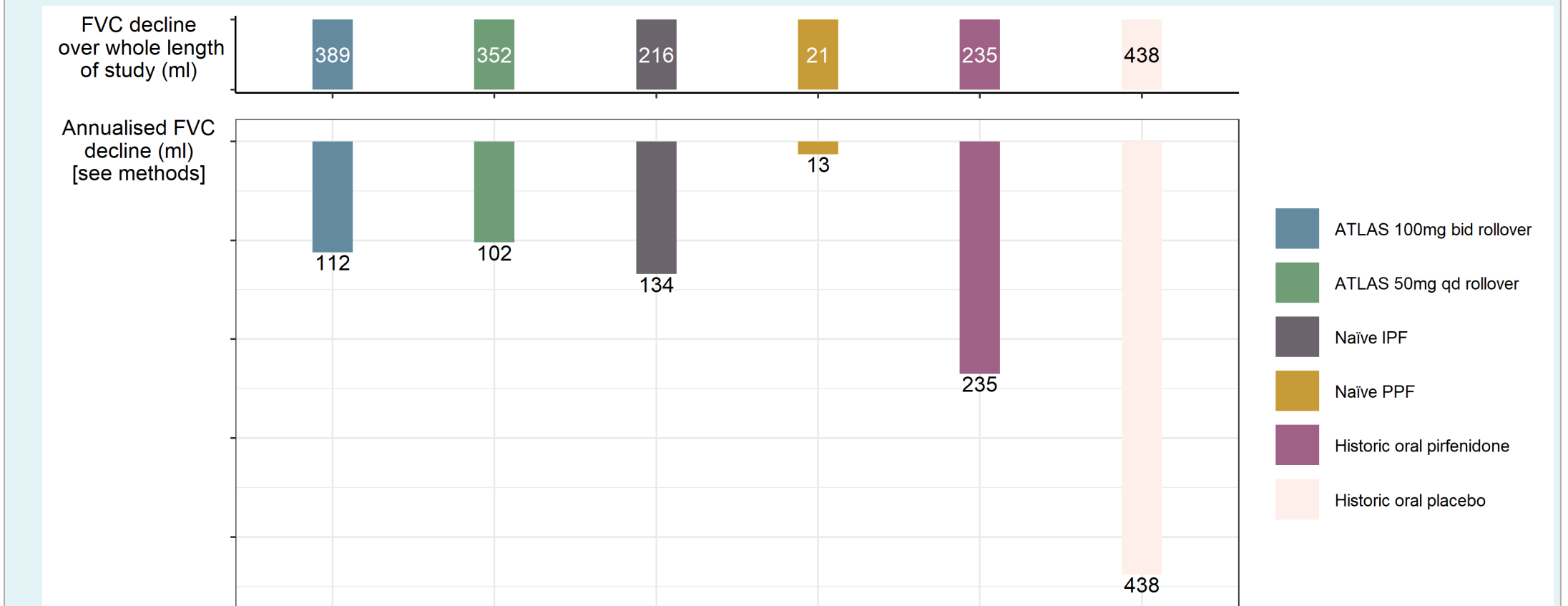
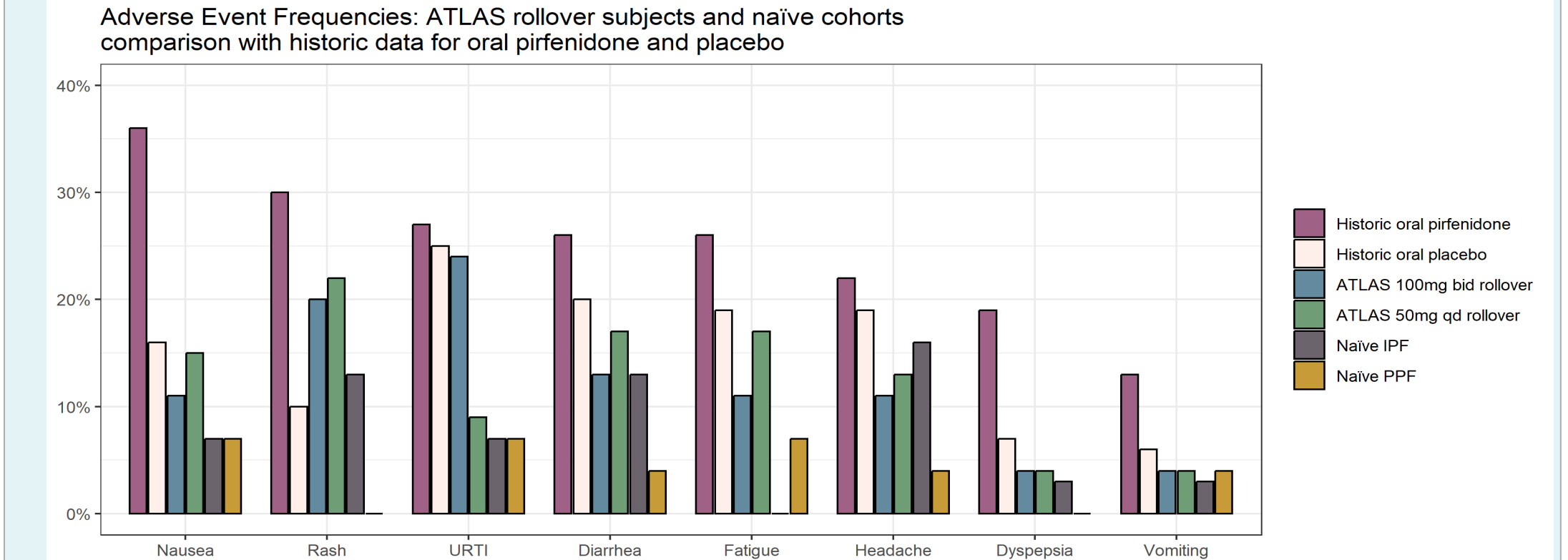
METHODS

- The ATLAS study randomized IPF patients to 50 mg AP01 once daily (qd) or 100 mg AP01 twice daily (bid) for 24 weeks.
- At the recommendation of the DSMB, ATLAS subjects originally randomized to AP01 50 mg qd transitioned to 100 mg bid by week 72.
- All cohorts in the OLE study received 100mg bid AP01 throughout.
- Forced Vital Capacity (FVC) was recorded every 3 months.
- We report
 - the change in FVC from baseline to latest visit (calculated as the mean of the difference between the earliest and latest FVC for each subject remaining on the study)
 - the annualized decline: overall $\Delta FVC \times (52/\text{study length [weeks]})$, and
 - the percentage of subjects experiencing any adverse event.
- Results were compared with published pooled data from the 3 pivotal clinical trials of oral pirfenidone².

STUDY DESIGN



SAFETY AND EFFICACY



CONCLUSIONS

The data from this OLE suggest that AP01 shows continuing efficacy at up to 180 weeks with a safety profile superior to oral pirfenidone. AP01 is currently being investigated in the MIST PPF study, a phase 2b randomized controlled study in Progressive Pulmonary Fibrosis³

REFERENCES:

1. West, A. et al. Inhaled pirfenidone solution (AP01) for IPF: a randomised, open-label, dose-response trial. Thorax (2023) doi:10.1136/thorax-2022-219391
2. Noble, P. W. et al. Pirfenidone for idiopathic pulmonary fibrosis: analysis of pooled data from three multinational phase 3 trials. European Respiratory Journal ERJ-00026 (2015).
3. <https://mistppfstudy.com/>