

European Respiratory Society Annual Congress 2013

Abstract Number: 2861

Publication Number: P3409

Abstract Group: 4.3. Pulmonary Circulation and Pulmonary Vascular Disease

Keyword 1: Pulmonary hypertension **Keyword 2:** Treatments **Keyword 3:** Monitoring

Title: Characteristics of idiopathic pulmonary arterial hypertension (IPAH) and PAH associated with connective tissue disease (APAH-CTD) patients in PROSPECT

Dr. Harrison 11869 Farber hfarber@bu.edu MD ¹, Dr. Robert 11870 Frantz frantz.robert@mayo.edu MD ², Dr. Robert 11871 Schilz Robert.Schilz@UHhospitals.org MD ³, Dr. Murali 11872 Chakinala mchakina@dom.wustl.edu MD ⁴, Dr. Brian 11873 Hartline brian.hartline@actelion.com MD ⁵, Mr. Dave 11874 Miller dave.miller@iconplc.com ⁶, Dr. Daniel 11878 Rosenberg Daniel.Rosenberg@actelion.com ⁷, Dr. Wade 11879 Benton wade.benton@actelion.com ⁵ and Dr. Robyn 11880 Barst robyn.barst@gmail.com MD ⁸. ¹ Department of Medicine, Boston University School of Medicine, Boston, United States ; ² Cardiovascular Diseases and Internal Medicine, Mayo Clinic College of Medicine, Rochester, United States ; ³ Pulmonary/Critical Care, University Hospitals of Cleveland, Cleveland, United States ; ⁴ Pulmonary & Critical Care Medicine, Washington University School of Medicine, St. Louis, United States ; ⁵ Medical Affairs, Actelion Pharmaceuticals US, Inc., South San Francisco, United States ; ⁶ Late Phase & Outcomes Research, ICON Plc, San Francisco, United States ; ⁷ Epidemiology & Observational Studies, Actelion Pharmaceuticals, Ltd., Allschwil, Swaziland and ⁸ Pediatrics, Columbia University College of Physicians and Surgeons, New York, United States .

Body: Background: The PROSPECT Registry evaluates use of Epoprostenol for injection (Veletri®, EFI), an IV prostacyclin (PGI₂) with improved stability. We compare characteristics and clinical outcomes of APAH-CTD and IPAH registry patients (pts). Methods: PROSPECT is a 48-center US, observational drug registry following pts for 1 year including pts naïve to IV prostacyclin therapy (NV) or transitioned from another IV PGI₂ therapy (TR) at enrollment. Disease characteristics at time of enrollment, and outcomes and dosing regimens up to 1 year of follow-up are presented. Results: PROSPECT enrolled 90 pts with APAH-CTD (n=39 NV, 51 TR) and 172 pts with IPAH (n=76 NV, 96 TR). The majority were NYHA functional class III/IV and female. Mean (± SD) six minute walk distance (6MWD) was 317±110 meters (m) for NV IPAH and 298 ±114 m for NV APAH-CTD pts. Mean (±SD) cardiac index (CI) was 2.1±0.7 L/min/m² for NV IPAH and 2.6±1.4 L/min/m² for NV APAH-CTD pts. The rate per 1000 pt days of blood stream infections (BSIs) was lower in NV APAH-CTD than in NV IPAH pts (0.10 vs. 0.23); in contrast, TR APAH-CTD pts had a higher rate of BSIs than TR IPAH pts (0.21 vs. 0.10). One year Kaplan-Meier survival estimates for IPAH pts were (NV=85.2%, TR=88.7%) compared to APAH-CTD pts (NV=74.8%, TR=84.3%). There was no significant difference in median EFI dose between NV APAH-CTD and NV IPAH pts: (10 and 25 ng/kg/min at one and 12 months). Conclusion: Compared to IPAH pts, APAH-CTD pts in PROSPECT had a lower 6MWD and higher CI. EFI dosing did not differ between the two populations.