Characteristics of Idiopathic Pulmonary Arterial Hypertension (IPAH) and PAH Associated With Connective Tissue Disease (APAH-CTD) Patients in PROSPECT

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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 enrolment. 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).

CONCLUSIONS: 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.

TYPE: Clinical Science

*The authors are saddened to report the passing of Robyn J. Barst, MD in April 2013. She was an esteemed physician, investigator, and colleague, and a distinguished leader in the field of pediatric pulmonary hypertension. Her contributions to the field are invaluable.