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**Title:** An open label study of ambrisentan in patients with exercise induced pulmonary arterial hypertension (EiPAH)

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**Body:** EiPAH, defined by: mPAP>30mmHg at peak exercise, PVRmax>80 dynes sec cm-5, and PCWPmax<20mmHg in the absence of resting PH, is increasingly recognized as clinically important and may represent an early form of PAH. Identifying the disease and initiating treatment at an early stage may improve functional status and prevent progression to severe forms of PAH. Methods: Single-center, open-label 6-month study to evaluate the effects of ambrisentan on exercise capacity utilizing invasive cardiopulmonary exercise testing (iCPET) in EiPAH. Patients were recruited after a clinically indicated ICPET for evaluation of unexplained exertional dyspnea. After 6-months of treatment patients underwent repeat iCPET, and exercise capacity, symptoms and hemodynamics were assessed. We compared change from baseline in peak exercise values of VO<sub>2</sub>, mPAP, PAWP, PVR, and CO. Results: To date 5 patients (4/5 male, age 58.2±7.0 years) have completed the 6-month treatment phase and undergone repeat ICPET. Baseline resting hemodynamics were mPAP=18.4±4.2mmHg, PAWP=9.4±4.3mmHg, CO=4.3±1.3l/min and PVR=141.2±70.8 dynes sec cm-5; peak values were mPAP=37.6±5.2, PAWP=14.0±4.1, CO=9.8±2.0 and PVR=176.4±63.7 dynes sec cm-5. After 6 months of treatment there was an increase in peak CO (+3.4±2.4l/min), with a decline in peak mPAP (-7.6±3.5, range -2 to -11; p=0.008) and PVR (-94.8±59.2, range -35 to -176; p=0.02). Neither peak PCWP (+0.8±8.2, p=0.84) nor peak VO<sub>2</sub> (+48.4mL/min, p=0.29) changed significantly. Conclusions: These preliminary findings suggest that treatment of EiPAH with ambrisentan results in improved hemodynamics over a 6-month period. Further study of EiPAH and early treatment is warranted.