

CT findings of pulmonary hypertension predict treatment response in idiopathic pulmonary fibrosis

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PURPOSE: To determine whether CT signs of pulmonary hypertension predict treatment response to sildenafil in idiopathic pulmonary fibrosis (IPF).

MATERIALS AND METHODS: In the Sildenafil Trial of Exercise Performance in IPF, individuals were randomized to treatment with oral sildenafil or placebo for 12 weeks. The primary outcome was improvement in 6-minute walk distance (6MWD) after treatment. Diameters of main pulmonary trunk (PT), left pulmonary artery (LPA), right pulmonary artery (RPA), and ascending aorta (AA) were measured on baseline CT's of 170 participants. To evaluate treatment effect, we compared those with values above and below the median for each measurement. A general linear model estimated 12 week change in 6MWD as a function of CT-derived indices for pulmonary hypertension, sildenafil treatment, and the interaction between CT indices and sildenafil, adjusted for age, height, sex, race and visually estimated severity of IPF on CT.

RESULTS: Median (25th, 75th percentile) values (in cm) for PT, LPA, RPA, AA were 3.27 (2.98, 3.51), 2.64 (2.38, 2.84), 2.71 (2.46, 2.91), and 3.56 (3.34, 3.79), respectively. Median PT/AA ratio was 0.92 (0.84, 0.98). The individuals were divided by PT/AA ratio into 2 groups: > 0.92, and < 0.92. Subjects with PT/AA ratio > 0.92 and treated with sildenafil showed improvement in 6MWD (mean 50.6 m) as compared with placebo treatment (95% CI: 5.8, 95.3; p = 0.027). A significant sildenafil effect was not observed when PT/AA ratio was ≤ 0.92.

CONCLUSION: In this population of subjects with moderate or advanced IPF, pulmonary artery diameters were generally above the normal range. PT/AA ratio may predict treatment response to sildenafil in patients with IPF.