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Title: Change in diffusing capacity for carbon monoxide as a predictor of outcomes in connective tissue disease-associated pulmonary arterial hypertension: Analysis from the REVEAL registry

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Body: Background: Change in diffusing capacity for carbon monoxide (DLCO) is a more powerful predictor of outcomes than baseline DLCO in connective tissue disease (CTD)-associated pulmonary arterial hypertension (PAH). Aims and Objectives: We sought to validate these findings in a larger, multi-center CTD-APAH cohort. Methods: CTD-APAH patients (pts) in the Registry to EValuate Early And Long Term PAH Management (REVEAL), comprising 3515 pts with PAH from 56 US centers, were divided into 3 cohorts based on change in DLCO over time: 1) decreased by >10% ($DLCO_{dec}$), 2) unchanged within ±10% ($DLCO_{stab}$), 3) increased by >10% ($DLCO_{inc}$). Two-year survival after second DLCO assessment was evaluated. Results: 249 CTD-APAH pts were analyzed. Most pts were women (87.9%), with Ssc-APAH (73.9%), functional class II & III symptoms (77.9%), and on PAH therapies (83.9%). While 66.3% of pts were stable over time, 25.3% had reduced (-20.1±12.4%) and 8.4% improved (25.1±18.2%) DLCO. Mean time between DLCO measurements did not differ between $DLCO_{dec}$ and $DLCO_{inc}$ pts (34.6±28.6 vs. 29.7±25.2 mos., P>0.05). All deaths (80) occurred in $DLCO_{dec}$ and $DLCO_{stab}$ cohorts. The $DLCO_{inc}$ cohort had the best 24-month survival (log-rank p=0.007, $DLCO_{inc}$ vs. $DLCO_{stab}$). Multivariable analysis showed $DLCO_{dec}$ as a predictor of worst survival (HR 2.8, CI=1.65-4.73, P<0.001). Conclusion: Increase in DLCO by >10% was associated with improved outcomes in CTD-APAH. Identification of mechanisms and role of therapies in DLCO enhancement needs further investigation and may provide significant insights into future clinical care of patients with PAH.