**Supplementary Table 3 Cardiopulmonary exercise testing and prognostic parameters in SSc**

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| --- | --- | --- | --- | --- |
| **Reference** | **Author** | **Number of patients (N)**  | **Main parameter** | **Result** |
|  |  |  |  |  |
| **SSc and CPET** |
| 8 | Cuomo G 2010 | 46 | peakVO2 | Inversely correlates with heart and lung involvement |
| 9 | Rosato E 2014 | 40 | VE/VCO2-slope  | Correlates with disease activity index |
| 10 | Boutou AK 2016 | 78 | peakVO2VO2@ATVE/VCO2-slope pETCO2RV | Discriminates respiratory, left ventricular and pulmonary vascular limitation  |
| 11 | Walkey AJ 2010 | 19 | AADO2 at exercise | Discriminates pulmonary-vascular and left ventricular limitation |
| 36 | Sudduth CD 1993 | 11 | peakVO2VO2@ATVO2/HR | Circulatory impairment  |
| 49 | Michelfelder M 2017 | 54 | peakVO2DLCOFEV1%FVC | SSc and PAH patients, parameters do not discriminate coexisting interstitial lung disease |
| 58 | Vandecasteele E  | 3185 (meta-analysis) | 6-MWD | PAH and ILD both shorten 6-MWD in SSc |
| 64 | Dumitrescu D 2010 | 30 | peakVO2VO2@ATVO2/HRVE/VCO2@ATpETCO2@AT | VE/VCO2@AT and pETCO2@AT distinguish pulmonary vasculopathy from left ventricular limitation |
| 65 | Reichenberger F 2009 | 33 | peakVO2 | Lower in exercise-induced PH  |
| 66 | Dumitrescu D 2017 | 173 | peakVO2VE/VCO2@AT | Both parameters highly correlated with PAPmean |
| 67 | Morelli S 2000 | 18 | peakVO2VO2@ATVO2/HR | All associated with PH |
| 68 | Schwaiblmair M 1996 | 78 | peakVO2DLCOAADO2 at exercise | Discriminate lung involvement  |
|  |  |  |  |  |
| **SSc patients vs normal subjects**  |
| 32 | Chia EM 2016 | 25 | Δs' of right ventricle | Reduced in SSc vs normal  |
| 33 | de Oliveira NC 2007 | 13 (female only) | peakVO2DLCO | Reduced even without ILD |
| 34 | Hargardottir H 2010 | 11 | peakVO2VE/VCO2-slope  | Impaired in 9 of 11, inversely correlated with Interleukin-6 |
| 35 | Plazak W 2011 | 46 | peakVO2VE/VCO2@ATPAWP | PAWP>10 correlates with decreased peakVO2 and increased VE/VCO2-slope  |
|  |  |  |  |  |
| **SSc and prognosis** |
| 37 | Swigris JJ 2009 | 83 | SpO2SaO2 | Worse prognosis in SSc-ILD if saturation <89% or -4% at exercise |
| 42 | Poormoghim H 2011 | 91 | DLCO | In lcSSc later impaired than in dcSSc |
| 43 | Winstone TA 2014 | 1616 (meta-analysis) | DLCOFVC | Predictive in both SSc and SSc-ILD |
| 44 | Lefevre G 2013 | 2244 (meta-analysis) | DLCO6-MWDPAPmean | DLCO and pericardial effusion only prognostic factors in ILD-related PH |
| 45 | Trad S 2006 | 86 | DLCOTLCFVC | None of these parameters, but only PAH predicts survival in dcSSc |
| 47 | Launay D 2011 | 47 | DLCO | DLCO and pericardial effusion only prognostic factors in ILD-related PH |
| 52 | Coghlan JG 2014 | 73 | FVC/DLCO | DETECT and ASIG algorithms out-perform the ESC/ERS guidelines due to early inclusion of FVC/DLCO |
| 57 | Distler O, Behrens F | Expert consensus | 6-MWDSpO2at exercise | recommended in prognosis assessment |
| 59 | Ryerson CJ 2015 | 156 | 6-MWDFVC | Both predict 1-year mortality |
| 60 | Le Pavec J 2011 | 70 | 6-MWD | PH-Specific treatment does not change mortality in ILD-related PH |
| 61 | Zhao J 2017 | 190 | 6-MWD | Mortality in SSc-PAH higher than in Lupus erythematodes –PAH |
| 63 | Wensel R 2013 | 226 | peakVO2 | Predicts prognosis in PAH |

6-MWD: walking distance in 6 minutes; AADO2: alveolar-arterial difference of oxygen; CPET: cardiopulmonary exercise testing; DLCO: diffusion capacity of carbon monoxide;FEV1: forced expiratory volume in one second; FVC: forced vital capacity; ILD: interstitial lung disease; KCO: Krogh factor (DLCO per alveolar volume); lcSSc: limited cutaneous manifestation; PAH: pulmonary arterial hypertension; PAPmean:mean pulmonary arterial pressure; PAWP: pulmonary artery wedge pressure; peakVO2: peak oxygen uptake; petCO2: end tidal pressure of carbon dioxide; petCO2@AT: end tidal pressure of carbon dioxide at anaerobic threshold; VE/VCO2@AT: ratio of ventilation to carbon dioxide output at anaerobic threshold; VE/VCO2-slope: slope of the relation between ventilation and carbon dioxide output; VO2@AT: oxygen uptake at anaerobic threshold; VO2/HR: ratio of oxygen uptake to heart rate.