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**Title:** Evaluation and correlations of the extension of pulmonary cysts in lymphangioleiomyomatosis

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**Body:** Introduction: The best method to assess the extension of pulmonary cysts in HRCT and whether it correlates with six-minute walk test (6MWT) variables, metalloproteinases (MMPs) or vascular endothelial growth factor-D (VEGF-D) dosage have not been defined in lymphangioleiomyomatosis (LAM). Objectives: To study the extension of pulmonary cysts in HRCT and to determine its correlations with pulmonary function tests, MMPs and VEGF-D dosage, the walking distance and desaturation–distance ratio (DDR) in 6MWT in LAM (Pimenta, S.P. et al. Clinics 2010;65:841-6). Methods: Data from 23 women with LAM followed at the University of Sao Paulo were evaluated. The extension of pulmonary cysts in HRCT was assessed by computing the cyst volume over the entire lung volume. Spearman correlation coefficient was used to establish the association between the extension of cysts and the other variables. Results: The mean age was  $46 \pm 8$  years. The extension of cysts in HRCT was 6.8% (median). Mean  $FEV_1/FVC$ ,  $FEV_1$ ,  $D_LCO$  and  $RV/TLC$  were, respectively,  $0.68 \pm 0.19$ ,  $76 \pm 25\%$ pred,  $65 \pm 25\%$ pred and  $0.37 \pm 0.08$ . The distance walked was  $508 \pm 112$ m, while the DDR was 4 (median). VEGF-D serum level was 464 (median). The extension of cysts correlated best with  $D_LCO$  ( $r=-0.82$ ,  $p<0.0001$ ),  $FEV_1/FVC$  ( $r=-0.84$ ,  $p<0.0001$ ),  $RV/TLC$  ( $r=0.64$ ,  $p<0.0001$ ), and DDR ( $r=0.76$ ,  $p<0.0001$ ). A weak correlation was found with VEGF-D ( $r=0.45$ ,  $p=0.03$ ). There was no correlation with the walking distance or the level of MMPs. Conclusions: The severity of pulmonary cystic involvement was mild and correlated with airway obstruction, air trapping,  $D_LCO$ , and DDR. The use of VEGF-D and MMPs to assess the severity of LAM remains controversial.