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Quantitative analysis of airway obstruction in lymphangioleiomyomatosis

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This study demonstrates a 4-fold reduction in the number of airways and terminal bronchioles in end-stage LAM lungs using a combination of CT, microCT and histopathology, compared to a matched control group http://bit.ly/2tBTiJy

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ABSTRACT Lymphangioleiomyomatosis (LAM) is a rare, cystic lung disease with progressive pulmonary function loss caused by progressively proliferating LAM cells. The degree of airway obstruction has not been well investigated within the pathogenesis of LAM.

Using a combination of *ex vivo* computed tomography (CT), microCT and histology, the site and nature of airway obstruction in LAM explant lungs was compared with matched control lungs (n=5 each). The total number of airways per generation, total airway counts, terminal bronchioles number and surface density were compared in LAM *versus* control.

Ex vivo CT analysis demonstrated a reduced number of airways from generation 7 on ($p<0.0001$) in LAM compared with control, whereas whole-lung microCT analysis confirmed the three- to four-fold reduction in the number of airways. Specimen microCT analysis further demonstrated a four-fold decrease in the number of terminal bronchioles ($p=0.0079$) and a decreased surface density ($p=0.0079$). Serial microCT and histology images directly showed the loss of functional airways by collapse of airways on the cysts and filling of the airway by exudate.

LAM lungs show a three- to four-fold decrease in the number of (small) airways, caused by cystic destruction which is the likely culprit for the progressive loss of pulmonary function.