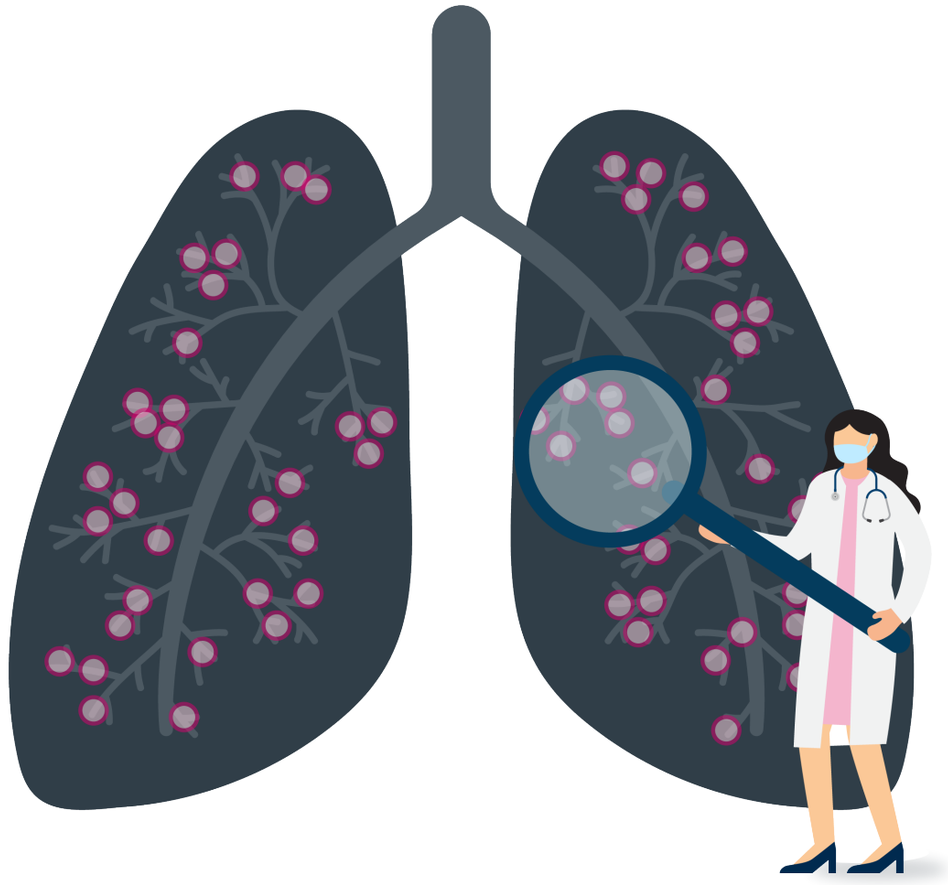
Guidelines for Diagnosing & Treating Lymphangioleiomyomatosis (LAM)

Always use the least invasive means for diagnosis.

Diagnosis

- CT findings of cystic lung disease alone are not sufficient to make a confirmed diagnosis of LAM.
- VEGF-D testing is useful for diagnosis and can help avoid lung biopsy.
- Other findings that can help establish a confirmed diagnosis of LAM include: presence of tuberous sclerosis complex (TSC), kidney angiomyolipomas, and lymphatic manifestations such as chylous effusions or lymphangioleiomyomas.
- In patients where non-invasive means have failed to provide a confirmed diagnosis, consider transbronchial lung biopsy before surgical lung biopsy.



Treatment

- Sirolimus is the first line treatment option for LAM patients with:
  1. Abnormal or rapidly declining lung function
  2. Substantial disease burden
  3. Problematic chylous effusions
- Do not use doxycycline or hormonal therapy for routine treatment of LAM.
- Offer pleurodesis following the initial episode of spontaneous pneumothorax rather than waiting for a recurrent event.
- Prior pleurodesis is not contraindication to lung transplantation.

McCormack FX, Gupta N, Finlay GA, et al. Am J Respir Crit Care Med. 194(6):748-761. Gupta N, Finlay GA, Kotloff RM, et al. Am J Respir Crit Care Med. 196(10):1337-1348.

For more information on LAM medical guidelines, point your phone camera to the QR code to the right, or visit: thelamfoundation.org/LAM-Treatment-Guidelines

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