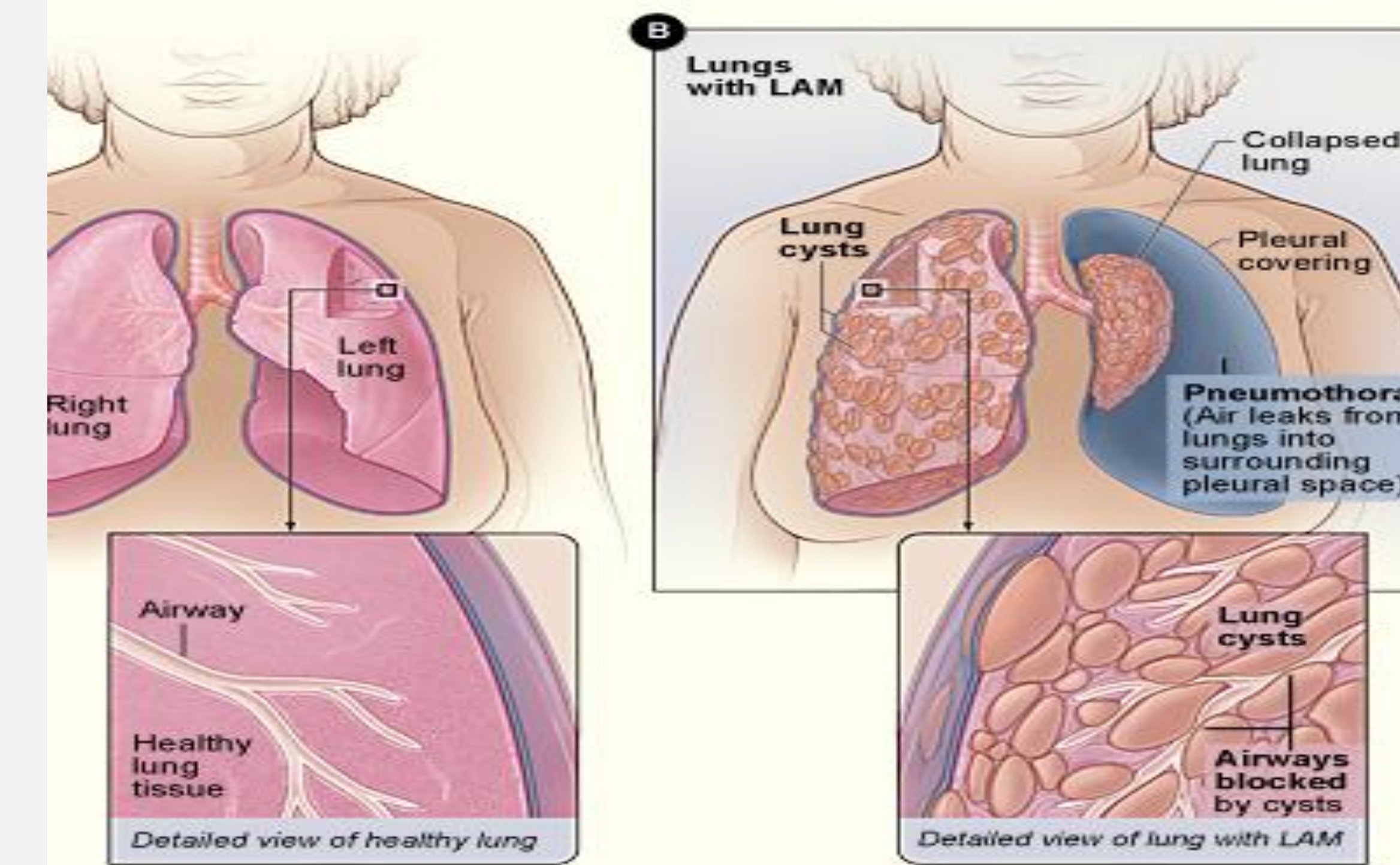


Peripartum Management of Lymphangioleiomyomatosis (LAM)

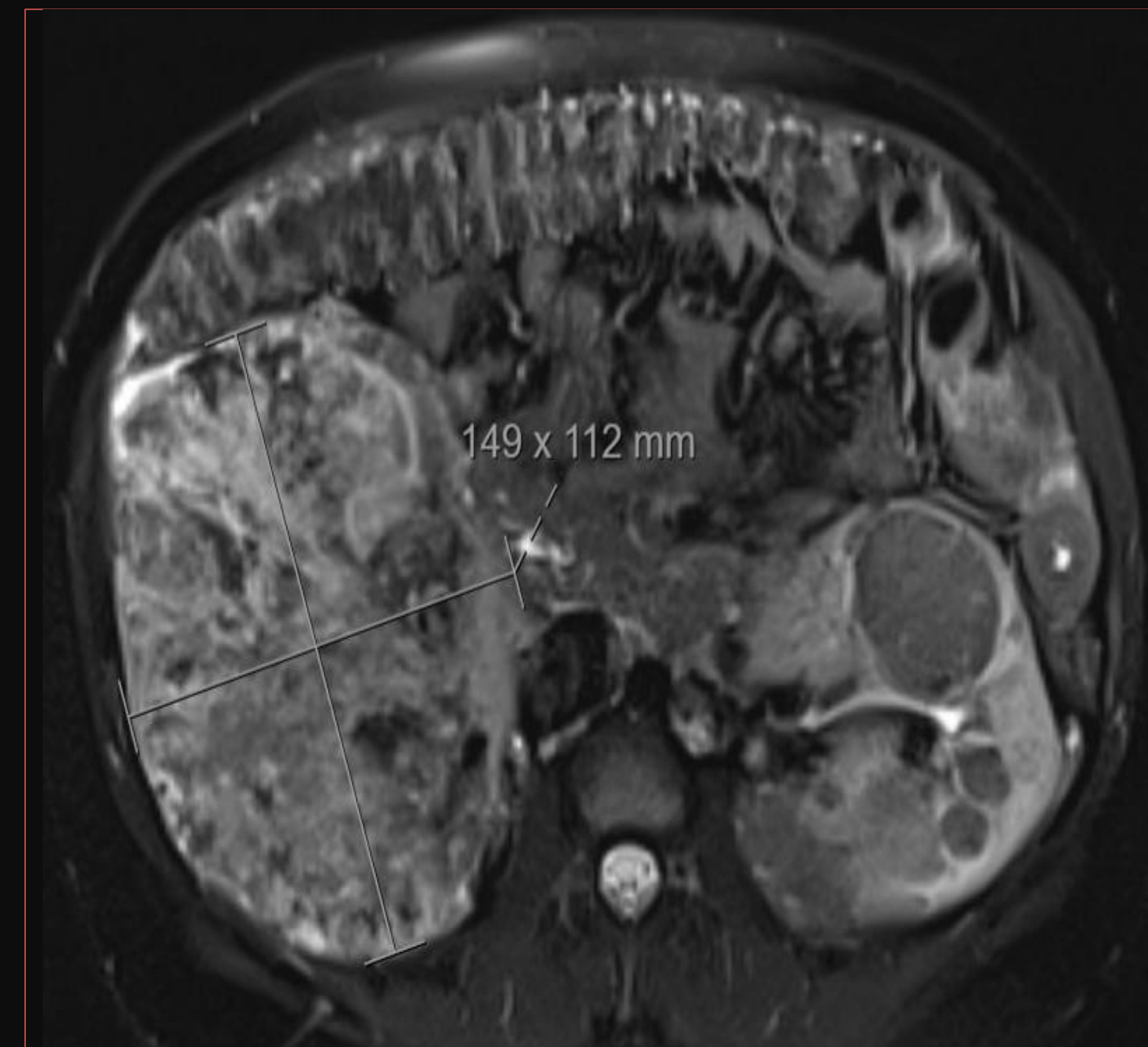
- LAM is a rare, sporadic, multisystem disease (~1:400,000) characterized by proliferation of abnormal smooth muscle-like cells that infiltrate the lungs, lymphatics, and kidneys.
- Two types of LAM:
 - Tuberous Sclerosis Complex-LAM-inherited genetic disorder
 - Sporadic LAM-Sporadic mutations occur in TSC ½ genes
- LAM leads to progressive cystic lung disease causing recurrent pneumothorax and may coexist with renal angiomyolipomas (AMLs). Symptoms include progressive dyspnea, chest pain, and fatigue
- Pregnancy is associated with increased disease activity and risks such as pneumothorax, AML rupture, and respiratory compromise
- Anesthetic considerations: multidisciplinary care and delivery planning, respiratory complications, potential hemorrhagic complications, avoid barotrauma, neuraxial preferred



High-resolution CT demonstrating diffuse bilateral thin-walled cysts consistent with cystic lung disease in lymphangioleiomyomatosis (LAM)

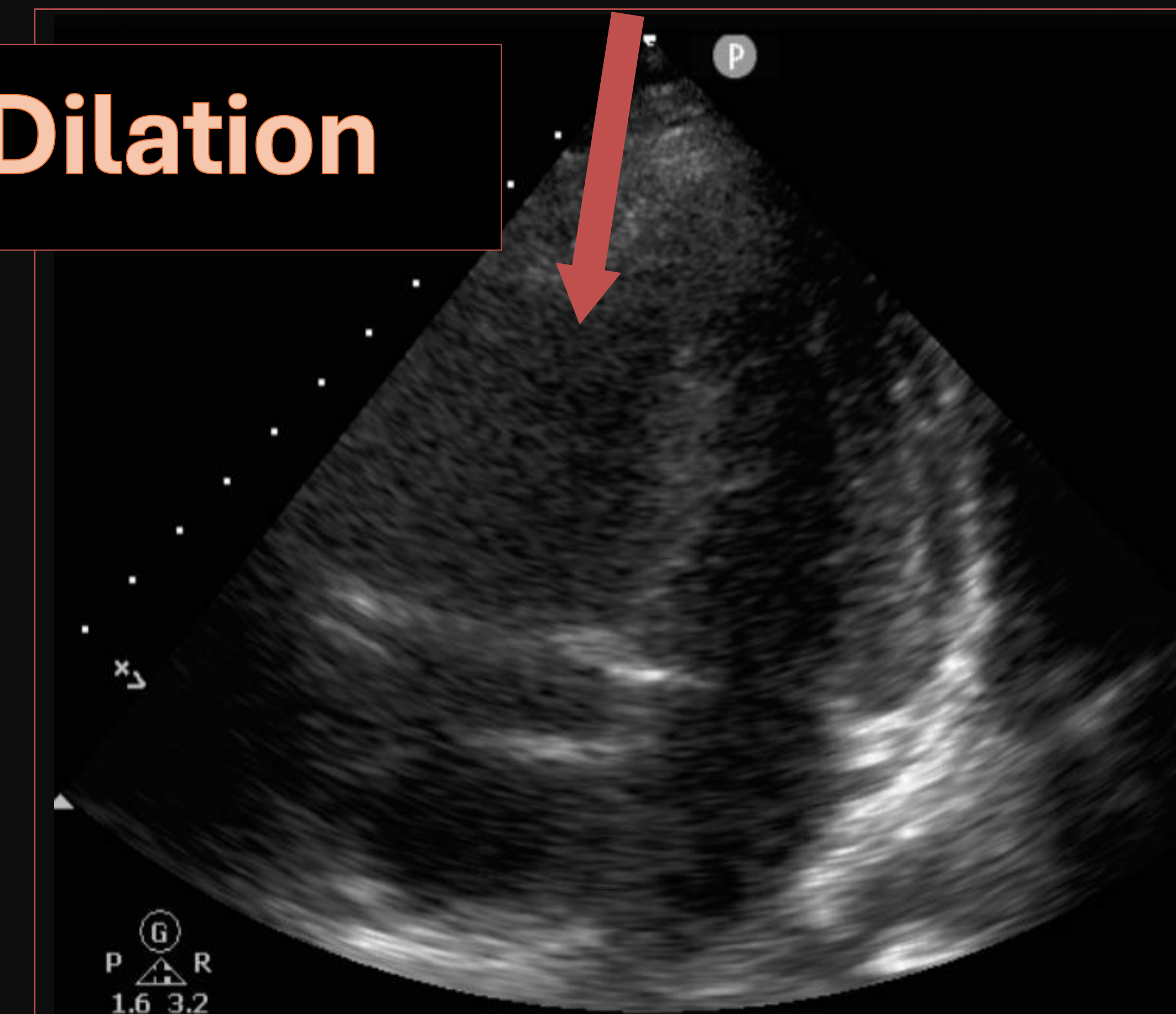
Case Presentation

- 32y/o F G2P1 at 32 weeks with bilateral AMLs (14.9 cm, 8.8 cm)
- Incidental mass → imaging → Right pneumothorax → chest tube prior to IR embolization
- Epidural + MAC to avoid positive pressure ventilation
- Acute hypoxia → pulmonary gel-foam embolization
- SICU admission → Acute Hypoxic Respiratory failure(HFNC) and heparinization
- Preterm vaginal delivery → recurrent pneumothorax
- Wedge resection + pleurodesis → final diagnosis: sporadic LAM



MRI abdomen demonstrating multiple bilateral renal masses consistent with angiomyolipomas; largest measuring 14.9 cm (right) and 8.8 cm (left).

RV Dilation



Apical four-chamber POCUS view demonstrating right ventricular dilation following suspected pulmonary gel-foam embolization.

Teaching Points

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- Suspect LAM in reproductive-age women with AMLs and pneumothorax
 - Pregnancy increases risk of pneumothorax and AML rupture (Hemorrhage)
 - Prefer neuraxial anesthesia; avoid nitrous oxide
 - Consider keeping epidural in place with heparinization
 - Chest tube in place until after delivery
 - If general anesthesia required, use lung protective ventilation strategies
 - Multidisciplinary planning and tertiary care essential