Langerhans cell histiocytosis

- More common in the paediatric population, with a peak incidence between one and three years of age [5].
- Incidence is estimated at ~5 per million children.
- 1-2 cases per million adults
- Male predilection (M:F ~ 1.5:1)
- Pediatric LCH not to be confused with adult pulmonary LCH (typically single system disease associated with smoking)

Etiology

- Neoplastic proliferation of monoclonal Langerhans cells leading to formation of destructive granulomas
- Childhood PLCH: Clonal cellular process unrelated to smoking
- Adult PLCH: Immune-mediated nonclonal proliferation related to smoking
  - Postulated to be allergic reaction to constituent of cigarette smoke
  - Smoke postulated to stimulate cytokine production, causing activation of Langerhans cells

Typical eponyms

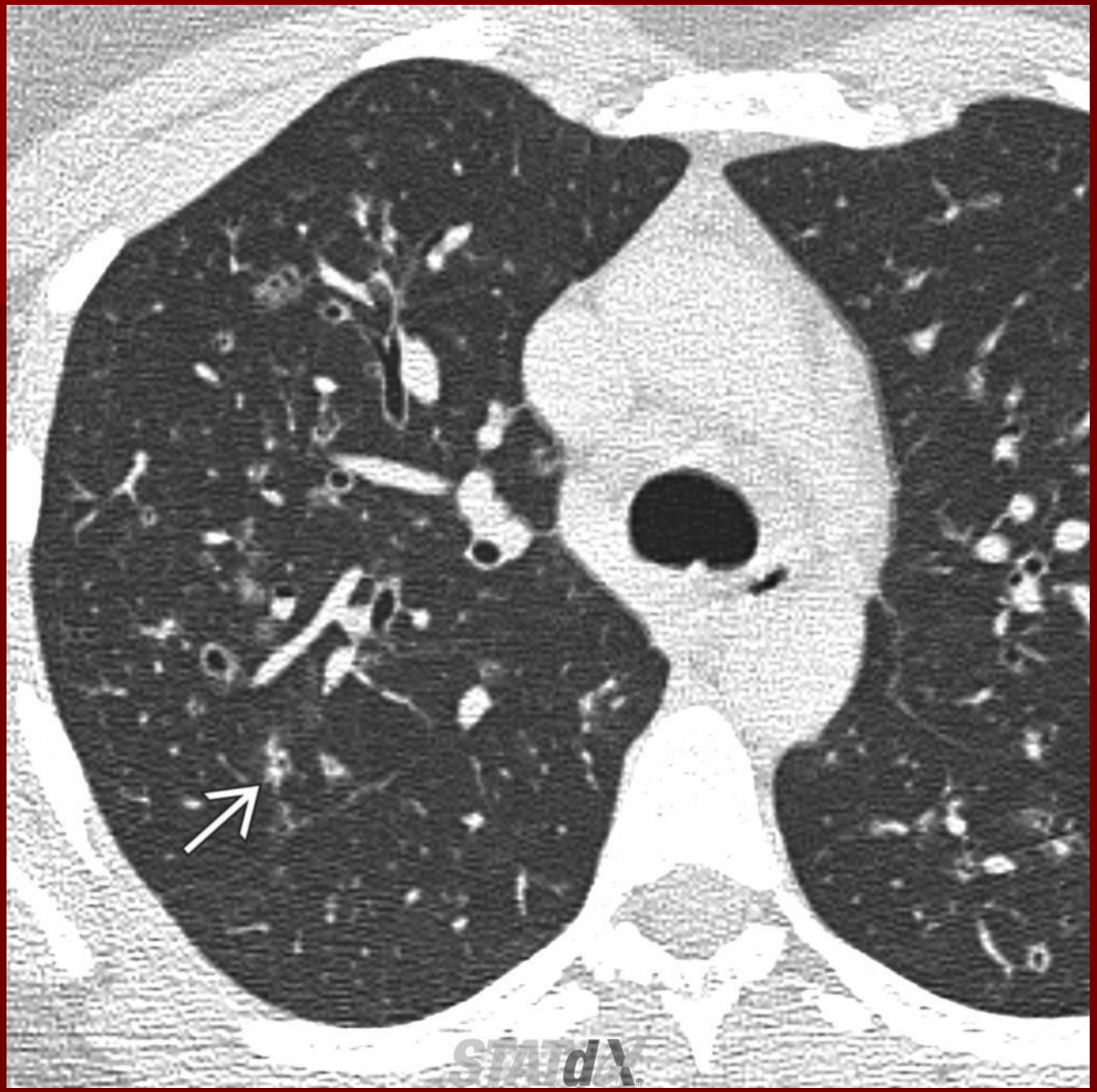
- Letterer-Siwe disease
  - disseminated multi-organ disease
  - typically young children/infants less than one year-old
  - fulminant course with poor prognosis
- Hand-Schüller-Christian disease
  - multiple lesions
    - some authors confine the term to patients with solitary organ involvement [4]
    - other authors accept multi-organ involvement (e.g. bone and spleen) [6]
  - confined to the one bone (usually bone)
  - typically children
  - intermediate prognosis
- eosinophilic granuloma (EG)
  - lesions are confined to one organ system
    - some authors confine the term to patients with a solitary lesion [4]
    - other authors accept multiple lesions [6]
  - 70% of cases affects bone
  - typically children
  - best prognosis

More Useful

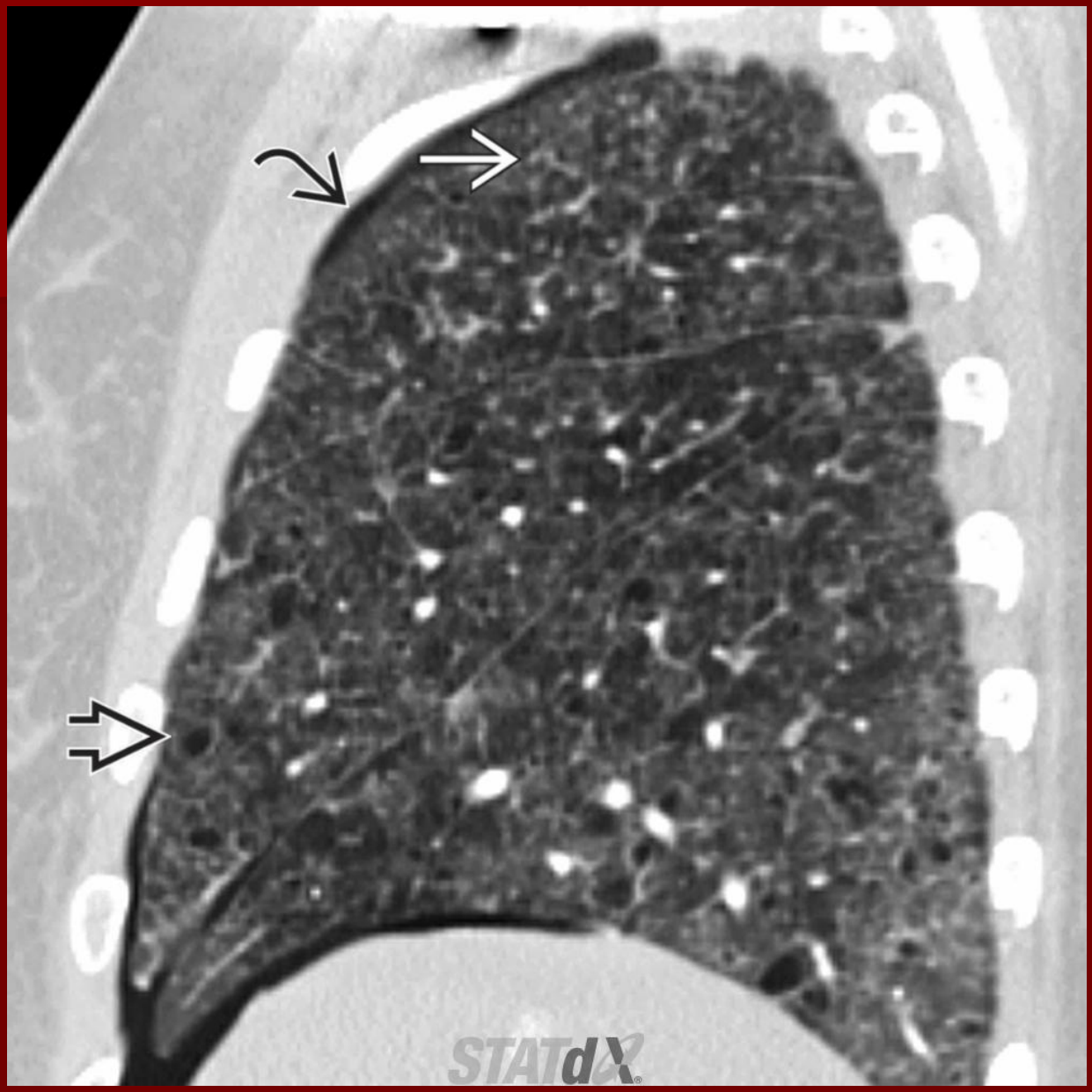
- Multiple organ systems, multiple sites involved
- Single-organ system, multiple sites involved
- Single lesion

CT

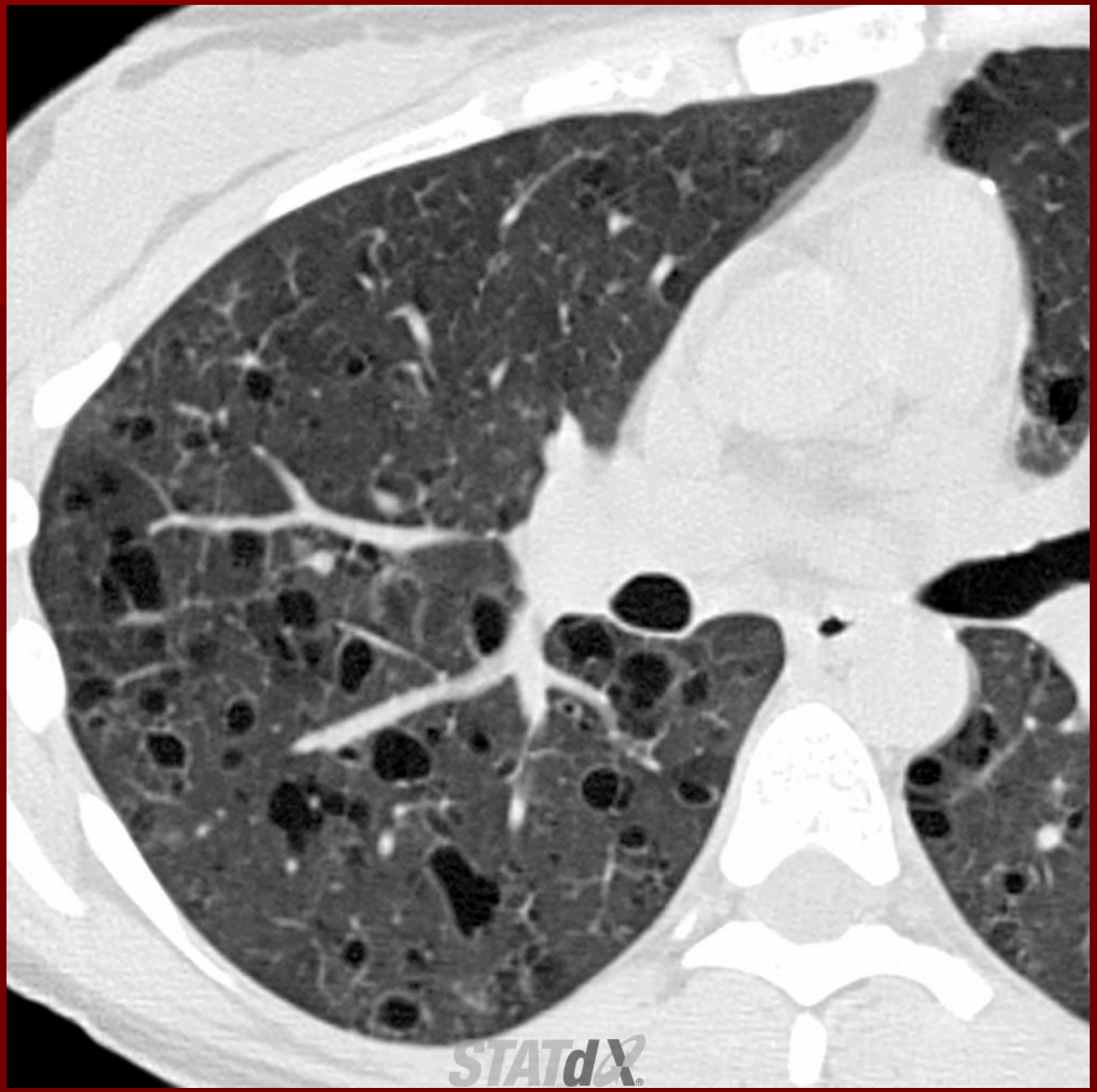
- Irregular, small nodules & cysts with upper/mid lung zone predominance
- Bronchiolocentric nodules, irregular/stellate borders, 1-10 mm in size
- Cysts: Variable sizes & bizarre shapes, thin or thick/nodular irregular walls



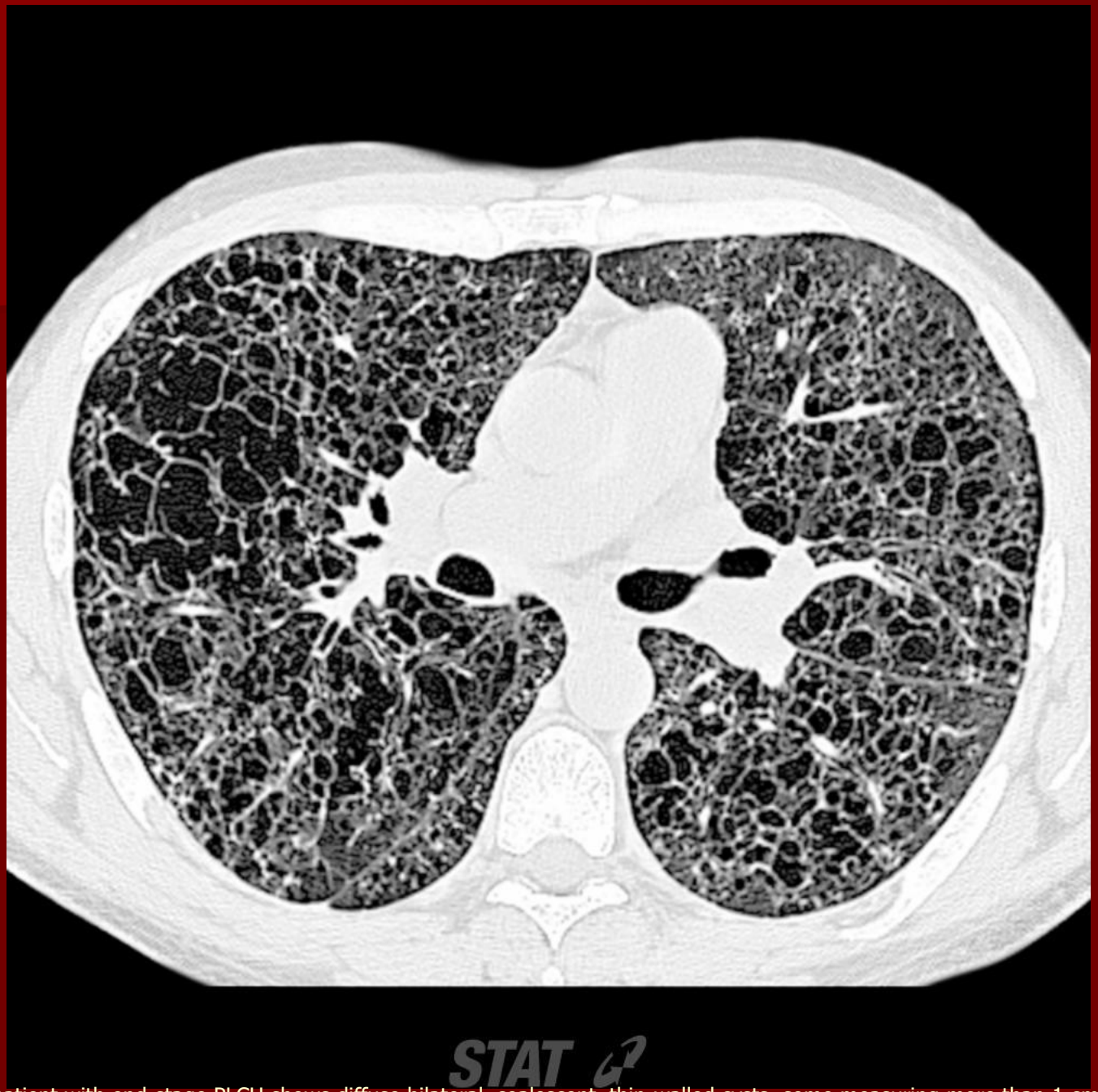
Axial NECT of the same patient demonstrates upper lung zone predominant small cavitary and noncavitary nodules with normal-appearing intervening lung parenchyma. Some of the noncavitary nodules (white solid arrow) exhibit a stellate morphology characteristic of PLCH lesions.



Sagittal NECT of a patient with PLCH who presented with acute chest pain shows profuse involvement of the lung with cysts (black open arrow), small nodules (white solid arrow), and a small anterior right-sided pneumothorax (black curved arrow). PLCH is a recognized cause of secondary spontaneous pneumothorax.



Axial NECT of a patient with PLCH demonstrates thin-walled pulmonary cysts of varying sizes, some with bizarre shapes. PLCH cysts may exhibit smooth or irregular cyst walls and may mimic other cystic lung diseases.



Axial HRCT of a patient with end-stage PLCH shows diffuse bilateral, coalescent, thin-walled cysts, some measuring more than 1 cm in diameter, and some partially septated and confluent.