The differentiation between congenital lung malformations (CLMs) and pleuropulmonary blastoma (PPB) is a diagnostic conundrum with potentially life-altering consequences. Congenital lung malformations are a group of diagnoses that include benign congenital pulmonary airway malformations (CPAMs), bronchogenic cysts, bronchopulmonary sequestrations, and congenital lobar emphysema. Congenital pulmonary airway malformations are the most common type of CLM. Congenital pulmonary airway malformations can be derived from the acinar structures of the lung and therefore may have a multiseptated cystic appearance strikingly similar to type I (purely cystic) PPB. In fact, the historic term type IV CPAM is now recognized as being the same clinical and pathologic entity as type I PPB.1 Pleuropulmonary blastomas are DICER1-familial cancer predisposition syndrome (OMIM 601200)-associated malignant embryonal tumors of the mesenchymal cells of the lungs and pleura. Pleuropulmonary blastomas start as type I (purely cystic) and can progress to type II (cystic and solid) or type III (purely solid) tumors, the latter 2 of which have the propensity for aggressive pleural spread and metastasis. While the standard of care for type I PPB is surgical resection with negative margins, debate remains in the pediatric surgical community about the optimal management of CLMs. For CLMs, some centers advocate for resection of all lesions owing to the potential for repeated infection, malignant degeneration, or misdiagnosis of CLM in the face of actual malignant tumors. In support of this approach, thoracoscopic lobectomy can be performed safely for infants with reduction of the short-term and long-term morbidity associated with open thoracotomy.2 In contrast, many centers advocate for observation of asymptomatic CLMs (particularly when diagnosed prenatally) because regression during infancy is possible, because asymptomatic lesions often remain asymptomatic throughout childhood, and to avoid the risks of surgery.3 A previous single-center retrospective study (in which the default management of asymptomatic CLM was observation) suggested that an algorithm based on prenatal vs postnatal diagnosis, radiographic appearance, and DICER1 germline testing can be used to differentiate between PPB and CLM with high sensitivity and specificity.5

A more recent study from the Midwest Pediatric Surgery Consortium demonstrated that 0 of 344 prenatally diagnosed cystic lung lesions were found to harbor malignant tumors on final pathological findings.6 However, for cystic lesions first diagnosed after birth, the risk of type I PPB approached 10%. This critical observation, enabled by multi-institutional collaboration, therefore refined the scope of the PPB vs CLM diagnostic conundrum to focus on postnatally diagnosed cystic lung lesions. The current article by Engwall-Gill et al7 is a follow-up study by the Midwest Pediatric Surgery Consortium with focused assessment of 40 computed tomography (CT) scans from infants with postnatally diagnosed cystic pulmonary lesions. This retrospective case-control study comprised a blinded assessment of preoperative CT scans from lesions determined to be type I PPB (cases) vs age-matched, postnatally detected CLMs (controls) by 9 experienced pediatric radiologists. The study demonstrated that the overall sensitivity and specificity for CT scan detection of PPB were 58% and 83%, respectively, with poor interrater reliability (κ = 0.36; P < .01). No specific imaging characteristics could be confirmed that differentiated PPB from CLM. Therefore, the authors concluded that CT scans are unreliable in differentiating CLM from PPB.

Given the inability to reliably differentiate CLM from PPB, these data provide additional support for operative management of cystic pulmonary lesions that are initially diagnosed after birth. These
data also suggest that DICER1 germline testing should be performed for infants with asymptomatic cystic pulmonary lesions first diagnosed after birth for whom the proposed management strategy is observation without surgical intervention. For cystic pulmonary lesions first appreciated on prenatal imaging, observation without DICER1 germline testing may continue to be a reasonable approach. Furthermore, these data may modulate surgical decision-making for postnatally diagnosed cystic lung lesions. In the circumstance of a postnatally diagnosed cystic lung lesion, an attempt to achieve negative margins, avoidance of tumor rupture, avoidance of specimen morcellation or division, and a lower threshold to start with or convert to an open approach should be considered.

ARTICLE INFORMATION
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REFERENCES