Thoracoscopic Lobectomy for Congenital Lung Lesions

Jarrett Moyer, MD[a,*], Hanmin Lee, MD[b], Lan Vu, MD[b]

KEYWORDS

• Congenital • Lung • Lesion • Malformation • Thoracoscopic • Resection
• Lobectomy • CPAM

KEY POINTS

- Most congenital lung lesions are diagnosed on prenatal ultrasound.
- Most fetuses with congenital lung lesions proceed to uncomplicated term delivery.
- Thoracoscopic lobectomy is safe, well-tolerated, and results in a shorter length of hospital stay compared with open resection.
- Symptomatic neonates should undergo resection shortly after birth.
- Asymptomatic neonates can be observed and undergo prophylactic resection at 6 months of life or later.

INTRODUCTION

Congenital lung lesions (CLLs) comprise a heterogeneous collection of rare developmental parenchymal lung abnormalities present in utero and at birth, including congenital pulmonary airway malformation (CPAM), bronchopulmonary sequestration (intralobar and extralobar), bronchial atresia, and congenital lobar emphysema. Additionally, pleuropulmonary blastoma (PPB), mediastinal teratoma, and bronchogenic cysts can mimic CLLs on fetal ultrasonography, postnatal radiograph, and postnatal contrast-enhanced computed tomography (CT) or MRI. CLLs are often diagnosed on prenatal ultrasound and have overlapping radiologic features. Thus, definitive diagnosis relies on histopathologic analysis of resected tissue.[1] Given the heterogeneity of CLL, it is not surprising that these lesions carry unpredictable and vastly different clinical outcomes if left untreated. On one hand, CLLs can produce mediastinal shift,

Disclosures: The authors have nothing to disclose.
[a] Department of Surgery, University of CA - San Francisco, 513 Parnassus Avenue, S-321, San Francisco, CA 94143, USA; [b] Division of Pediatric Surgery, Department of Surgery, University of CA - San Francisco, 550 16th Street, 5th Floor, UCSF Box 0570, San Francisco, CA 94143, USA
* Corresponding author.
E-mail address: jarrett.moyer@ucsf.edu

Clin Perinatol ■ (2017) ■–■
http://dx.doi.org/10.1016/j.clp.2017.08.003
perinatology.theclinics.com

polyhydramnios, and fetal nonimmune hydrops in 5% to 30% of cases,[2,3] requiring prenatal intervention to avoid fetal demise. On the other hand, approximately 1 in 5 cases are diagnosed outside of the neonatal period incidentally or because of infection or pneumothorax.[4] Finally, in utero behavior of CLLs is variable, with an early proliferative phase producing a peak in size around 25 weeks' gestation, often followed by size regression in the third trimester. In a case series of 600 CLLs, 68% of pulmonary sequestration and 15% of CPAM underwent marked spontaneous regression before birth.[2] Accordingly, the rarity of CLLs, heterogeneity in lesion histology for prenatally diagnosed lung lesions, and an unpredictable and widely variable natural history make the development of evidence-based treatment algorithms difficult. Thus, although the development of hydrops mandates prenatal intervention, and symptomatic neonates clearly benefit from resection, optimal treatment of the asymptomatic CLL is less clear and is determined on a case-by-case basis with the guidance of case series and expert opinion.

In patients who warrant surgical intervention, formal lobectomy is recommended over segmental resection.[5,6] Case reports and series have demonstrated precursors to mucinous bronchioloalveolar carcinoma harbored in CPAM type 1[7,8]; PPB can be indistinguishable from CPAM type 4, resulting in inadequate surgical margins and risk of recurrence with segmental resection.[9] Additionally, determining lesion margins both preoperatively on CT scan and intraoperatively is difficult, resulting in a high risk of incomplete resection with nonanatomic resection.[4,5] Traditionally, lobectomy occurred as an open surgical procedure through posterolateral thoracotomy. However, Albanese and colleagues[10] first described a completely thoracoscopic minimally invasive lobectomy in 2003. Subsequent case series and a meta-analysis have shown the thoracoscopic technique to provide improved or equivalent complication rates, decreased hospital length of stay, and decreased time of tube thoracostomy when compared with open techniques.[11–15] The focus of this article is on the indications, surgical approach, technical considerations, postoperative care, and outcomes of thoracoscopic resection of CLLs, in particular CPAM.

INDICATIONS/CONTRAINDICATIONS

As mentioned earlier, the clinical presentation of antenatally diagnosed CLLs varies widely, from fetal nonimmune hydrops to asymptomatic term live birth extending into childhood. Large lesions can produce mediastinal shift, cardiac compression, and obstruction of the vena cava, resulting in profound hemodynamic alterations and development of hydrops in the fetus. Lesions can be risk stratified according to the cystic adenomatoid volume ratio (CVR), which normalizes lesion volume to head circumference and predicts an 80% risk of hydrops for a CVR greater than 1.6.[16,17] Fetuses with a CVR greater than 1.6 should be monitored with weekly ultrasound examinations, as hydrops is associated with near 100% fetal mortality if left untreated.[3,18] Treatment strategies include maternal betamethasone administration as well as invasive techniques, such as fetal lobectomy via maternal laparotomy and hysterotomy, thoracoamniotic shunting, radiofrequency or laser ablation, or percutaneous ultrasound-guided sclerotherapy.[19] Steroid administration is especially effective in treating microcystic lesions producing nonimmune hydrops, with survival rates to delivery as high as 92%.[20–22] Given the effectiveness of prenatal steroids in the treatment of hydrops, there are now fewer clinical scenarios whereby invasive fetal intervention is indicated.

Most fetuses with a prenatal diagnosis of CLL proceed to live birth without the development of hydrops. Of these neonates, roughly one-quarter will be symptomatic