Current Operative Management of Congenital Lobar Emphysema in Children: A Report from the Midwest Pediatric Surgery Consortium

Shaun M. Kunisaki, MD, MSc, FAAP[1]; Jacqueline M. Saito, MD, MSCI, FAAP[2]; Mary E. Fallat, MD, FAAP[3]; Shawn D. St. Peter, MD, FAAP[4]; Aimee G. Kim, MD[1]; Kevin N. Johnson, MD[1]; Rodrigo A. Mon, MD[1]; Cheryl Adams[5]; Bola Aladegbami, MD[2]; Christina Bence, MD[6]; R. Cartland Burns, MD, FAAP[7]; Kristine S. Corkum, MD[8]; Katherine J. Deans, MD, MHSc, FAAP[9]; Cynthia D. Downard, MD, FAAP[3]; Jason D. Fraser, MD, FAAP[4]; Samir K. Gadepalli, MD, MBA, FAAP[1]; Michael A. Helmrath, MD, FAAP[5]; Rashmi Kabre, MD, FAAP[8]; Dave R. Lal, MD, MPH, FAAP[6]; Matthew P. Landman, MD, MPH, FAAP[7]; Charles M. Leys, MD, FAAP[10]; Allison F. Linden, MD, MPH[11]; Joseph J. Lopez, MD[9]; Grace Z. Mak, MD, FAAP[11]; Peter C. Minneci, MD, MHSc, FAAP[9]; Brooks L. Rademacher, MD[10]; Aimen Shaaban, MD, FAAP[5]; Sarah K. Walker, MD[6]; Tiffany N. Wright, MD[3]; Ronald B. Hirschl, MD, FAAP[1]; on behalf of the Midwest Pediatric Surgery Consortium

Author Affiliations:
[1]Section of Pediatric Surgery, Department of Surgery, University of Michigan and Michigan Medicine, C.S. Mott Children's and Von Voigtlander Women's Hospital, Ann Arbor, MI, USA
[2]Division of Pediatric Surgery, Department of Surgery, Washington University School of Medicine, St. Louis, MO, USA
[3]Division of Pediatric Surgery, Hiram C. Polk, Jr., M.D. Department of Surgery, University of Louisville, Norton Children's Hospital, Louisville, KY, USA
[4]Division of Pediatric Surgery, Department of Surgery, Children's Mercy Hospital, Kansas City, MO, USA
[5]Division of Pediatric Surgery, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA
[6]Division of Pediatric Surgery, Department of Surgery, Medical College of Wisconsin, Milwaukee, WI, USA
[7]Division of Pediatric Surgery, Department of Surgery, Indiana University School of Medicine, Indianapolis, IN, USA
[8]Division of Pediatric Surgery, Department of Surgery, Ann & Robert H. Lurie Children's Hospital of Chicago, Northwestern University Feinberg School of Medicine, Chicago, IL, USA
[9]Center for Surgical Outcomes Rsearch, The Research Institute and Department of Surgery, Nationwide Children's Hospital, Ohio State University College of Medicine, Columbus, OH, USA
[10]Division of Pediatric Surgery, Department of Surgery, University of Wisconsin, Madison, WI, USA
[11]Section of Pediatric Surgery, Department of Surgery, Comer Children's Hospital, University of Chicago Medicine and Biological Sciences, Chicago, IL, USA

Corresponding Author:
Shaun M. Kunisaki, MD, MSc, FAAP
Associate Professor of Surgery
Johns Hopkins Children's Center
1800 Orleans Street, Suite 7353
Baltimore, MD 21287-1005
skunisa1@jhmi.edu

Disclosure Statement: This project was supported by intramural funding from each institution within the Midwest Pediatric Surgery Consortium (www.mwpsc.org). No competing financial interests exist, and all authors have nothing to disclose.

This is the author's manuscript of the article published in final edited form as:
Kunisaki, S. M., Saito, J. M., Fallat, M. E., St. Peter, S. D., Kim, A. G., Johnson, K. N., … Hirschl, R. B. (2019). Current Operative Management of Congenital Lobar Emphysema in Children: a Report from the Midwest Pediatric Surgery Consortium. Journal of Pediatric Surgery. https://doi.org/10.1016/j.jpedsurg.2019.02.043

Abstract

Purpose: The purpose of this study was to evaluate the clinical presentation and operative outcomes of patients with congenital lobar emphysema (CLE) within a large multicenter research consortium.

Methods: After central reliance IRB-approval, a retrospective cohort study was performed on all operatively managed lung malformations at eleven participating children's hospitals (2009-2015).

Results: Fifty-three (10.5%) children with pathology-confirmed CLE were identified among 506 lung malformations. A lung mass was detected prenatally in 13 (24.5%) compared to 331 (73.1%) in non-CLE cases ($p<0.0001$). Thirty-two (60.4%) CLE patients presented with respiratory symptoms at birth compared to 102 (22.7%) in non-CLE ($p<0.0001$). The most common locations for CLE were the left upper (n=24, 45.3%), right middle (n=16, 30.2%), and right upper (n=10, 18.9%) lobes. Eighteen (34.0%) had resection as neonates, 30 (56.6%) had surgery at 1-12 months of age, and five (9.4%) had resections after 12 months. Six (11.3%) underwent thoracoscopic excision. Median hospital length of stay was 5.0 days (interquartile range, 4.0-13.0).

Conclusions: Among lung malformations, CLE is associated with several unique features, including a low prenatal detection rate, a predilection for the upper/middle lobes, and infrequent utilization of thoracoscopy. Although respiratory distress at birth is common, CLE often presents clinically in a delayed and more insidious fashion.

Abbreviations: CLE, congenital lobar emphysema; CLM, congenital lung malformation CPAM, congenital pulmonary airway malformation; BPS, bronchopulmonary sequestration; LUL, left upper lobe; RML, right middle lobe

Keywords: congenital lobar emphysema, congenital pulmonary airway malformation, congenital lung malformations

Level of Evidence: Level III

Introduction

Congenital lobar emphysema (CLE), also known as congenital lobar overinflation, is a rare but well described congenital lung malformation (CLM) known to cause progressive hyperinflation and air trapping within one or more pulmonary lobes [1-3]. Because CLE can lead to profound respiratory distress and even cardiac arrest, thoracotomy and lobectomy have been the mainstay treatment of choice in infants with severe symptomatic disease [4, 5]. Although the histologic characteristics of the affected lobe can be quite variable in CLE [6, 7], the unifying embryological basis may be related to transient bronchial obstruction during the canalicular phase of lung morphogenesis [8, 9].

Over the past decade, there have been a number of innovations that have made a significant impact on the contemporary clinical diagnosis and management of CLMs. These advances range from the enhanced resolution of prenatal ultrasound technology, which has led to improved detection rates and family counseling, to the increasing popularity of thoracoscopic techniques amongst the ranks of pediatric surgeons [10-12]. There has also been a growing body of work supporting the non-operative management of asymptomatic CLMs, based on the rationale that infection and malignant degeneration are relatively rare complications in patients followed without resection [13-15]. To date however, there has been relatively little focus on the diagnosis and management of CLE with respect to these changes in the field [16]. Given the relative paucity of published case series on CLE [1, 2, 17, 18], there remains a need to better understand this CLM if we are to develop evidence-based treatment recommendations that will optimize clinical outcomes in affected children [16, 19].

In this study, we sought to evaluate the clinical presentation and operative outcomes in a large cohort of pediatric CLE patients. Our group hypothesized that children with CLE represent a unique subgroup of CLM patients characterized by infrequent prenatal diagnosis, high rate of neonatal resection, and liberal use of open surgical techniques.

Methods

A central reliance agreement was approved by the institutional review boards (IRB) at each of the eleven hospitals associated with the Midwest Pediatric Surgery Consortium (wwww.mwpsc.org)[20]. We performed a multi-institutional retrospective cohort study of patients less than 18 years of age who underwent a resection of a pathologically-confirmed lung malformation between January 1, 2009 and December 31, 2015. Patients were identified for study inclusion based on Current Procedural Terminology (CPT) codes corresponding to removal of lung tissue (32140, 32440, 32480-4, 32505, 32662-71). We then used the International Classification of Disease, 9th Edition, Clinical Modification (ICD-9-CM) diagnosis code for CLE (770.2) to identify patients and compared them with those with codes for congenital pulmonary airway malformation (CPAM, 748.4), bronchopulmonary sequestration (BPS, 748.5), and other congenital lung lesions (748.6). At each institution, the pathology reports were reviewed to confirm the appropriate diagnosis. We have previously reported on the entire cohort of CLM patients elsewhere (JPS manuscript submitted).

Data for collection, including demographics, maternal records, birth history, prenatal and postnatal diagnostic imaging, operative approach and findings, and

postoperative morbidity and mortality, were entered and managed using REDCap (Research Electronic Data Capture, 8.1.20, Vanderbilt University, Nashville, TN) software hosted at the Michigan Institute for Clinical & Health Research at the University of Michigan. All study data entered were validated by trained reviewers at both central and institutional levels for completeness and accuracy.

Frequencies and percentages were used to describe categorical variables. Median and interquartile ranges were generally used to describe continuous variables. Statistical analysies were performed using STATA (StataCorp, College Station, TX) and Prism (GraphPad Software, La Jolla, CA). Significance was defined as $p<0.05$.

Results

3.1. Demographics

Of 506 lung malformation resections entered into the data registry, 53 (10.5%) were pathology-confirmed CLE (Figure 1A). There was marked variability in the number of CLE cases contributed by institution (Figure 1B). Five CLE specimens also had histologic evidence of another CLM, including CPAM (n=3, 5.6%), bronchogenic cyst (n=1, 1.9%), and extralobar BPS (n=1, 1.9%).

Figure 1

Thirty-two (60.4%) were male. Thirty-seven (69.8%) were of white/Caucasian descent, and 9 (17.0%) were non-white. Twenty-five (49.0%) had private insurance, and 19 (37.3%) were Medicaid or self-pay.

3.2. Perinatal characteristics

Prenatal detection was documented in only 13 (24.5%) of the CLE cases compared to 331 (73.1%) of the non-CLE cases ($p<0.0001$). In prenatal lesions that subsequently revealed CLE, the ultrasound diagnosis was CPAM (n=11, 84.6%), BPS (n=1, 7.7%), or unspecified (n=1, 7.7%). The mean CVR among fetal CLE lesions was 0.65±0.41, and there were no cases of fetal hydrops.

The median gestational age at birth was 39.0 weeks [interquartile range (IQR), 37.0-39.0). The median 1-minute and 5-minute Apgar scores were 7 and 9, respectively. Eleven (20.8%) were inborn. Thirty-two (60.4%) neonates with CLE presented with respiratory symptoms at birth compared to 102 (22.7%) non-CLE neonates with respiratory symptoms ($p<0.0001$). However, prenatally diagnosed children with CLE were no more likely to be symptomatic at birth compared to prenatally diagnosed children with non-CLE lesions (30.0% vs. 21.9%, respectively; $p=1.0$). Fifteen (46.9%) of the symptomatic neonates with CLE were supported by positive pressure mechanical ventilation prior to surgical resection.

Initial neonatal chest radiograph data were available in 40 (75.4%) children. The initial image demonstrated a parenchymal lucency in 36 (90.0%) cases. Additional radiographic findings included mediastinal shift (n=7, 17.5%) and pneumothorax (n=5, 12.5%). One (2.5%) child had a neonatal study that was interpreted as completely normal.

3.3. Preoperative evaluation

Fifty (94.3%) children with CLE had a pre-operative CT scan at a median age of 4.6 weeks (IQR, 1.6-10.7). Thirty-seven (74.0%) studies correctly predicted the

Figure 2A

pathologic diagnosis. CLE most commonly involved the left upper lobe (LUL; n=24, 45.3%), followed by right middle lobe (RML; n=16, 30.2%), and right upper lobe (n=10, 18.9%)(Figure 2A). The most common indication for operative intervention was respiratory symptoms (77.4%), which included tachypnea, chronic cough, or need for mechanical ventilatory support. The risk of having respiratory symptoms was not associated with the location of the affected lobe (LUL: 60.9%, RML: 86.7%; p=0.14). Almost one-third (31.7%) of symptomatic CLE children had surgery after 12 weeks of age. Other indications for surgery were to prevent future complications in an otherwise asymptomatic patient (n=10, 18.9%), prior pneumonia (n=6, 11.3%), failure to thrive (n=6, 11.3%), and prior pneumothorax (n=1, 1.9%). Nine (17.0%) children had preoperative co-morbidities, including pulmonary hypertension (n=2), structural heart disease (n=1), esophageal atresia with distal tracheoesophageal fistula (n=1), and congenital diaphragmatic hernia (n=1).

3.4. Operative data

Figure 2B

The age distribution at operative resection is shown in Figure 2B. Eighteen (34.0%) underwent resection during the neonatal period, 30 (56.6%) had surgery at 1-12 months of age, and five (9.4%) had operations after 12 months. Three (5.6%) children with CLE underwent surgical resection within the first week of life. The overall median age at resection was 11.1 weeks (IQR, 3.0-27.0). The median operative age in CLE cases that had respiratory symptoms at birth was 4.2 weeks (IQR, 2.0-13.5) compared to 29.6 weeks (IQR, 11.1-42.9) in those born asymptomatic (p<0.0001).

The median weight at operation was 4.8 kg (IQR, 3.5-7.5). Fifty-one (96.2%) lobectomies were performed. Mean operative duration was 160.7±76.9 minutes. Approximately one-third of cases utilized mainstem intubation (n=19, 35.8%), whereas another one-third of cases had no lung isolation (n=17, 32.1%). Surgeons favored open thoracotomy (n=45, 84.9%) over thoracoscopic approaches (n=6, 11.3%). Two additional resections were initially attempted by thoracoscopy but were converted to thoracotomy, yielding a conversion rate of 25.0%. In both cases, the reason for conversion to an open procedure was inadequate exposure. The conversion rate in non-CLE thoracoscopic resections was lower (15.1%) but not statistically significant (p=0.36). Some cases required an intraoperative blood transfusion (n=13, 23.1%). Approximately half of patients were extubated immediately after surgery (n=27, 51.9%).

3.5. Postoperative course

Thirty-four patients (65.4%) were monitored in the intensive-care unit postoperatively. Median chest tube duration and hospital length of stay were 4.0 days (2.0-5.0) and 5.0 days (IQR, 4.0-13.0), respectively. Fourteen (26.4%) children had at least one complication within 30 days after operative resection (Table 1). The most common complication was intubation greater than 48 hours (n=7, 13.2%), followed by pneumothorax necessitating tube thoracostomy (n=4, 7.5%). The overall mortality rate was 1.9%. One premature infant born at 25.5 weeks died from respiratory failure and sepsis following an open lobectomy performed at eight weeks of age.

Table 1

4. Discussion

Although CLE was originally described in 1932 [21], the term was first coined in a series of patients by Robertson and James in 1951 [22]. CLE can lead to progressive air trapping and hyperinflation within the affected lobe due to a ball valve effect, resulting in compression atelectasis of normal lung parenchyma and mediastinal shift. The etiology of the abnormal lung tissue is idiopathic in some cases but may also be secondary to intrinsic or extrinsic bronchial obstruction as well as polyalveolar lobe [6, 7, 23]. Historically, children with CLE have been thought to present with respiratory symptoms within days to weeks after birth, and the majority have been diagnosed by six months of age [1, 24]. Acute and rapidly worsening air trapping at birth during normal postnatal breathing, which can be exacerbated by intratracheal positive pressure ventilation, can result in tension pneumothorax-like physiology requiring emergent thoracotomy [25]. However more recently, CLE has been shown to have a much wider spectrum of segmental and lobar disease given that asymptomatic cases and those with minimal pulmonary symptoms have been managed successfully with serial observation alone [2, 17, 26, 27].

In this study, we evaluated the largest known series to date of pathologically-confirmed CLE cases. These CLEs were drawn from a multi-institutional registry of 506 operative CLMs, thereby constituting approximately 10% of all lung lesions. We found that surgical resection was associated with low mortality and that clinical outcomes were generally excellent. However, several interesting findings from this analysis were noted. First, the prenatal diagnosis of CLE was significantly less common compared to that of non-CLE lesions, despite widespread prenatal screening as well as ongoing improvements in ultrasound image quality [28-30]. While a majority of non-CLE cases

were identified prenatally, only a quarter of CLEs in our series were visualized by antenatal ultrasound. Moreover, none of the fetal lesions that were subsequently found to be CLE were diagnosed as such by ultrasound, perhaps reflecting their nonspecific echogenicity and overall rarity of CLE compared to microcystic CPAMs [31-33]. Apparent involution of CLEs, similar to the behavior observed in some microcytic CPAMs, has been described [34, 35], but infants with CLE may still encounter severe respiratory distress at birth [36, 37]. Fetal MRI may be beneficial in helping to narrow the differential diagnosis of CLMs but does not have the diagnostic accuracy of postnatal CT [38]. Since our data suggests that neonates with prenatally detected cases of CLE were no more likely to be symptomatic at birth compared to neonates with prenatally detected non-CLE lesions, it is unlikely that the addition of fetal MRI to the diagnostic evaluation would help to facilitate an alternative delivery plan compared to ultrasound alone.

A second important finding from our study was that, despite our relative inability to image CLE prenatally, there was a nearly three-fold increased risk for respiratory symptoms at birth in patients with CLE when compared to those with non-CLE lesions. It was therefore not surprising that approximately one-third of CLE children underwent operative resection in the neonatal period compared to a 10% neonatal resection rate in non-CLE patients. Nevertheless, our series also showed that only three (5.7%) patients required intervention within the first week of life. Based on these data, in conjunction with initial plain films suggesting the absence of mediastinal shift in the majority of affected neonates, the more common clinical scenario in CLE appears to be a more mild, chronic form of respiratory distress. Alternatively, it is possible that some of these

infants had progressive air trapping within the affected lobe over time, leading to a delayed onset of symptoms. Although some investigators have identified a high association between CLE and congenital heart disease [17, 39, 40], the low incidence of cardiac anomalies reported in our series does not support the routine use of echocardiography in the preoperative planning of these patients.

When CLE resections are performed beyond the neonatal period, our data also revealed that surgeons overwhelmingly still favored open thoracotomy over thoracoscopy in the operative management of these lesions. These findings are consistent with work published elsewhere [32, 41]. Features that make thoracoscopic lobectomy particularly difficult in CLE, when compared to CPAM and BPS, include the inability to substantially deflate the air trapped lobe to create adequate working space as well as the increased technical difficulty associated with upper and middle lobe resections. Transient in utero left upper lobe bronchial compression by the ductus arteriosis has been proposed as one theory for the predominance of left upper lobe CLE [42]. Our study also showed that one-quarter of lesions approached by thoracoscopy were converted to open, but this was not statistically significant when compared to thoracoscopic resections for non-CLE lesions.

Despite the aforementioned data presented here confirming our hypotheses on CLE, several caveats of this study deserve mention. First, these data were extracted from a retrospective cohort registry that is limited by the quality of the data present in the electronic medical record at each institution [43]. Second, although less susceptible to referral bias than large, single institution studies, our results may still not be generalizable to other hospitals, particularly smaller pediatric surgery centers or

institutions in other parts of the world. Finally, because this is an operative cohort registry, some CLE patients were obviously excluded, such as those who never had surgery due to non-operative management or those who may have died in the early postnatal period prior to resection. Based on work by others [2, 17, 26], there is likely a subset of CLE infants whose respiratory symptoms resolved or who remained asymptomatic in the long term. To better inform best practice, we are currently exploring prospective methods to evaluate operative and non-operative CLM outcomes within our consortium.

Among lung malformations, CLE is associated with several unique features, including a low prenatal detection rate, a predilection for the upper/middle lobes, and infrequent utilization of thoracoscopy. Although respiratory distress at birth is common, CLE often presents clinically in a delayed and more insidious fashion.

REFERENCES

[1] Hendren WH, McKee DH. Lobar emphysema of infancy. J Pediatr Surg 1966;1(1):24-38.

[2] Ozcelik U, Gocmen A, Kiper N, Dogru D, Dilber E, Yalcin EG. Congenital lobar emphysema: evaluation and long-term follow-up of thirty cases at a single center. Pediatric pulmonology 2003;35(5):384-91.

[3] Adzick NS. Management of fetal lung lesions. Clin Perinatol 2009;36(2):363-76, x.

[4] Man DW, Hamdy MH, Hendry GM, Bisset WH, Forfar JO. Congenital lobar emphysema: problems in diagnosis and management. Archives of disease in childhood 1983;58(9):709-12.

[5] McBride JT, Wohl ME, Strieder DJ, Jackson AC, Morton JR, Zwerdling RG, et al. Lung growth and airway function after lobectomy in infancy for congenital lobar emphysema. J Clin Invest 1980;66(5):962-70.

[6] Hislop A, Reid L. New pathological findings in emphysema of childhood. 1. Polyalveolar lobe with emphysema. Thorax 1970;25(6):682-90.

[7] Tapper D, Schuster S, McBride J, Eraklis A, Wohl ME, Williams A, et al. Polyalveolar lobe: anatomic and physiologic parameters and their relationship to congenital lobar emphysema. J Pediatr Surg 1980;15(6):931-7.

[8] Langston C. New concepts in the pathology of congenital lung malformations. Semin Pediatr Surg 2003;12(1):17-37.

[9] Kunisaki SM, Fauza DO, Nemes LP, Barnewolt CE, Estroff JA, Kozakewich HP, et al. Bronchial atresia: the hidden pathology within a spectrum of prenatally diagnosed lung masses. J Pediatr Surg 2006;41(1):61-5; discussion -5.

[10] Ehrenberg-Buchner S, Stapf AM, Berman DR, Drongowski RA, Mychaliska GB, Treadwell MC, et al. Fetal lung lesions: can we start to breathe easier? Am J Obstet Gynecol 2013;208(2):151 e1-7.

[11] Rothenberg SS. First decade's experience with thoracoscopic lobectomy in infants and children. J Pediatr Surg 2008;43(1):40-4; discussion 5.

[12] Kunisaki SM, Powelson IA, Haydar B, Bowshier BC, Jarboe MD, Mychaliska GB, et al. Thoracoscopic vs open lobectomy in infants and young children with congenital lung malformations. Journal of the American College of Surgeons 2014;218(2):261-70.

[13] Jaffe A, Chitty LS. Congenital cystic adenomatoid malformations may not require surgical intervention. Arch Dis Child Fetal Neonatal Ed 2006;91(6):F464.

[14] Ng C, Stanwell J, Burge DM, Stanton MP. Conservative management of antenatally diagnosed cystic lung malformations. Archives of disease in childhood 2014;99(5):432-7.

[15] Criss CN, Musili N, Matusko N, Baker S, Geiger JD, Kunisaki SM. Asymptomatic congenital lung malformations: Is nonoperative management a viable alternative? J Pediatr Surg 2018;53(6):1092-7.

[16] Baird R, Puligandla PS, Laberge JM. Congenital lung malformations: informing best practice. Semin Pediatr Surg 2014;23(5):270-7.

[17] Karnak I, Senocak ME, Ciftci AO, Buyukpamukcu N. Congenital lobar emphysema: diagnostic and therapeutic considerations. J Pediatr Surg 1999;34(9):1347-51.

[18] Thakral CL, Maji DC, Sajwani MJ. Congenital lobar emphysema: experience with 21 cases. Pediatric surgery international 2001;17(2-3):88-91.

[19] Choudhry M, Drake D. Antenatally diagnosed lung malformations: a plea for long-term outcome studies. Pediatric surgery international 2015;31(5):439-44.

[20] Hirschl RB, Minneci P, Gadepalli S, Saito JM, Askegard-Giesmann J, Deans K, et al. Development of a multi-institutional clinical research consortium for pediatric surgery. J Pediatr Surg 2017;52(7):1084-8.

[21] Nelson RL. Congenital cystic disease of the lung. J Pediatr 1932;1(2):233-8.

[22] Robertson R, James ES. Congenital lobar emphysema. Pediatrics 1951;8(6):794-804.

[23] Warner JO, Rubin S, Heard BE. Congenital lobar emphysema: a case with bronchial atresia and abnormal bronchial cartilages. Br J Dis Chest 1982;76(2):177-84.

[24] Haller JA, Jr., Golladay ES, Pickard LR, Tepas JJ, 3rd, Shorter NA, Shermeta DW. Surgical management of lung bud anomalies: lobar emphysema, bronchogenic cyst, cystic adenomatoid malformation, and intralobar pulmonary sequestration. The Annals of thoracic surgery 1979;28(1):33-43.

[25] Glenski JA, Thibeault DW, Hall FK, Hall RT, Germann DR. Selective bronchial intubation in infants with lobar emphysema: indications, complications, and long-term outcome. American journal of perinatology 1986;3(3):199-204.

[26] Mei-Zahav M, Konen O, Manson D, Langer JC. Is congenital lobar emphysema a surgical disease? J Pediatr Surg 2006;41(6):1058-61.

[27] Paramalingam S, Parkinson E, Sellars M, Diaz-Cano S, Nicolaides KH, Davenport M. Congenital segmental emphysema: an evolving lesion. European journal of pediatric surgery : official journal of Austrian Association of Pediatric Surgery [et al] = Zeitschrift fur Kinderchirurgie 2010;20(2):78-81.

[28] Burge D, Wheeler R. Increasing incidence of detection of congenital lung lesions. Pediatric pulmonology 2010;45(1):103; author reply 4.

[29] Lima JS, Camargos PA, Aguiar RA, Campos AS, Aguiar MJ. Pre and perinatal aspects of congenital cystic adenomatoid malformation of the lung. The journal of maternal-fetal & neonatal medicine : the official journal of the European Association of Perinatal Medicine, the Federation of Asia and Oceania Perinatal Societies, the International Society of Perinatal Obstet 2014;27(3):228-32.

[30] Lau CT, Kan A, Shek N, Tam P, Wong KK. Is congenital pulmonary airway malformation really a rare disease? Result of a prospective registry with universal antenatal screening program. Pediatric surgery international 2017;33(1):105-8.

[31] Babu R, Kyle P, Spicer RD. Prenatal sonographic features of congenital lobar emphysema. Fetal Diagn Ther 2001;16(4):200-2.

[32] Rahman N, Lakhoo K. Comparison between open and thoracoscopic resection of congenital lung lesions. J Pediatr Surg 2009;44(2):333-6.

[33] Pariente G, Aviram M, Landau D, Hershkovitz R. Prenatal diagnosis of congenital lobar emphysema: case report and review of the literature. J Ultrasound Med 2009;28(8):1081-4.

[34] Quinton AE, Smoleniec JS. Congenital lobar emphysema--the disappearing chest mass: antenatal ultrasound appearance. Ultrasound in obstetrics & gynecology : the official journal of the International Society of Ultrasound in Obstetrics and Gynecology 2001;17(2):169-71.

[35] Kunisaki SM, Ehrenberg-Buchner S, Dillman JR, Smith EA, Mychaliska GB, Treadwell MC. Vanishing fetal lung malformations: Prenatal sonographic characteristics and postnatal outcomes. J Pediatr Surg 2015;50(6):978-82.

[36] Winters WD, Effmann EL. Congenital masses of the lung: prenatal and postnatal imaging evaluation. Journal of thoracic imaging 2001;16(4):196-206.

[37] Olutoye OO, Coleman BG, Hubbard AM, Adzick NS. Prenatal diagnosis and management of congenital lobar emphysema. J Pediatr Surg 2000;35(5):792-5.

[38] Mon RA, Johnson KN, Ladino-Torres M, Heider A, Mychaliska GB, Treadwell MC, et al. Diagnostic accuracy of imaging studies in congenital lung malformations. Arch Dis Child Fetal Neonatal Ed 2018.

[39] Dogan R, Dogan OF, Yilmaz M, Demircin M, Pasaoglu I, Kiper N, et al. Surgical management of infants with congenital lobar emphysema and concomitant congenital heart disease. Heart Surg Forum 2004;7(6):E644-9.

[40] Moideen I, Nair SG, Cherian A, Rao SG. Congenital lobar emphysema associated with congenital heart disease. J Cardiothorac Vasc Anesth 2006;20(2):239-41.

[41] Kulaylat AN, Engbrecht BW, Hollenbeak CS, Safford SD, Cilley RE, Dillon PW. Comparing 30-day outcomes between thoracoscopic and open approaches for

resection of pediatric congenital lung malformations: Evidence from NSQIP. J Pediatr Surg 2015;50(10):1716-21.

[42] Krivchenya DU, Rudenko EO, Dubrovin AG. Congenital emphysema in children: segmental lung resection as an alternative to lobectomy. J Pediatr Surg 2013;48(2):309-14.

[43] Sulkowski JP, Deans KJ, Asti L, Mattei P, Minneci PC. Using the Pediatric Health Information System to study rare congenital pediatric surgical diseases: development of a cohort of esophageal atresia patients. J Pediatr Surg 2013;48(9):1850-5.

FIGURE LEGENDS

Figure 1. Retrospective cohort study of operative congenital lobar emphysema (CLE) from eleven hospitals associated with the Midwest Pediatric Surgical Consortium. **(A)** Pie graph showing the number and percent of CLE resections relative to that of other lung malformations (BPS, bronchopulmonary sequestration; BA, bronchial atresia; BC, bronchogenic cyst; CPAM, congenital pulmonary airway malformation). **(B)** Bar graph demonstrating marked variability in CLE cases contributed by institution.

Figure 2. Clinical characteristics of operative congenital lobar emphysema (CLE). **(A)** Bar graph of lobar distribution of CLE lesions, by percent. **(B)** Bar graph of timing of operative resection (in months) among CLEs.

TABLES

Table 1: Postoperative complications following CLE resection (n=53)

Complication	n (%)
Any	14 (26.4)
Intubation>48 hours	7 (13.2)
Pneumothorax requiring chest tube	4 (7.5)
Airleak>48 days	2 (3.8)
Bronchial plug requiring bronchoscopy	1 (1.9)
Re-operation	1 (1.9)
Any post-operative blood transfusion	2 (3.8)
Pneumonia	2 (3.8)
Hospital readmission	3 (5.7)
Other	1 (1.9)
Death	1 (1.9)

Abbreviations: CLE, congenital lobar emphysema

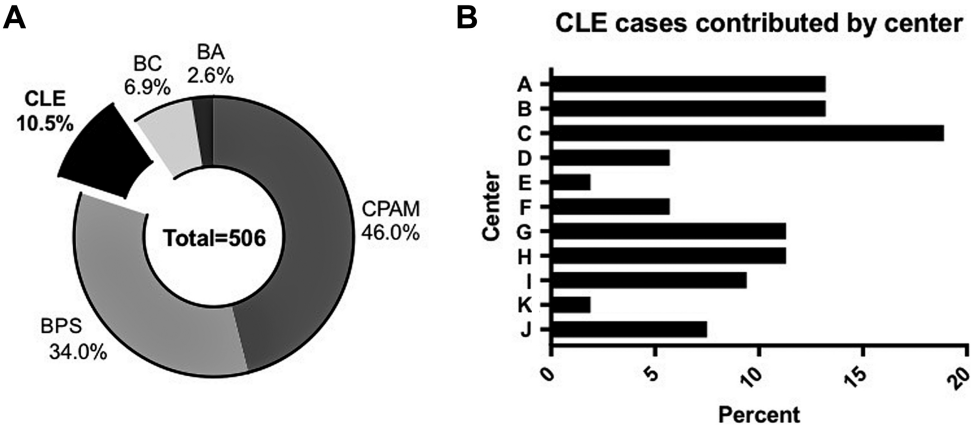
A
BC
6.9%
BA
2.6%
CLE
10.5%
CPAM
46.0%
Total=506
BPS
34.0%
B
CLE cases contributed by center
Center
A
B
C
D
E
F
G
H
I
K
J
0
5
10
15
20
Percent

Figure 1

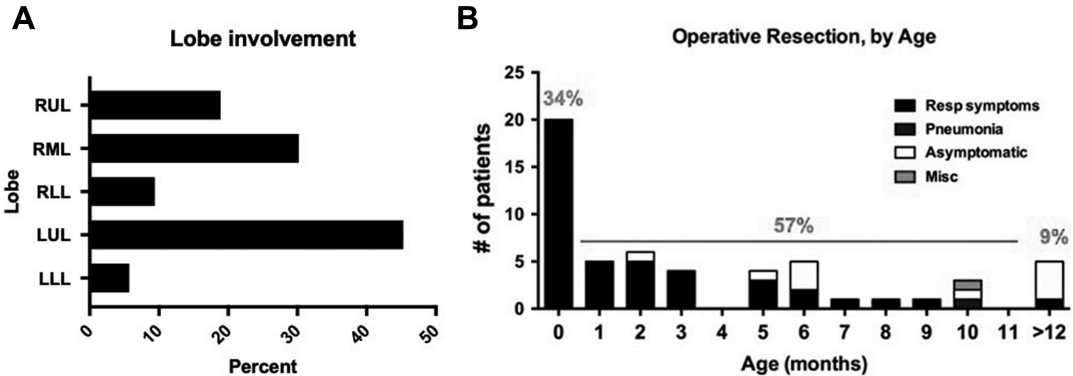
A
Lobe involvement
RUL
RML
RLL
LUL
LLL
Lobe
0
10
20
30
40
50
Percent
B
Operative Resection, by Age
34%
57%
9%
Resp symptoms
Pneumonia
Asymptomatic
Misc
# of patients
25
20
15
10
5
0
0 1 2 3 4 5 6 7 8 9 10 11 >12
Age (months)

Figure 2