Development of a Multi-Institutional Registry for Children with Operative Congenital Lung Malformations

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Abstract

Introduction: The purpose of this study was to develop a multi-institutional registry to characterize the demographics, management, and outcomes of a contemporary cohort of children undergoing congenital lung malformation (CLM) resection.

Methods: After central reliance IRB approval, a web-based, secure database was created to capture retrospective cohort data on pathologically-confirmed CLMs performed between 2009-2015 within a multi-institutional research collaborative.

Results: Eleven children's hospitals contributed 506 patients. Among 344 prenatally diagnosed lesions, the congenital pulmonary airway malformation volume ratio was measured in 49.1%, and fetal MRI was performed in 34.3%. One hundred thirty-four (26.7%) children had respiratory symptoms at birth. Fifty-eight (11.6%) underwent neonatal resection, 322 (64.1%) had surgery at 1-12 months, and 122 (24.3%) had operations after 12 mos. The median age at resection was 6.7 months (interquartile range, 3.6-11.4). Among 230 elective lobectomies performed in asymptomatic patients, thoracoscopy was successfully utilized in 102 (44.3%), but there was substantial variation across centers. The most common lesions were congenital pulmonary airway malformation (n=234, 47.3%) and intralobar bronchopulmonary sequestration (n=106, 21.4%).

Conclusion: This multicenter cohort study on operative CLMs highlights marked disease heterogeneity and substantial practice variation in preoperative evaluation and operative management. Future registry studies are planned to help establish evidencebased guidelines to optimize the care of these patients.

Abbreviations: CPAM, congenital pulmonary airway malformation; BPS, bronchopulmonary sequestration; congenital lung malformation, CLM; congenital pulmonary airway malformation volume ratio, CVR

Keywords: congenital pulmonary airway malformation, congenital cystic adenomatoid malformation, bronchopulmonary sequestration, congenital lung malformations, registry

Level of Evidence: Level II

Introduction

Congenital lung malformations (CLM) consist of a broad range of lung anomalies, including congenital pulmonary airway malformations (CPAM), bronchogenic cysts, bronchial atresia, bronchopulmonary sequestrations (BPS), and congenital lobar emphysema (CLE) [1-3]. The incidence of CLMs has increased over time, with more recent studies suggesting an incidence of approximately 1 in 2500 births, likely secondary to widespread prenatal screening as well as improvements in ultrasound image quality [4-6]. From 1997 to 2009, the number of operative CLM cases increased almost three-fold [7]. Although the etiology of CLMs is unclear, the inciting event may be related to transient bronchial obstruction during the canalicular phase of lung morphogenesis [1, 8]. More recently, dysregulated signaling in the Ras and PI3K-AKT-mTOR pathways has been implicated [9].

In contrast to other rare anomalies managed by pediatric surgeons, CLMs have several features, including highly variable symptomatology, unclear natural history, and diverse histopathology, which make clinical outcomes research uniquely challenging [10, 11]. Furthermore, most of these children are now diagnosed prenatally and therefore have clinically relevant data that must be extracted from the maternal medical record prior to analysis [12]. Delivering evidence-based, quality care with complication rates that are as low as possible is especially critical for asymptomatic CLMs given that we, as pediatric surgeons, are asking families to consent to a major operation performed relatively infrequently for an incidentally identified lesion that may or may not cause any problems later in life [13, 14].

In this study, we sought to develop a multi-institutional registry to better characterize the demographics, management approaches, and outcomes in a contemporary cohort of children undergoing CLM resection. The rationale for creating a registry was to assist in establishing evidence-based benchmarks by which individual institutions can measure their outcomes in a more directed fashion. Our group hypothesized that there is substantial practice variation in the preoperative evaluation and operative management of these lesions.

1. Methods

Following approval through a central reliance institutional review board process (#96707), 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. The eleven hospitals associated with the Midwest Pediatric Surgery Consortium (wwww.mwpsc.org) were invited to participate [15]. Patients were identified for study inclusion based on International Classification of Disease, 9th Edition, Clinical Modification (ICD-9-CM) diagnosis codes for a CPAM (748.4), BPS (748.5), CLE (770.2), and other congenital lung lesions (748.6). Current Procedural Terminology (CPT) codes corresponding to removal of lung tissue (32140, 32440, 32480-4, 32505, 32662-71) were confirmed in each patient. Pathology reports were reviewed to verify the presence of a CLM. Since specific CLM pathologic terminology may not be standardized across institutions, pathologic diagnosis categories were specifically defined as detailed elsewhere [16]. All lesions with an associated systemic feeding artery were classified as either BPS or a

hybrid CPAM, depending on appearance of the affected lung parenchyma.

Collaborative consensus was obtained for identifying and defining relevant data elements for collection. These included demographics, maternal records, other congenital anomalies, birth history, prenatal and postnatal diagnostic imaging, operative approach and findings, and postoperative morbidity and mortality. A multi-institutional CLM database containing 103 different data fields organized into seven modules (demographics, fetal diagnosis, fetal treatment, birth history, preoperative, intraoperative, and postoperative) was created within REDCap (Research Electronic Data Capture, 8.1.20, Vanderbilt University, Nashville, TN) software hosted by the Michigan Institute for Clinical & Health Research at the University of Michigan (Table 1). All study data were validated by trained reviewers at both the central and institutional levels for completeness and accuracy. Outliers were reconfirmed to be appropriate, and no patients were excluded on the basis of missing patient information. Frequencies and percentages were used to describe categorical variables. Median and interquartile ranges were generally used to describe continuous variables. Statistical analysis was performed as appropriate using STATA (StataCorp, College Station, TX).

Table 1

2. Results

2.1. Demographics

All eleven children's hospitals within the consortium participated and contributed a total of 506 cases. The number of children reported from each institution ranged from 9 to 74 patients, yielding a mean of 46.0±23.1 cases per member hospital (Figure 1). Two hundred ninety-eight (59.0%) were male. Race/ethnicity data showed 314 (62.2%)

Fig 1

white/Caucasian, 127 (25.1%) non-white, and 64 (12.7%) other or unknown. Two hundred sixty-four (53.7%) had private insurance, and 152 (30.9%) had public insurance such as Medicaid or were self-pay.

2.2. Fetal diagnosis and treatment

Three hundred forty-four cases (68.0%) were diagnosed in utero. The most common ultrasound diagnosis was CPAM without a systemic feeding vessel (n=234, 70.9%). Calculation of an initial CPAM volume ratio (CVR) was reported in 169 (49.1%) cases but was variable by institution. Five institutions reported initial CVR data at referral in more than 60% of cases, whereas two institutions did not record CVR measurements for any patients in the database. Fetal magnetic resonance imaging (MRI) was performed in 118 (34.3%) cases. Thirty-four (10.4%) fetuses were diagnosed with additional congenital anomalies (Table 2). Fifteen (4.4%) fetuses had hydrops, which was defined as the presence of fluid in two or more spaces, including ascites, pleural effusion, pericardial effusion, or skin edema, at some point in gestation. The most common finding consistent with hydrops was ascites (n=13, 86.7%) followed by skin edema (n=8, 53.3%) and pleural effusion (n=4, 26.7%).

Table 2

Maternal steroids were administered to 41 (11.9%) fetuses at eight (72.7%) different institutions. Invasive prenatal procedures were uncommon (n=6, 1.7%), ranging from thoracentesis (n=2), thoracoamniotic shunt placement (n=1), and ex utero intrapartum treatment (EXIT, n=3). Among the 15 fetuses with hydrops, only three (20%) received an invasive therapeutic intervention prior to delivery.

2.3. Birth history

The median gestational age at birth was 39.0 weeks [interquartile range (IQR), 37.0-39.2]. The median birth weight was 3.2 kg (IQR, 2.8-3.6). Excluding those who underwent the EXIT procedure, 131 (26.3%) patients had respiratory symptoms at birth. One hundred eighteen newborns (23.0%) received supplemental oxygen, and 49 (9.8%) were intubated prior to resection. Plain film chest radiograph reports were available in 317 (64.6%) newborns. However, among inborn neonates, 165 (88.2%) had a chest radiograph shortly in the newborn period. Two hundred forty-four (77.7%) studies suggested an opacity or lucency consistent with a pulmonary parenchymal lesion. A pneumothorax was seen in 13 (4.1%) newborns.

2.4. Preoperative and intraoperative data

At least one chest CT was performed preoperatively in 462 (91.7%) children at a median age of 3.2 months (IQR, 1.2-6.8). Eighty-seven (18.9%) children had more than one preoperative CT scan (range, 2-5). The most common CT diagnoses were CPAM without a systemic feeding vessel (n=320, 49.9%) and BPS (n=104, 22.6%). There were 236 (47.5%) right-sided and 259 (52.1%) left-sided lesions. Thirty-two (6.4%) patients had unilateral lesions involving more than one lobe, and two (0.4%) additional children had bilateral disease. The most common anatomic locations were left lower lobe (n=147, 29.4%) and right lower lobe (n=145, 29.0%). Seventy-five (14.5%) children had co-morbidities, the most common of which were pulmonary (n=26, 5.2%), cardiac (n=24, 4.8%), and neurologic (n=12, 2.4%) in nature.

The median age at resection was 6.7 months (IQR, 3.6-11.4). Fifty-eight (11.6%) underwent neonatal resection, 322 (64.1%) had surgery at 1-12 months, and 122 (24.3%) had operations after 12 months. A bar graph depicting the age distribution at operative resection is shown in Figure 2. The median patient weight at surgical resection was 7.5 kg (IQR, 5.9-9.8). The indications for operative management, based on final pathologic diagnosis, can be found in Table 3. The most common overall reason for surgery was to prevent future complications associated with a CLM in an otherwise asymptomatic child (n=309, 61.3%). One hundred twenty-two (24.2%) resections were performed because of ongoing respiratory symptoms. Sixty-one (12.1%) patients had an operation because of a prior episode of pneumonia [median age, 35.8 months (IQR, 9.6-82.1)]. Pneumothorax, failure to thrive, elevated concern for malignancy (e.g., pleuropulmonary blastoma), and heart failure were uncommon indications for resection. The operative report identified 96 different primary surgeons. The mean operative time was 177.9±101.2 minutes. The vast majority of procedures were lobectomies (n=374, 74.8%). Segmentectomies (n=21, 4.2%) and non-anatomic resections (n=33, 6.6%) were rarely performed.

Fig 2

Table 3

There were 257 (51.4%) open and 203 (40.6%) thoracoscopic resections. An additional 37 cases were thoracoscopic converted to open resections, yielding an overall conversion rate of 15.4%. Among 230 elective lobectomies performed in asymptomatic patients [median age, 6.4 months (IQR, 4.5-9.4)], thoracoscopy resection was accomplished in 102 (44.3%). The median age at elective thoracoscopic lobectomy was 6.2 months (IQR, 5.3-8.7). However, there was substantial variation in percentage

of elective thoracoscopic resections (range, 0-100%) across consortium member hospitals.

2.5. Postoperative data

One hundred seventy-eight (36.6%) were initially managed in the intensive care unit post-operatively. An epidural catheter was used for postoperative pain control in 128 (26.0%) patients, most (96.1%) of whom had a thoracotomy incision. Among symptomatic neonates, the median total duration of perioperative mechanical ventilator support was 6.0 days (IQR, 2.5-19.5).

The median chest tube duration was 3.0 days (IQR, 2.0-4.0), and the median postoperative hospital length of stay was 3.0 days (IQR, 2.0-6.0). Age at resection was significantly correlated with hospital length of stay ($p<0.0001$). Five (1.0%) neonates were managed with ECMO postoperatively. The median length of stay among those on ECMO support was 35.0 days (range, 30.0-159.0).

Five hundred four (99.6%) patients survived to hospital discharge. Two children died secondary to respiratory failure. Post-operative complications occurred in 91 (18.4%) patients (Table 4). The most common complications were pneumothorax requiring insertion of a chest tube (n=40, 8.1%) and intubation greater than 48 hours (n=20, 4.0%). However, further query of the latter event revealed that 12 (60%) of these children were already intubated pre-operatively, and 17 (85%) had respiratory symptoms prior to resection. Rare but notable post-operative complications in the entire cohort included hospital re-admission (n=9, 1.8%), superficial skin infections (n=5,

Table 4

1.0%), postoperative blood transfusion (n=7, 1.4%), pneumonia (n=8, 1.6%), and unplanned re-operation (n=6, 1.2%).

The distribution of pathologically confirmed CLMs, stratified by age at resection, is shown in Table 5. The most common type among all age groups was CPAM without a feeding vessel, formerly referred to as congenital cystic adenomatoid malformation (CCAM), in 234 (47.3%) followed by BPS in 172 (34.7%). One hundred six (61.6%) of the BPS lesions were intralobar. When pathologic diagnosis was compared to ICD code for each patient, there was a 25.0% discordance rate. The most common error in ICD coding was the failure to properly document the presence of a systemic feeding vessel in association with a CPAM (i.e., hybrid lesion) in 77.8% of discordant cases.

Table 5

3. Discussion

Disease-specific, multicenter cohort registries are becoming increasingly popular in pediatric surgery [17-19]. Interest in these studies has largely been driven by the desire for more granular clinical data, particularly for rare disorders, than what is currently provided by administrative and clinical databases [20, 21]. Contemporary examples of registries that have impacted clinical care in pediatric surgical patients include the Congenital Diaphragmatic Hernia Study Group, Extracorporeal Life Support Organization Registry, the International Serial Transverse Enteroplasty Data Registry, and the Japanese Biliary Atresia Registry [22-25].

In this study, we established a large, comprehensive, multi-institutional database to help study the contemporary surgical management of CLMs. Based on over 500 operative lesions collected retrospectively at eleven major children's hospitals, there

were several interesting findings. First, despite having several centers within the consortium with established capabilities to perform the full spectrum of in utero invasive procedures [26, 27], fetal interventions, including shunts and EXIT procedures, were strikingly uncommon, accounting for <2% of all prenatally diagnosed lesions. Given the historically strong interest in fetal surgical intervention for CLMs espoused by some quaternary care prenatal referral centers [2, 28-30], these data are enlightening in this current era where maternal steroids have been adopted nationally as first line therapy for large fetal CLMs. Second, our registry demonstrated a marked practice variation regarding the diagnostic evaluation of fetal CLMs. For example, only one-third of fetuses had an MRI as part of their prenatal diagnostic evaluation. The role of MRI in this setting remains controversial [31, 32]. Less than half of prenatally diagnosed patients were followed by a calculated CVR despite multiple studies now demonstrating its prognostic value, both in terms of predicting hydrops risk as well as determining likelihood of respiratory distress at birth, among other outcome measures [12, 33-36].

Postnatal surgical management among asymptomatic CLMs was also variable among consortium centers as revealed by notable differences in surgical approach at resection. Excluding cases performed during the neonatal period, approximately half of all resections were performed by thoracotomy. Although the thoracoscopic approach has now been described for over two decades and is likely to be associated with similar perioperative outcomes, less postoperative pain, and improved cosmesis compared to traditional thoracotomy [37, 38], open surgery was still exclusively performed at a number of centers during the entire study period. Data suggesting this restrained enthusiasm for thoracoscopy for elective resections has also been demonstrated in both

large single-center reports [39] as well as large administrative and clinical database studies [40, 41].

Our study also highlights the incredible diversity of CLMs with respect to disease presentation and pathology. First, slightly less than half of the operative CLMs in our registry were isolated CPAMs based on a review of the pathology reports. Second, while the majority were prenatally diagnosed by ultrasound and were asymptomatic at birth, one-third were still initially diagnosed in the postnatal period in spite of the widespread availability and enhanced resolution of ultrasound technologies. This prenatal diagnosis rate is comparable to that recently reported for congenital diaphragmatic hernia [42]. Nevertheless, the true CLM prenatal diagnosis rate may be slightly higher given that some member institutions without well-established fetal care programs may have had difficulty accessing outside hospital maternal records. Finally, we showed that approximately 90% of resections now occur outside of the neonatal period and are largely elective in nature, and this has been corroborated by other recent studies [12, 43]. These findings suggest a possible referral bias skewed towards neonates with symptomatic CLMs in many single center studies, particularly in those managed at highly developed fetal care centers [28, 44]. On the other end of the spectrum, we identified 61 cases (12.1%) in predominantly older children who had resections performed in the setting of a prior pneumonia or infection. These data are congruent with work by others, who have also shown that infection is a bona fide complication in some CLMs initially managed nonoperatively [45-47].

Our CLM registry offers a number of distinct advantages compared to recently published multicenter CLM data based on the American College of Surgeons National

Surgical Quality Improvement Program- Pediatric (NSQIP-P), Healthcare Cost and Utilization Project Kid's Inpatient Database, and Nationwide Inpatient Sample, among others [7, 40, 48]. For example, because the NSQIP-P has yet to contain a lung surgery-specific module, the majority of complications recorded after elective CLM surgery occur infrequently [43, 49]. Our database revealed that air leak and pneumothorax, both major predictors for chest tube removal and hospital length of stay, were the most common complications following elective resection. It is also difficult in the NSQIP-P and other databases to elucidate which patients underwent a planned thoracoscopic lobectomy that was converted intraoperatively to an open procedure [40, 49]. The use of other hospital databases may be problematic for evaluating CLM outcomes because cases cannot be adequately stratified based on rationale for the procedure [49, 50]. In an attempt to address this important confounder, some investigators have used urgent/emergent and elective status, age at operation, or other surrogates for operative indication [41, 48]. Conversely, data from our CLM registry would allow us to better understand the rationale for operative resection (e.g., tachypnea, prior pneumonia), factors that are not easily captured otherwise. Finally, clinical databases, such as the NSQIP-P, do not contain hospital or surgeon identifiers, which would be critical for evaluating volume-outcome relationships that may exist in pediatric thoracic surgery [51].

Another noteworthy result from this study was the relative inaccuracy of ICD coding as the sole basis for capturing and evaluating CLM health services outcomes. Based on pathology reports, we identified the use of an incorrect ICD code in 25% of cases, a finding that may not be surprising given the overlapping and complicated

terminology often used to describe different CLMs as well as the potential for hybrid lesions with more than one pathologic diagnosis [8, 32, 52]. Moreover, ICD codes for these lesions can sometimes be non-specific, such as in the case of bronchogenic cysts and CPAMs, which share the exact same ICD code despite the fact that they are pathologically distinct, have unique disease presentations, and require different operative procedures. Based on our registry data, 13% with the ICD-9-CM code, 748.4, were indeed bronchogenic cysts and not CPAMs. Adding to further confusion is the fact that some surgical investigators have considered BPS or other malformations to be under the umbrella of CPAM [41, 50] whereas our group, among others, maintain a more rigid definition of CPAMs, formerly referred to as CCAMs, as articulated by Stocker [3, 10].

Several caveats regarding this study should be acknowledged. First, this is a retrospective registry that is limited by the quality of the data present in the electronic medical record at each institution [53]. Although the vast majority of data points can be extracted in accurate fashion [54], there were missing data in some fields, particularly if they required an extensive query of the maternal or birth record (e.g., ultrasound mass characteristics, Apgar scores, neonatal chest radiographs) from an outside hospital. Second, despite a relatively large number of CLM patients, our ability to perform subgroup analyses in subsequent studies may still be limited. Third, because this is an operative cohort registry, some CLM patients were obviously excluded, such as those who never had surgery due to non-operative management or those who may have died in utero or in the early postnatal period prior to resection. Finally, although less susceptible to referral bias than large, single institution studies [55, 56], our results may

still not be generalizable to other pediatric centers, such as highly specialized, quaternary fetal care units as well as institutions where lung resections are performed on a very infrequent basis.

Despite these aforementioned limitations, we believe that our registry data will be useful in helping surgeons and other health care providers to better counsel families and manage these patients. Future studies are ongoing looking in more detail at radiology-pathology correlation, postnatally diagnosed CLMs and malignancy risk, thoracoscopic resection, and volume-outcome relationships, among other topics. Because the CLM registry was intentionally designed to optimize participation and compliance, it can also be easily modified to capture long-term outcomes data or embraced by other institutions to create a larger national or international consortium. Finally, we are exploring prospective evaluation using this registry concept, as has already done elsewhere [34, 57], to help inform best practice in the management of CLMs.

CONFLICT OF INTEREST

The authors indicate no potential conflicts of interest.

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FIGURE LEGENDS

Figure 1. Bar graph of number of cases contributed by member hospitals within the Midwest Pediatric Surgery Consortium.

Figure 2. Timing of resection (in months) among operative congenital lung malformations.

TABLES

Table 1. Selected data fields (45 of 103) captured in the Midwest Pediatric Surgery Consortium congenital lung malformation registry

Module 1: Demographics		
hospital name	date of birth	gender
prenatal diagnosis	race	Insurance
Module 2: Fetal diagnosis		
gestational age	ultrasound diagnosis	CPAM volume ratio
cyst description	mediastinal shift	hydrops
MRI	MRI date	
Module 3: Fetal treatment		
maternal steroids	fetal resection	EXIT
Module 4: Birth history		
gestational age	Apgar scores	respiratory symptoms
birthweight	inborn	ventilator days
radiographs		
Module 5: Preoperative		
diagnosis age	CT date	CT diagnosis
co-morbidities		
Module 6: Intraoperative		
date of resection	operative approach	chest tube
weight at operation	lobe involved	blood transfusions
operative indication	extent of resection	surgeon (de-identified)
operative duration		
Module 7: Postoperative		
epidural use	survival	date of discharge
chest tube duration	complications	date of last follow up
pathology diagnosis		

CPAM, congenital pulmonary airway malformation; MRI, magnetic resonance imaging; ex utero intrapartum treatment, EXIT; CT, computed tomography

TABLES

Table 2. Selected concomitant anomalies in 344 fetal lung malformation patients, by system

Neurologic (2.3%)	
myelomeningocele	Dandy-Walker syndrome
agenesis of corpus callosum	
Cardiac (4.9%)	
coarctation of the aorta	Tetralogy of Fallot
ventricular septal defect	pulmonary valve atresia
atria septal defect	
Renal (0.6%)	
hydronephrosis	duplex collecting system
Gastrointestinal (0.9%)	
cystic abdominal mass	anorectal malformation (dilated rectum)
Other thoracic (0.6%)	
congenital diaphragmatic hernia	eventration of the diaphragm

Table 3. Indications for postnatal operative resection, by final pathologic diagnosis.

Pathology	*Respiratory symptoms*	*Asymptomatic, Prevent future complications*	*Previous pneumonia or infection*	*PTX*	*Failure to thrive*	*Cancer concern*	*Heart failure*
CPAM, n=233	18.5%	67.4%	10.3%	2.6%	0.9%	1.7%	0.0%
CPAM w/ feeder n=46	2.2%	87.0%	4.3%	2.2%	0.0%	2.2%	0.0%
Intralobar BPS, n=106	10.4%	67.9%	18.9%	0.0%	1.9%	0.0%	0.0%
Extralobar BPS, n=66	16.7%	65.2%	6.1%	1.5%	1.5%	4.5%	1.5%
CLE, n=53	77.4%	18.9%	11.3%	1.9%	11.3%	0.0%	0.0%
BC, n=35	48.6%	37.1%	14.3%	2.9%	2.9%	5.7%	0.0%
Bronchial atresia, n=13	23.1%	69.2%	15.4%	0.0%	0.0%	0.0%	0.0%

PTX, pneumothorax; CPAM, congenital pulmonary airway malformation; BPS, bronchopulmonary sequestration; CLE, congenital lobar emphysema; BC, bronchogenic cyst

Table 4. Post-operative complication rates after lung malformation resection, overall and by age group

Complication	*All ages n=91*	*Neonatal n=22*	*Age 1-12 mo n=49*	*Age>12 mo n=20*
Intubation>48 hrs	20 (4.0%)	9 (15.0%)	9 (2.8%)	2 (1.7%)
Re-intubation	10 (2.0%)	3 (5.0%)	7 (2.2%)	0 (0.0%)
Pneumothorax	40 (8.1%)	9 (15.0%)	21 (6.6%)	10 (8.5%)
Air leak>48 hrs	7 (1.4%)	1 (1.7%)	3 (0.9%)	3 (2.5%)
Bronchial plug	4 (0.8%)	0 (0.0%)	3 (0.9%)	1 (0.8%)
Chylothorax	4 (0.8%)	1 (1.7%)	2 (0.6%)	1 (0.8%)
Phrenic or recurrent laryngeal nerve injury	1 (0.2%)	0 (0.0%)	1 (0.3%)	0 (0.0%)
Re-do thoracotomy/thoracoscopy	6 (1.2%)	0 (0.0%)	2 (0.6%)	4 (3.4%)
Postop blood transfusion	7 (1.4%)	2 (3.3%)	5 (1.6%)	0 (0.0%)
Superficial skin infection	5 (1.0%)	2 (3.3%)	3 (0.9%)	0 (0.0%)
Pneumonia	8 (1.6%)	0 (0.0%)	6 (1.9%)	2 (1.7%)
Urinary tract infection	2 (0.4%)	1 (1.7%)	0 (0.0%)	1 (0.8%)
Hospital re-admission	9 (1.8%)	1 (1.7%)	6 (1.9%)	2 (1.7%)

Table 5. Pathologic diagnosis, overall and by age group.

Group	*CPAM*	*CPAM w/ feeder*	*Intralobar BPS*	*Extralobar BPS*	*CLE*	*BC*	*Bronchial atresia*
All ages, n=495	234 (47%)	46 (9%)	106 (21%)	66 (13%)	53 (11%)	35 (7%)	13 (3%)
Neonatal, n=58	27 (48%)	1 (2%)	6 (11%)	5 (9%)	18 (32%)	2 (4%)	1 (2%)
Age 1-12 mo, n=318	150 (48%)	38 (12%)	71 (23%)	48 (15%)	30 (10%)	13 (4%)	11 (4%)
Age>12 mo, n=119	55 (46%)	7 (6%)	28 (24%)	11 (9%)	5 (4%)	20 (17%)	1 (1%)

CPAM, congenital pulmonary airway malformation; BPS, bronchopulmonary sequestration; CLE, congenital lobar emphysema; BC, bronchogenic cyst

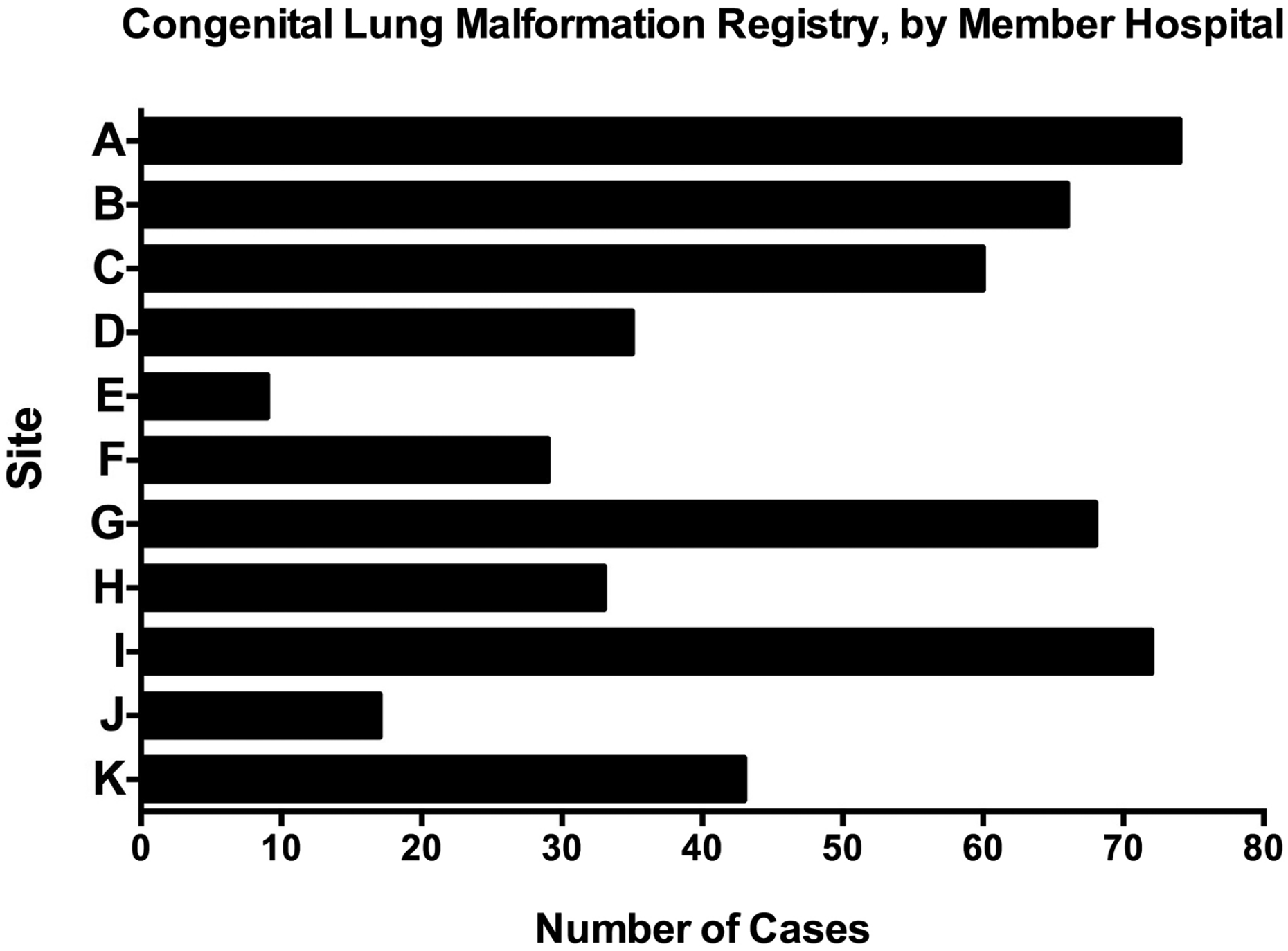
Congenital Lung Malformation Registry, by Member Hospital
Site
A
B
C
D
E
F
G
H
I
J
K
0
10
20
30
40
50
60
70
80
Number of Cases

Figure 1


Operative Resection, by Age
# of patients
70
60
50
40
30
20
10
0
Fetal EXIT
0
2
4
6
8
10
12
14
16
18
20
22
24
Age (months)

Figure 2