Journal of Cardiothoracic Surgery

RESEARCH Open Access

Clinical outcomes after surgical resection in asymptomatic and symptomatic children with congenital lung malformations

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Abstract

Purpose Our study aims to evaluate the outcomes of children with congenital lung malformation (CLM) who have undergone surgical resection.

Methods A retrospective analysis was conducted among children under 18 who were diagnosed with CLM and underwent surgery at Srinagarind Hospital, Khon Kaen University between January 2007 and December 2023. We collected data on surgical outcomes, including operative time, postoperative complications, and mortality rate.

Results During our study period, a total of 38 children with CLM were undergone surgery. The median time for diagnosis was 9 months (IQR 1–33 months). Congenital pulmonary airway malformation was the most common diagnosis, affecting 26 children (68.4%). Of these, 25 children were operated on when they presented with symptoms, while 13 children were operated on even though they were asymptomatic. The median age at surgery was 12 months (IQR 3–32 months) for symptomatic children and 6 months (IQR 3–12 months) for asymptomatic children ($P=0.201$). After the surgery, symptomatic children had a higher rate of postoperative complications than asymptomatic children, with 52% and 15.4%, respectively ($P=0.028$). The median length of stay for symptomatic children was 17 days, compared to 11 days for asymptomatic children ($P=0.280$).

Conclusions Early surgery of CLM in asymptomatic children was associated with a lower rate of postoperative complications. Further studies are needed to investigate long-term complications.

Keywords Congenital lung malformation, Surgery, Children, Outcome

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Background

Congenital lung malformation (CLM) is an umbrella term for various conditions including congenital pulmonary airway malformation (CPAM), pulmonary sequestration, bronchogenic cyst, congenital lobar overinflation, and bronchial atresia. This is a relatively uncommon disease, with an incidence of around 30–42 cases per 100,000 population [1]. Prenatal ultrasonography has made it possible to detect this condition more frequently. However, the actual prevalence may be higher than reported since some patients are discovered incidentally without symptoms [2].

Newborns born with CLM may experience significant complications, including fetal hydrops, which can be found in 5–30% of cases [3]. Additionally, even after the neonatal period, approximately 3.2% of undiagnosed or untreated patients may develop subsequent complications such as recurrent pulmonary infections, pulmonary hemorrhage, pleural effusion, decreased lung function due to compressed lung parenchyma, or the occurrence of malignancies such as pleuropulmonary blastoma, bronchoalveolar carcinoma, and adenocarcinoma [4, 5].

Surgical intervention or embolization, which involves blocking the abnormal blood vessels that supply the lung tissue, is recommended for patients experiencing symptoms. The primary goal of surgery is to remove the affected lung tissue, thereby reducing lung compression and the risk of subsequent complications. Additionally, it allows the remaining lung tissue to expand and function better [6]. In patients who do not experience any symptoms, there is currently no clear treatment approach. Surgical resection is the definitive treatment to mitigate these risks, but the debate persists regarding whether early intervention in asymptomatic children offers superior outcomes compared to waiting for symptom development. Therefore, our study aims to investigate the outcomes of surgery for CLM compared to cases where symptoms are absent.

Methods

Study design and participants

A retrospective review was conducted on all children under the age of 18 who were admitted to Srinagarind Hospital, Khon Kaen University between January 1, 2007, and December 31, 2023. The study's main inclusion criteria were children with CLM who underwent surgical resection during the study period, while children with cystic lung lesions from other causes and CLM who did not undergo surgical resection were excluded. The study followed the Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) guideline.

Data collection

The objective is to review the electronic medical records in Health Objects of Srinagarind Hospital, Khon Kaen University. The diagnosis of CLM was identified using the International Statistical Classification of Diseases and Related Health Problems, 10th Revision, Thai Modification (ICD-10-TM) code Q33.0, Q33.2, Q33.8, Q33.9, and J43.0. We recorded demographic data including age at diagnosis, gender, age at surgery, diagnosis, and location of the lesion. We also gathered details about the surgery, perioperative data, and operative outcomes. Operative outcomes encompassed operative time, duration of intubation, length of stay in the intensive care unit (ICU), length of hospital stay, postoperative complications, and mortality. Postoperative complications were defined as condition-related operations that presented after the surgery.

Statistical analysis

The data analysis included presenting categorical data as numbers and percentages and comparing them using Chi-square or Fisher's exact test. Continuous data were represented as the median with interquartile range (IQR) depending on the data distribution as determined by the Shapiro-Wilk test. For non-normally distributed data, the Mann-Whitney U test was used. A p-value of < 0.05 was considered statistically significant. The data analysis was carried out using STATA software version 10 (Stata Corp., College Station, TX, USA).

Results

Baseline characteristics

During the study period, 54 children were diagnosed with CLM. There were 19 children excluded from our study. Seven children with CPAM were diagnosed prenatally and experienced spontaneous resolution of their lesions after birth. Additionally, three children with CPAM and two with bronchopulmonary sequestration were asymptomatic and awaiting surgical intervention. Furthermore, two children with CPAM and one with bronchopulmonary sequestration were lost to follow-up. The child who passed away before surgery was diagnosed with CPAM, alongside co-morbidities, including ruptured myelomeningocele, and subsequently developed severe sepsis. A total of 38 children with CLM underwent surgical resection and were included in the study (Fig. 1.). Of these patients, 21 (55.3%) were male. Only 10 (26.3%) children had a prenatal diagnosis, and 2 (8%) of them were symptomatic, while the remaining 8 children (61.5%) were asymptomatic. Prior to their surgery, 25 children (65.8%) exhibited symptoms such as respiratory distress, history of previous pulmonary infection, pneumothorax, and hemoptysis, while 13 children (34.2%) were

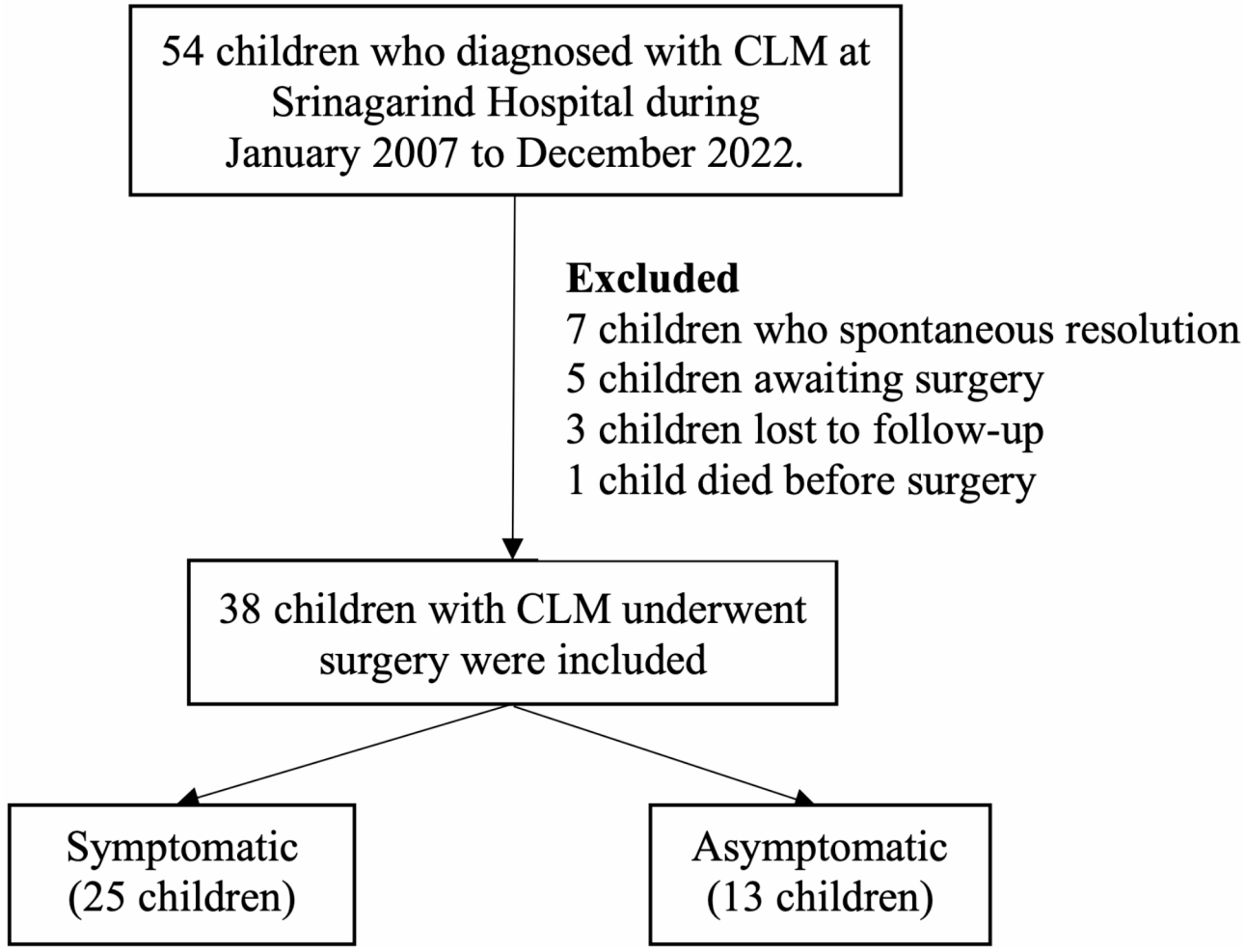

Fig. 1 Flow diagram of children diagnosed with congenital lung malformation (CLM) included in the study

Table 1 Baseline characteristic of children diagnosed with congenital lung malformation who underwent surgery

Characteristic	Total (*n* = 38)	Symptomatic (*n* = 25)	Asymptomatic (*n* = 13)	*p*-value
Gender, n (%)				0.575
Male	21 (55.3)	13 (52.0)	8 (61.5)	
Female	17 (44.7)	12 (48.0)	5 (38.5)	
Prenatal diagnosis, n (%)	10 (26.3)	2 (8.0)	8 (61.5)	< 0.001
Age at diagnosis (months), median (IQR)	9 (1–33)	15 (3–33)	1 (1–3)	0.181
Age at surgery (months), median (IQR)	8.5 (3–28)	12 (3–32)	6 (3–12)	0.201
Lesion location, n (%)				0.668
Left lung	14 (36.8)	8 (32.0)	6 (46.2)	
Right lung	23 (60.5)	16 (64.0)	7 (53.8)	
Mediastinum	1 (2.6)	1 (4.0)	0 (0)	

asymptomatic. All patients underwent surgical resection via open thoracotomy (Table 1).

The median age at diagnosis and surgery was higher in the symptomatic group compared to the asymptomatic group, 15 months (IQR 3–33 months) and 1 month (IQR 1–3 months), respectively ($p = 0.18$). The median age at surgery was 12 months (IQR 3–32 months) and 6 months (IQR 3–12 months) for the symptomatic group and the asymptomatic group ($p = 0.201$). In the symptomatic group, the lesions were left-sided in 8 children (32%)

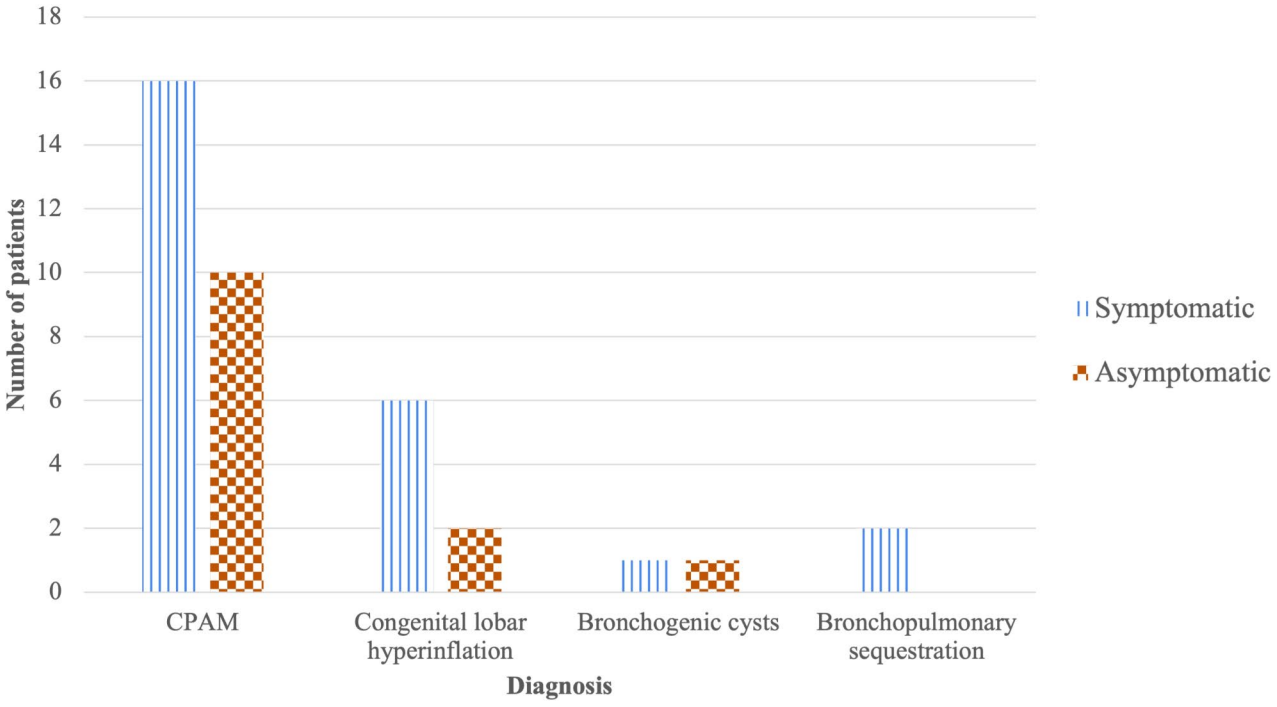

Fig. 2 The number of children diagnosed with congenital lung malformations (CLM) categorized by symptomatic and asymptomatic cases

Table 2 Operative outcomes of children diagnosed with congenital lung malformation who underwent surgery

Characteristic	Symptomatic ($n=25$)	Asymptomatic ($n=13$)	*p*-value
Operation time (minutes), median (IQR)	145 (125, 165)	135 (115, 180)	0.548
Duration of intubation (hours), median (IQR)	42 (16, 139)	45 (23, 117)	0.805
Duration of ICU (hours), median (IQR)	80 (45, 184)	71 (44, 141)	0.633
Length of hospital stay (days), median (IQR)	17 (8, 26)	11 (8, 15)	0.280
Postoperative complication, n (%)	13 (52)	2 (15.4)	0.028

Abbreviations: ICU, intensive care unit; IQR, interquartile range

and right-sided in 16 children (64%). The lesion was in the mediastinum in 1 child (4%). In the asymptomatic group, the lesions were located left-sided in 6 children (46.2%) and right-sided in 7 children (53.8%). There was no significant difference in the lesion location between the groups. CPAM was the most common type of CLM in our study (26 children, 68.4%), followed by congenital lobar overinflation (8 children, 21%), bronchopulmonary sequestration (2 children, 5.3%), and bronchogenic cyst (2 children, 5.3%), respectively. The comparison of the number of diagnoses distinguishing between those exhibiting symptoms and those classified as asymptomatic was presented in Fig. 2.

Operative outcomes

We found that symptomatic patients had a longer median time of operation than asymptomatic patients, 145 min (IQR 125–165 min) compared with 135 min (IQR 115–180 min), $p=0.548$. The median duration of postoperative intubation in symptomatic children was 42 h (IQR 16–139 h), while it took 45 h (IQR 23–117 h) among asymptomatic children ($p=0.805$). Symptomatic children stayed in the ICU for a median time of 80 h (IQR 45–184 h) compared to 71 h (IQR 44–141 h) for asymptomatic children ($p=0.633$). The median time of hospital stays was also longer for symptomatic children than for asymptomatic children, but no statistical significance ($p=0.28$) (Table 2).

Postoperative complications were significantly higher in symptomatic children (13 children, 52.0%) compared to asymptomatic children (2 children, 15.4%), $p=0.028$. Ventilator-associated pneumonia was the most common complication (5 children, 13.1%). Other complications included hospital-acquired pneumonia, pneumothorax, pleural effusion, atelectasis, septicemia, pulmonary hemorrhage, and residual lung cyst. There was only one death in the study, which was caused by ventilator-associated pneumonia and *K. pneumoniae* septicemia. This patient

had undergone surgery for CPAM at the age of 5 days and required mechanical ventilation prior to the operation.

Discussion

CLM represents a heterogeneous group of abnormalities. Several children exhibited complications associated with CLM, including fetal hydrops and lung hypoplasia, beginning from the prenatal period [3]. Although the number of cases that are suspected or diagnosed early has increased, many management questions remain unanswered. Especially in cases where children show no symptoms, there is no universally accepted surgical management [3]. While there is a general consensus that any symptomatic CLM should be treated, the best management of asymptomatic children remains controversial [6]. In this retrospective study, we analyzed children with CLM who underwent surgical resection at a tertiary care center in Thailand. Most of them had respiratory symptoms before surgery.

Our study found that children who had surgery while experiencing symptoms had a higher incidence of postoperative complications compared to those who did not have symptoms. This finding is consistent with previous research in the field [7–9]. Our research revealed that the most common complication encountered was ventilator-associated pneumonia. There are studies that suggest that prior infections can lead to the scarring of lung tissue, which can make surgeries more difficult. This could result in an increased number of complications post-surgery. Thus, our findings support the notion of undertaking an early resection before any symptoms begin to manifest.

The adoption of early surgical resection as a customary procedure may effectively mitigate the likelihood of severe respiratory complications and abbreviate hospital stays, thereby contributing to enhanced patient outcomes and decreased healthcare expenditures. This is consistent with previous research demonstrating that early resection in asymptomatic children results in improved postoperative recovery and fewer complications [10–12]. Adopting these findings into routine clinical protocols, coupled with the development of guidelines for the management of asymptomatic CLM, could provide clinicians with a clearer decision-making framework and optimize patient care.

Our study demonstrates that early surgical intervention in asymptomatic children with CLM potentially enhances lung growth and regeneration. Although the alveolarization process remains incompletely comprehended [13], the dominant theory holds that alveoli cease to proliferate by 2–3 years of age [14] and subsequently experience a growth in both volume and surface area. However, certain authors have proposed that alveolar formation could persist for an additional 7–8 years of age [15]. By performing surgical resections at a younger age, we can take advantage of this critical window for lung growth. Additionally, human hepatocyte growth factor (hHGF) has been identified as the most potent mitogen for alveolar type II cells. It plays a crucial role in the repair of the alveolar epithelium and compensatory lung growth. A study evaluating changes in serum hHGF levels in patients undergoing thoracic surgical procedures found that hHGF levels significantly increased following lung resection, indicating its role in lung regeneration [16]. These findings suggest that early resection in clinical practice could reduce immediate complications and foster optimal lung growth during this crucial developmental phase. This approach underscores the importance of developing robust clinical guidelines that advocate for early surgical intervention in asymptomatic children, ensuring that therapeutic decisions are grounded in both immediate and long-term benefits.

Our study did not reveal a significant difference in operative time, length of ventilation, and hospital stay between the two groups. These results contrast with the previous study [10]. In 2021, Elhattab et al. compared surgical outcomes via thoracoscopic surgery in 28 patients with a history of pulmonary infection and 62 patients with no such history. They reported that the operative time was significantly shorter in asymptomatic patients [8]. However, data regarding the frequency of infections or the duration from the onset of symptoms to surgery were not collected. Recurrent episodes of infection may lead to increased lung fibrosis, potentially affecting surgical outcomes [17].

Our study suggests that patients who display symptoms tend to have longer stays in the hospital as compared to those who are asymptomatic. Although the difference is not statistically significant, the median duration of hospitalization for symptomatic patients was 17 days, while for asymptomatic patients it was 11 days. The reason behind this could be a higher occurrence of postoperative complications among symptomatic patients. Consistent with the previous study in Belgium, they discovered postoperative complications and longer hospital stays were significantly higher in patients with preoperative clinical signs of CPAM [18].

Children with CLM who present asymptomatically and undergo early surgical resection in our study encounter a markedly reduced incidence of postoperative complications in comparison to those who exhibit symptoms. This observation lends support to the expanding corpus of evidence indicating that implementing early intervention measures before symptoms manifest could potentially result in improved surgical outcomes and decreased healthcare costs. In addition, to guide clinical decision-making, our findings highlight the significance of developing standardized protocols for the management of asymptomatic CLM. Considering the constraints of our retrospective study conducted at a single center, it is recommended that further prospective multicenter investigations be undertaken to

validate these findings and examine the enduring clinical consequences, such as pulmonary function and quality of life, in older children and adolescents.

This retrospective study included only children who underwent surgical resection, excluding those who spontaneously regressed or remained under observation, which may introduce selection bias. Additionally, we did not determine the optimal age for surgery in asymptomatic patients or document the rationale behind such decisions, highlighting the need for prospective research. Our small sample size reduced statistical power, and as a single-institution study, the findings may not be widely applicable. Furthermore, we did not collect detailed data on intraoperative complications; however, no redo surgeries or recurrent symptoms were observed. Only short-term outcomes were evaluated, so long-term results remain unknown, underscoring the necessity of larger, multi-center studies with standardized follow-up protocols.

Conclusions

Early surgery of CLM in asymptomatic children was associated with lower rate of post operative complications. Further studies are needed to investigate long-term complications.

Abbreviations

CLM	Congenital lung malformation
CPAM	Congenital pulmonary airway malformation
ICU	Intensive care unit
IQR	Interquartile range

Acknowledgements

We express our thanks to all the residents, fellows, and staff of Srinagarind Hospital, Khon Kaen University, for their invaluable support and dedicated care for our patients.

Author contributions

RU: Conceptualization, Data curation, Formal analysis, Investigation, Supervision, Validation, Writing – original draft, Writing – review & editing. PU: Data curation, Formal analysis, Investigation, Writing –original draft. SN: Supervision, Writing – review & editing. LT: Supervision, Writing – review & editing. SS: Supervision, Writing – review & editing. PS: Conceptualization, Data curation, Formal analysis, Investigation, Supervision, Validation, Writing – original draft, Writing – review & editing.

Funding

This study was supported by Research and Graduate Studies, Khon Kaen University, Thailand.

Data availability

No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate

The study protocol was approved by the human research ethics committee of Khon Kaen University (HE651065). The informed consent was waived as it was a retrospective observational study with no more than minimal risk.

Consent for publication

Not applicable.

Competing interests

The authors declare no competing interests.

Received: 6 July 2024 / Accepted: 8 February 2025
Published online: 15 February 2025

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