ORIGINAL ARTICLE

SURGICAL MANAGEMENT OF CONGENITAL THORACIC DISORDERS: A 15-YEAR CENTER EXPERIENCE

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Abstract

***Introduction**: Congenital thoracic disorders represent a spectrum of fetal lung bud development abnormalities, which may affect breathing capacity and quality of life. We aim to evaluate the impact of surgery in the treatment of 4 major congenital conditions.*

***Materials and Methods:** We performed a retrospective cohort analysis of patients who underwent surgical treatment in our tertiary center, from 2007 to 2022.*

***Results:** Over the 15-year period, we treated 33 patients, with a male predominance of 55%. 22 patients (67%) were asymptomatic. When symptomatic, the recurrence of respiratory infections was the most common clinical presentation (18%). In 13 patients (39%), diagnosis was achieved through fetal ultrasonography.*

This study encompassed 13 patients with pulmonary sequestration (39%), 11 patients with bronchogenic cysts (33%), 7 patients with congenital pulmonary airway malformation (21%) and 2 patients with congenital lobar emphysema (6%).

Considering solely lung malformation conditions, we accounted 22 patients with a median age of 3 [1-67] years-old. Surgery comprised bilobectomy (9%), lobectomy (77%), lobectomy with wedge resection (5%), segmentectomy (5%) and wedge resection (5%).

Concerning bronchogenic cysts, we treated 11 patients with a median age of 19 [14-66] years-old. We identified 1 hilar, 1 intrapulmonary and 9 mediastinal lesions, of which 4 were paraesophageal, 4 were subcarinal and 1 was miscellaneous.

Overall, surgery was conducted by thoracotomy in 61% of patients, VATS in 33% and RATS in 6%.

The median drainage time was 3 [1-40] days and median hospital stay was 4 [1-41] days. There were no cases of mortality. Ensuing, 94% of patients experienced clinical improvement after surgery.

***Conclusion:** Early diagnosis of congenital thoracic malformations increased considerably with the improvement in imaging technology and prenatal screening. Treatment may include expectant conservative treatment. However, in selected cases, surgery may play an important role in symptomatic control and prevention of disease progression.*

***Keywords**: Congenital, Thoracic Malformation, Congenital Lung Malformation, Congenital Pulmonary Airway Malformation, Bronchogenic Cyst, Pulmonary Sequestration, Congenital Lobar Emphysema*

INTRODUCTION

Congenital thoracic malformations represent rare developmental anomalies, encompassing a large spectrum of disorders, with particular emphasis on congenital pulmonary airway malformation (CPAM), pulmonary sequestration, congenital lobar emphysema (CLE) and, bronchogenic cyst, among other entities.[1,2] These disorders show an overlapping biological genesis as well as similar clinical features and pathologic outcomes and, as a result, they are often considered conjointly for the purpose of discussion and treatment evaluation.[2]

Literature demonstrates a recent increase in the incidence of congenital thoracic malformations. Historically, they were estimated between 1 per 11,000 to 35,000 births[3-5] while currently, they can be identified in 1 per 2,500 births[1,5]. This upsurge seems to be due to an improvement in fetal ultrasound sensitivity and comprehension as well as

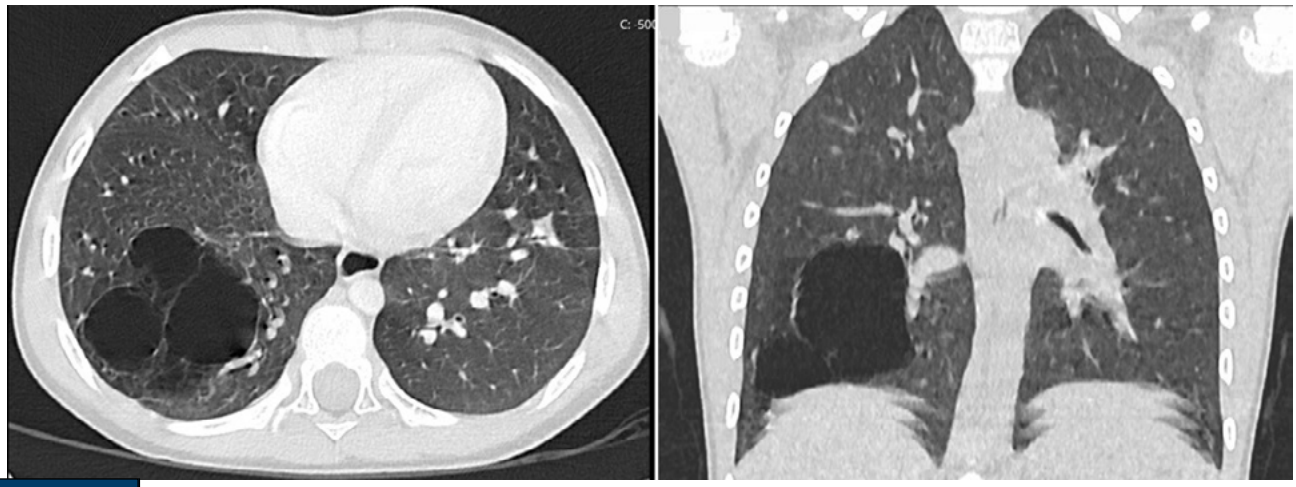

Figure 1 *CPAM type I: Preoperative Thoracic CT-scan.*

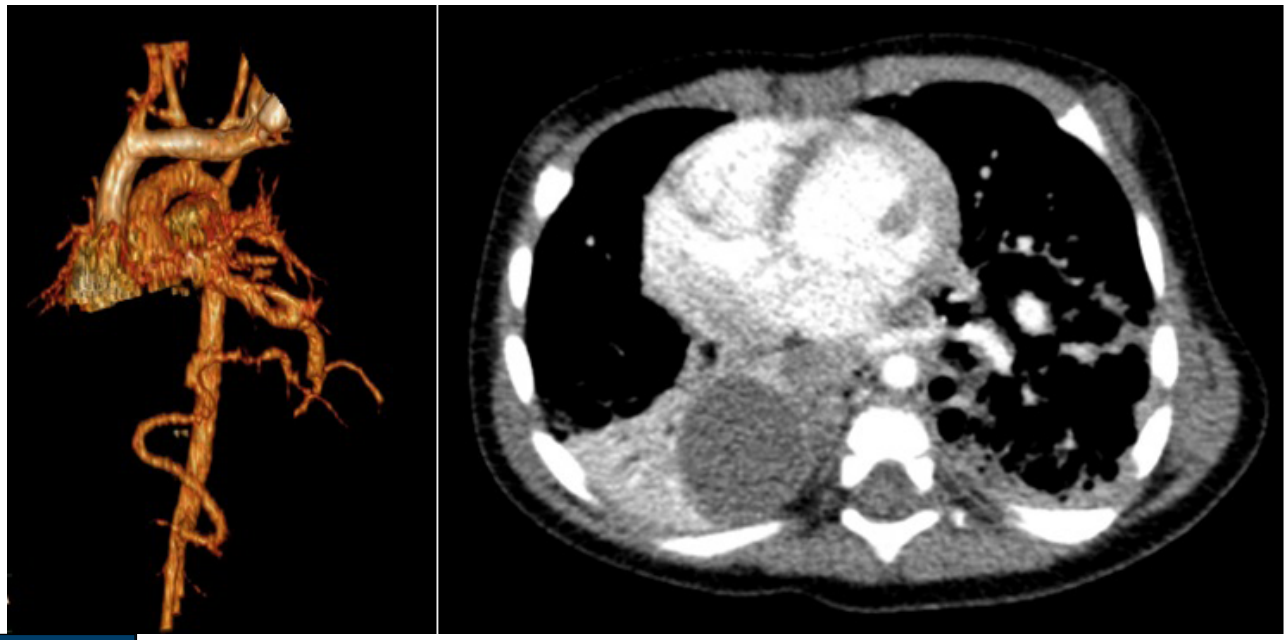

Figure 2 *Pulmonary Intralobar Sequestration: Preoperative Thoracic CT-scan.*

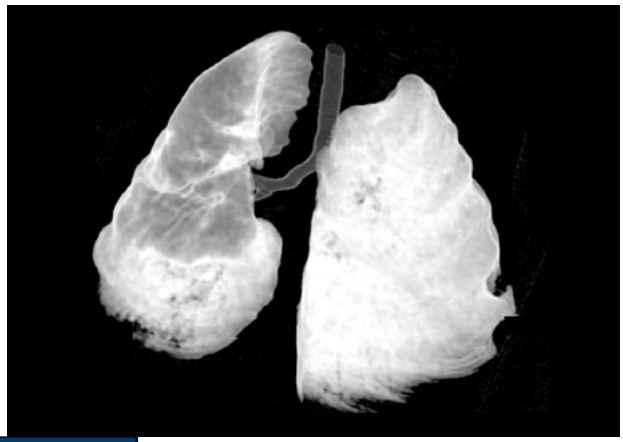

Figure 3 *CLE: Preoperative Thoracic CT-scan.*

widespread accessibility of prenatal screening programs.[5]

Congenital thoracic malformations tend to remain stable or even regress during pregnancy. The majority of patients are asymptomatic at birth with radiologically parenchymal abnormalities.[4,6] However, symptoms may develop with postnatal respiratory distress or, ensue during infancy or early adult life, with respiratory complications, comprehending recurrent infection, pneumothorax, hemothorax, hemoptysis or the rare development of lung malignancy.[4,6]

CPAM is the most common fetal lung disorder (30-40%), resulting from an abnormal overgrowth of distal bronchial and alveolar structures, as represented in figure 1. This malformation preserves the communication with the

Table 1 **Patient Characteristics**

	n (%)
Gender	
Male	18 (55)
Mean Tobacco Exposure (pack-years)	33
Comorbidities	
Hypertension	6 (18)
Chronic Obstructive Pulmonary Disease	2 (6)
Diabetes Mellitus	2 (6)
Hemolytic Anemia	2 (6)
Chronic Kidney Disease	1 (3)

airway and pulmonary blood supply, although generally incapable of adequate gas exchange.[1,3,5] The modified Stocker classification (initially proposed in 1977 and later improved in 2002), proposes five types of CPAM according to the cyst level and histopathological characterization.[5]

Bronchopulmonary sequestration consists of a section of nonfunctional lung parenchyma with no connection with the tracheobronchial tree and sustained through an aberrant arterial supply. There are two categories: the intralobar sequestration is the most frequent subtype (75%) and it is located within the lobar structure and surrounded by normal lung parenchyma, as seen in figure 2, and, the extralobar subtype identified outside of the lobar structure, with systemic venous drainage.[1,3,5] In this condition due to the abnormal arterial irrigation, percutaneous embolization emerged recently as a therapeutic option, that may complement or constitute an alternative to surgery, in selected patients, allowing for occlusion of the arterial supply with consequent involution of the sequestration.[7,8]

CLE is characterized by progressive hyperinflation and air trapping of one of the pulmonary lobes (most frequently the upper left lobe), leading to atelectasis of the functioning parenchyma.[9,10] The primary explanation is airway collapse during expiration, stemming from cartilaginous deficiency and consequent emphysematous lobe distortion, as perceived in figure 3.[1]

Finally, bronchogenic cysts are mucus-filled lesions, lined by respiratory epithelium and with a defined cartilaginous wall, arising from an abnormal development of the lung bud (figure 4). They can be located intrapulmonary or contained in the mediastinum (Maier's classification comprehends 5 possible locations).[11,12]

Management of congenital thoracic malformations may comprise expectant monitoring or surgery. In symptomatic patients, surgical treatment has soundly demonstrated a positive impact in symptom control and prevention of disease progression whereas in asymptomatic patients at birth, its role remains controversial.[1,3,4] Furthermore, the optimal timing and degree of surgical resection has not been established.[4]

The present study reviewed the clinical features and management of patients with congenital thoracic malformations referred to our center for surgical treatment over the past 15 years. The objective is to report our experience and highlight the impact of surgery in the treatment of these congenital disorders.

METHODS

A retrospective cohort study was performed of all patients with congenital respiratory malformation pathologies who underwent surgery at the Thoracic Surgery department of a tertiary center, comprehended between 2007 and 2022.

Data was obtained through review of medical records. Preoperative information included personal demographic characteristics (such as age and gender), smoking habits when applicable, medical history, symptoms at presentation, timing of diagnosis (prenatal or postnatal), preoperative imaging, laterality of the malformation, age at the time of the surgical procedure and span between diagnosis and surgical treatment. Compilation of intraoperative variables comprehended surgical approach, extent of resection, need for conversion to open surgery and intraoperative findings. Additionally, postoperative information comprised histopathology, imaging reevaluation, symptomatic status, duration of chest drainage and hospital stay, evidence of complications and length of follow-up by the thoracic surgical team. No patient was lost during follow-up.

Complications occurring during or within 60 days of surgery were stratified according to the Clavien-Dindo classification. Minor morbidity comprehended type I-II whereas major surgical morbidity included type III-IV Clavien-Dindo complications.

Categorical variables are presented in absolute value and percentage format and the continuous variables with median value and range. Statistical analysis was performed using IBM SPSS 27 software

RESULTS

Over the 15-year period (2007 to 2022), 33 patients were treated in our center, displaying male predominance (55%, n=18). 24% of patients (n=8) were smokers with a mean amount of 33 (±21) pack-years. 9 patients presented with known history of major medical comorbidities; arterial hypertension being the most frequent (18%, n=6). The demographic features are compiled in table 1.

The clinical presentation of congenital thoracic malformations was variable. The majority of patients (67%, n=22) were asymptomatic but showed persistent radiological abnormalities. When symptomatic, recurrent respiratory infections was the most common symptom, occurring in 18% of patients (n=6). A summary of the clinical presentation is present in table 2.

Antenatal diagnosis was made in 13 patients (39%)

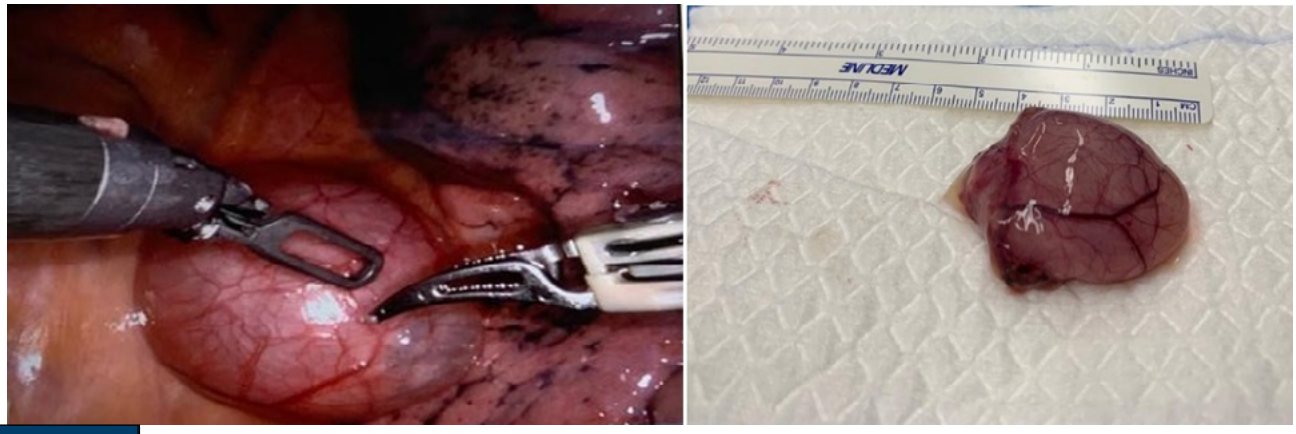

Figure 4 *Bronchogenic Cyst: Intraoperative Lesion.*

through fetal ultrasonography. Diagnostic evaluation included a thoracic radiography and CT scan for all patients. Preoperative assessment also comprised clinical history, physical examination, routine blood tests, electrocardiogram and when possible, spirometry (n=9).

This cohort encompassed 13 patients with pulmonary sequestration (39%), 11 patients with bronchogenic cysts (33%), 7 patients with CPAM (21%) and 2 patients with CLE (6%).

Considering solely lung malformation conditions (pulmonary sequestration, CPAM and CLE), we account for 22 patients with a median age of 3 [1-67] years-old.

In terms of pulmonary sequestration, 77% (n=10) constituted intralobar sequestrations. In turn, the majority of patients with CPAM were type I (57%, n=4) while the remaining, were type II in 29% and type IV in 14%.

Surgery comprised bilobectomy (9%), lobectomy (77%), lobectomy with wedge resection (5%), segmentectomy (5%) and wedge resection (5%). In 55% of patients, surgery was performed on the left lung. Thoracotomy was the favored approach, employed in 82% of patients (n=18). In this sense, the reminiscing 18% (n=4), in this case, amounting to lobectomies, were performed through video-assisted thoracoscopic surgery (VATS).

Concerning bronchogenic cysts, 11 patients were contemplated, showcasing a median age of 19 [14-66] years-old. They presented as 1 hilar, 1 intrapulmonary and 9 mediastinal bronchogenic cysts, of which, according to Maier's classification, 4 were paraesophageal, 4 were subcarinal and 1 was miscellaneous (transdiaphragmatic).

Surgery consisted of excision of the lesions. The intrapulmonary cyst required middle lobe lobectomy. Conversely, the miscellaneous cyst due to transdiaphragmatic extension, demanded diaphragmatic left pillar disinsertion in order to allow for the resection of the lesion and subsequent suture of the diaphragmatic defect.

Complete resection (R0) was achieved in 97% (n=32) of patients. There was one case of planed partial excision of the bronchogenic cyst (R2) due to strict adherences to the left atrial heart appendage, distal trachea and carina. It was a subcarinal bronchogenic cyst of around 6 cm in diameter and surgery consisted of partial excision with ample opening of the wall structure, mucus content drainage and double cerclage of the base of the lesion.

We performed VATS in 64% (n=7) of the bronchogenic cysts and robotic-assisted thoracoscopic surgery (RATS) in 18% (n=2). 2 VATS procedures were converted to thoracotomy. In one case due to iatrogenic injury of the main bronchus, there was need for correction suture, whereas in the case of the transdiaphragmatic bronchogenic cyst, technical surgical difficulties dictated preventive conversion to open surgery.

Overall, surgery was conducted by thoracotomy in 61% (n=20) of patients, VATS in 33% and RATS in 6%. The conversion rate to open surgery was 6% (n=2).

The distribution of surgical approach and type of resection, according to the pathological diagnosis is summarized in tables 3 and 4. The median time between diagnosis and surgical treatment was 20 months [1-84 months].

Additionally, all patients had a chest tube placed intraoperatively. 6 patients, submitted to open surgery, required more than one chest tube. The median drainage time was 3 [1-40] days and comparably, the median hospital length of stay was 4 [1-41] days. The difference in median length of pleural drainage between minimally invasive and open surgery (2 vs. 3 days) was not statistically significant (p=0.099) and though, the length of hospital stay was

Table 2 **Clinical Presentation**

Clinical Presentation	At Diagnosis n (%)	After Surgery n (%)
Asymptomatic	22 (67)	31 (94)
Recurrent Respiratory Infections	6 (18)	2 (6)
Cough	3 (9)	-
Pain	2 (6)	-
Dyspnea	1 (3)	-
Hemoptysis	1 (3)	-

Table 3 **Surgical Ressection**

	n (%)
Pulmonary Sequestration	13 (39)
Lobectomy	10
Bilobectomy	2
Wedge Resection	1
Bronchogenic Cysts	11 (33)
Complete Excision	9
Partial Excision	1
Lobectomy	1
Congenital Pulmonary Airway Malformation	7 (21)
Lobectomy	6
Lobectomy + Wedge Resection	1
Congenital Lobar Emphysema	2 (6)
Segmentectomy	1
Lobectomy	1

Table 4 **Surgical Approach**

	n (%)
Pulmonary Sequestration	13 (39)
Thoracotomy	10
VATS	3
Bronchogenic Cysts	11 (33)
VATS	7
RATS	2
Thoracotomy (conversion)	2
Congenital Pulmonary Airway Malformation	7 (21)
Thoracotomy	6
VATS	1
Congenital Lobar Emphysema	2 (6)
Thoracotomy	2

shorter in minimally invasive surgery (2 vs 5 days), was also not statistically significant (p=0.033).

Major surgical morbidity rate was 9%, concerning Clavien Dindo type III to IV complications. These were identified in patients submitted to open surgery. Postoperative complications included development of empyema, that needed chest tube placement and intravenous antibiotic treatment; hemothorax, demanding of surgical hemostasis and, prolonged air leak, ultimately requiring surgical pneumostasis.

The median postoperative follow-up time by the thoracic surgical team was 12 [1-66] months. Afterwards, all patients maintained regular medical surveillance by the referral physician (commonly, in Pediatrics or Pulmonology consultation).

Clinical improvement after surgical treatment was verified in 94% of patients (n=31), who remain asymptomatic, as presented in table 2.

There was no evidence of mortality. To this day, overall survival rate remains 100%.

DISCUSSION

Congenital thoracic malformations represent a spectrum of conditions resulting from respiratory system growth anomalies during the fetal period. The incidence of congenital defects has ascended hand in hand with the improvement in the diagnostic assessment, predominantly in the prenatal phase, underlining the importance of defining an adequate plan of care.[2,4]

In symptomatic patients, surgical resection is the optimal treatment, allowing for symptom control and prevention of disease complications, such as oppressive recurrent respiratory infections that may lead to diminishing lung capacity.[3]

However, symptomatic patients constitute only 25% while for the remaining 75% of patients, who are initially asymptomatic at birth,[5] the role of surgical resection is not as well established.

The risk of mounting complications is generally one of the most accepted indications for surgery in asymptomatic patients.[1,5] The true risk of developing respiratory complications has not been determined. A recent meta-analysis described 505 asymptomatic antenatal detected cases, of which only 3,2% became symptomatic at a median age of 6,9 months. When symptoms did develop, they were generally more severe.[6] The compiled reports were limited by short follow-up periods that may underrate respiratory implications. [1,6]

Another meta-analysis determined that 25% of patients, specifically diagnosed with CPAM, went on to develop symptoms at a median age of 7 months. [13] Moreover, a Japanese multicentric nationwide survey of 428 patients, showed 65% of patients developed symptoms before the age of 3. Additionally, this study supported that early surgical resection before lung infection allowed

for significant recovery of the remaining lung, measured though higher vital capacity.[14]

Multiple studies have been conducted, investigating the repercussion of surgery in pulmonary function tests and lung capacity. They show a generally preserved total lung capacity, normal to elevated residual volume and lower forced expiratory volume (FEV1),[4,5] suggesting compensatory growth of the remaining lung after parenchyma removal. Evidence has not proved lung regeneration after surgery, even in earlier ages.[5]

The optimal timing for surgical treatment is difficult to define but early surgical procedures seem to deter disease progression, preventing repeated and prolonged hospitalizations and averting technical difficulties in surgical dissection in consequence of infectious flares. [3] In this sense, Naito et al, performed a retrospective study in 2012 that concluded there was a trend favoring lower levels of maximal oxygen uptake during exercise (VO2max) in delayed surgical procedures.[4]

At our institution, we propose surgery as a treatment option in symptomatic patients or in larger lesions with considerable lung parenchyma distortion. When possible, we prefer to delay the procedure until the child is 1-2 years of age and can better tolerate and recover after surgery, allowing for an easier perioperative management. In patients with small lesions and minimal disease affection, conservative treatment with close radiological monitoring may be acceptable. Surgery can be reconsidered in case of lesion growth or symptomatic development.

Open surgery remains the preferred approach concerning the management of congenital thoracic malformations in early infancy. Thoracoscopy is an accepted safe and effective approach; however, in small children some challenges have limited its application, namely reduced lung reserve that may restrict the use of one lung ventilation and hinder surgical dissection as well as a narrow thoracic cavity space and delicate anatomic structures. Kulaylat et al, performed a propensity score matching of 258 pediatric patients and showed there was no significant statistical difference in surgical outcome, length of hospital stay or postoperative complications between the two techniques.[2,9] Established advantages of thoracoscopic surgery are the minimized musculoskeletal injury and the cosmetic aspect of reduced incisions. [1-3,9] Previous infection is a major risk factor for conversion to open surgery. [5]

In our study, thoracotomy was the more prevalent approach in surgical resection of congenital lung malformations, with the exception of excision of bronchogenic cysts, in which VATS prevailed. There was also no significant difference in terms of length of pleural drainage and hospitalization. Patients showed a positive outcome to surgical treatment with low morbidity rate and high symptomatic improvement. We aim to employ minimally invasive techniques more and more as we ascend to increasingly more complex procedures. The RATS program in our center was introduced in March of 2022, justifying the small sample of cases. Further investigation is necessary to evaluate the role of robotic-assisted surgery in the treatment of these pathologies.

The extent of resection is another matter of debate. Formal lobectomy is predominant as reflected in our article; but parenchyma-saving resections with employment of segmentectomy and wedge resection should be considered in a case-to-case basis. The most important consideration is allowing for the complete removal of the lesion.

Recent cohorts such as Fasceitti-Leon et al and Bagrodia et al, compared patients submitted to lobectomy and segmentectomy, showcasing, in the latter, a good clinical outcome associated with a reduced hospitalization time. These surgical resections may be performed safely in smaller lesions while conserving healthy lung parenchyma.[1,3]

Lastly, a comprehensive preoperative plan and elective surgery are associated with better outcomes and lower rate of postoperative complications when compared with emergency procedures.[1,6]

Limitations to our study are the retrospective nature and inherent small sample of cases considered. Moreover, only patients with congenital thoracic malformations referred for surgical treatment evaluation were considered.

CONCLUSION

Significant improvements have been reported in comprehending the behavior and treatment response of congenital thoracic malformations. Their incidence has increased in recent years parallel to the upsurge in prenatal imaging diagnosis.

Evidence favors surgical treatment in symptomatic patients but is more controversial when concerning asymptomatic patients. The decision for surgery lies in the balance between the lifelong risk of complications and respiratory function decline and the risk of postoperative complications.

Our cohort demonstrates low morbidity and mortality rates and positive clinical outcomes, highlighting the role of surgery in symptomatic control and prevention of disease progression in congenital thoracic malformations.

Further investigation with a larger sample of patients is of paramount importance to confirm our study results.

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Conflicts of interest

The authors have no conflicts of interest to declare concerning this work.

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