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Postoperative outcomes in symptomatic pediatric patients with congenital pulmonary airway malformations

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Abstract

Objectives: The aim of our study was to investigate the short-term postoperative outcomes of symptomatic congenital pulmonary airway malformations (CPAMs) and compare them with asymptomatic CPAMs.

Patients and methods: Between January 2007 and January 2024, medical records of a total of 18 patients (10 males, 8 females; median age: 120 days; range, 4 to 2,160 days) who were operated for CPAM and whose CPAM was confirmed histopathologically were retrospectively analyzed. The patients were divided into two groups: Group 1 included symptomatic patients, while Group 2 included asymptomatic patients. Presence of postoperative complications such as oxygen requirement, ventilator requirement, pneumothorax, persistent air leak (≥7 days), pleural effusion, transfusion requirement, surgical site infection, postoperative pneumonia and sepsis were recorded.

Results: The median postoperative follow-up was 29.2 (range, 1 to 84) months. The surgical procedure in all patients was thoracotomy. Group 1 had a higher postoperative complication rate and all patients with complications in this group were younger than three months. The postoperative oxygen requirement rate was significantly higher in Group 1 compared to Group 2 ($p<0.05$). In addition, in Group 1, the median duration of hospitalization was prolonged, which was associated with the occurrence of complications ($p<0.05$). There was no significant relationship between the chest tube duration, operation technique, and age in Group 1 and Group 2 ($p>0.05$).

Conclusion: The presence of symptomatic CPAM and surgery before the age of three months may increase the risk of postoperative complications, particularly postoperative oxygen requirement.

Keywords: Congenital pulmonary airway malformations, postoperative complications, symptomatic.

Congenital lung malformations (CLMs) encompass a varied group of disorders, which include congenital pulmonary airway malformations (CPAMs, previously known as congenital cystic adenomatoid malformation), bronchopulmonary sequestration, congenital lobar overinflation (previously known as congenital lobar emphysema), and bronchogenic cysts. Congenital pulmonary airway malformations characterized by an abnormal overgrowth of terminal bronchioles and development of alveoli are one of the most common CLMs, accounting for 30 to 40% of all diagnosed CLMs.[1] According to a European registry, its prevalence is estimated at 0.81/10,000 fetuses.[2] The majority of these lesions are diagnosed before birth through routine prenatal ultrasound.[2,3] Treatment in symptomatic patients is surgical excision. It is recommended that asymptomatic lesions be electively resected in the first few years of life due to the risk of recurrent infection and malignant transformation.[3]

In the present study, we aimed to assess the effect of symptomatic course of disease on postoperative outcomes of patients with CPAM who underwent lobectomy or segmentectomy.

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PATIENTS AND METHODS

This single-center, retrospective study was conducted at Bursa Uludağ University Faculty of Medicine, Department of Pediatric Surgery between

January 2007 and January 2024. Medical records of a total of 18 patients (10 males, 8 females; median age: 120 days; range, 4 to 2,160 days) who were operated for CPAM and whose CPAM was confirmed histopathologically were analyzed. Demographic features, clinical findings, pathological findings, treatment, and follow-up were analyzed. A written informed consent was obtained from the parents and/or legal guardians of the patients. The study protocol was approved by the Bursa Uludag University Health Research Ethics Committee (date: 03.04.2024, no: 2024-5/5). The study was conducted in accordance with the principles of the Declaration of Helsinki.

The patients were divided into the symptomatic group (Group 1, n=9) and asymptomatic group (Group 2, n=9). Those with complaints such as acute respiratory distress, pneumothorax, recurrent pneumonia, cough, and inability to feed were considered symptomatic. If these findings were not present, they were considered asymptomatic. Based on our clinical experience, in symptomatic cases in CPAM, surgery is performed even during the neonatal period. In asymptomatic cases, we adopt the surgical plan after computed tomography (CT) imaging in the postnatal third month or later. The preoperative surgical evaluation is based on CT images and segmentectomy or lobectomy are preferred techniques.

The outcomes of the patients were compared in terms of the presence of postoperative complications such as postoperative oxygen requirement, ventilator requirement, pneumothorax, persistent air leak (≥7 days), pleural effusion, transfusion requirement, surgical site infection, postoperative pneumonia, and sepsis.

Statistical analysis

Statistical analysis was performed using the IBM SPSS version 28.0 software (IBM Corp., Armonk, NY, USA). Descriptive data were presented in median (min-max) or number and frequency, where applicable. The Mann-Whitney U test was used for non-normally distributed data. Categorical variables were analyzed using the chi-square test and Fisher exact test. A p value of <0.05 was considered statistically significant.

RESULTS

In this study, a total of 18 patients (19 operations due to bilateral lesions) were included. Table 1 summarizes patient demographics, preoperative, and operative characteristics. Cardiac risk factors, gastrointestinal, and neurologic comorbidities were not significantly different across age groups (p>0.05). Four patients had minor congenital cardiac disease.

The median postoperative follow-up was 29.2 (range, 1 to 84) months. A total of 67% of the lesions were located on the right side (n=12), 22% on the left side (n=4), and 11% bilaterally (n=2).

TABLE 1
Demographic characteristics, clinical features and operative technique

	0-3 month age cases		Over 3 months age cases	
	Symptomatic (n)	Asymptomatic (n)	Symptomatic (n)	Asymptomatic (n)
Number of cases	6	3	3	6
Sex ratio				
Female	1	1	2	1
Male	1	2	1	2
Prenatal CPAM diagnosis history	3	3	0	5
Preoperative pneumonia history	1	0	3	0
Operative technique				
Lobectomy	6	2	1	4*
Segmentectomy	0	1	2	3*

CPAM: Congenital pulmonary airway malformation; * In one patient, lobectomy and segmentectomy were performed at separate times due to bilateral CPAM.

TABLE 2
Operation technique

	Lobectomy (n)	Segmentectomy (n)
Number of operation*	13	6
Affected lobe, right side		
Upper**	3	3***
Middle	1	0
Lower	5	1
Upper and middle	1	-
Affected lobe, left side		
Upper	2	2
Lower	1	0

CPAMs: Congenital pulmonary airway malformations; * In one patient, right lobectomy and left segmentectomy were performed at different times. The same patient is shown separately as lobectomy and segmentectomy in the table; ** In one patient, there was CPAM in bilateral upper lobes, but only right upper lobectomy was performed, the other side was followed; *** In one patient, right upper segmentectomy + simultaneous bronchogenic cyst removal were performed.

In cases younger than three months, the chief symptoms were respiratory distress and inability to feed, while in older infants, the main symptom was cough. In the preoperative period, four patients had a history of pneumonia, and one patient had pneumothorax. There were five symptomatic newborn cases.

All patients underwent either lobectomy or segmentectomy via open thoracotomy. Variations in the type of procedure performed were due to surgeon preference. In one of the two patients with bilateral lesions, lobectomy was performed on one side only, while in the other patient, lobectomy was performed on one side and segmentectomy on the other side with intermittent bilateral thoracotomy (Table 2). All pathological specimens were compatible with CPAM. In cases with lobectomy, the pathological diagnosis was compatible with

TABLE 3
Postoperative outcomes in Group 1 and Group 2

	Group 1 (n=9)				Group 2 (n=9)				
	n	%	Median	Min-Max	n	%	Median	Min-Max	*p*
Age at operation (Day)			30	4-2160			285	60-360	0.156
Mean operative time (min)			65	60-120			95	60-150	0.356
Mean chest tube duration (Day)			6	4-9			5.5	4-10	0.905
Mean length of hospital stay (Day)			8	6-17			6	5-12	**0.028**
Postoperative oxygen requirement	5	56			0	0			**0.011**
Postoperative ventilator requirement	1	11			0	0			0.474
Pneumothorax	2	22			1	10			0.582
Persistent air leak (≥7 days)	0	0			0	0			-
Pleural effusion	0	0			0	0			-
Surgical site infection	1	11			0	0			0.474
Bleeding requiring transfusion	1	11			0	0			0.474
Postoperative pneumonia and sepsis	2	22			0	0			0.211

Stocker's classification as type 1 in seven and type 2 in three patients, while in cases with segmentectomy, it was type 1 in two, type 2 in three, and type 4 in one patient. Pathological typing was not performed in three patients.

Postoperative outcomes are shown in Table 3. The median operating time was 65 min in the symptomatic group and 95 min in the asymptomatic group ($p>0.05$). Complications were seen in seven out of 18 cases, and most children with complications were in the neonatal period (5/7) and symptomatic (6/7). Although there was no significant difference between the groups in terms of age distribution ($p>0.05$), six patients with postoperative complications in the symptomatic group were younger than three months and five were in the neonatal period. The patients without complications in the symptomatic group were older than three months. Three of the asymptomatic patients were younger than three months and had no complications.

The overall complication rate was 67% in Group 1 and 11% in Group 2. The postoperative complications in Group 1 were postoperative oxygen requirement (56%), pneumothorax (22%), postoperative pneumonia and sepsis (22%), ventilator requirement (11%), transfusion requirement (11%), and surgical site infection (11%). However, only pneumothorax, which caused the thorax tube to remain for a long time, was observed in one patient in Group 2. In Group 1, two neonatal patients had a longer hospital stay due to postoperative pneumonia and sepsis.

There were significant differences between the two groups in terms of postoperative oxygen requirement ($p<0.05$). In the symptomatic group, the duration of hospitalization was prolonged due to complications ($p<0.05$). However, there was no significant relationship between the chest tube duration, operation technique, and age in either group ($p>0.05$). There was no operative mortality.

DISCUSSION

If symptoms are present, CLM can lead to a broad range of complications such as varying from fetal hydrops in utero to postnatal respiratory problems or infections. Varying degrees of respiratory distress occur in approximately 25 to 30% of infants during the postnatal period, and early surgery is required.[4] The symptoms in infancy and childhood are mostly related to infections and to respiratory distress associated with obstruction or spontaneous pneumothorax. Although many lesions would remain asymptomatic, there is a small risk of neoplasia.[5]

Patients with CPAM appear to have better perioperative outcomes before the beginning of symptoms, with shorter operating times, shorter postoperative mechanical ventilation, shorter chest tube duration, and shorter postoperative hospital stays.[6] In our study, the asymptomatic group had improved postoperative outcomes than the symptomatic group, including shorter postoperative oxygen requirement and shorter postoperative hospital stays. However, we found no significant difference in the chest tube time.

In the literature, the complication rates in symptomatic patients vary between 9.8 and 31.8%.[3,6,7] Xia et al.[8] reported that, compared to CPAM patients, asymptomatic patients were associated with improved clinical outcomes. They showed that 11 out of 19 patients in the symptomatic group had major postoperative complications (pleural effusion, hemothorax, and pneumothorax under tension). In our study, the complication rate was 67% (6/9) in the symptomatic group; however, we did not categorize complications as major and minor. Most children with complications were in the neonatal period and were symptomatic. In the study conducted by Engwall-Gill et al.[9] among newborns, the overall postoperative complication rate in CPAM cases was 45.5%. The high complication rate in symptomatic patients in our study compared to the literature can be explained by the employment of different criteria in published studies for symptoms and complications and the large number of newborn patients in the symptomatic group. Surgeries performed in the neonatal period are primarily done for emergency purposes. In a meta-analysis, Stanton et al.[10] reported that elective surgery was associated with significantly fewer complications than emergency surgery.

The management of symptomatic CPAM has been well established and resection should be done, but for asymptomatic patients, resection still remains controversial. Some authors have suggested that delayed surgery in asymptomatic CPAM patients is associated with fewer complications.[4] There is

a consensus suggesting that asymptomatic lesions should be removed before the first year of life, although the optimal timing of the surgery remains unknown.[11] According to the comprehensive study including the infants undergoing elective resection of CPAM from the American College of Surgeons National Surgical Quality Improvement Program (ACS NSQIP) pediatric database, the morbidity composite was significantly more common among infants younger than three months of age compared to infants older than three months.[3] In our study, more complications were seen in symptomatic patients younger than three months.

The risk of infection for congenital lung lesions has been estimated to range between 10 and 30% within the first year of life.[12] In several studies, symptomatic patients experienced significantly more intra- and postoperative complications and longer hospitalizations compared to infants who were asymptomatic at the time of operation.[12,13] Rothenberg et al.[14] suggested that there was less pulmonary tissue inflammation, if the operation was performed before six months of life. Early resection may be technically easier than late resection due to less inflammation associated with recurrent infections and would reduce the risk of complications.[6] The optimal timing of elective resection remains controversial. However, according to several studies, age is an important factor and removal is recommended before symptoms occur.[2,3,11,15]

The main limitations to this study are its single-center, retrospective design and a relatively small sample size. In the literature, studies on complications and studies that differentiate symptomatic and asymptomatic patients are heterogeneous. Therefore, further multi-center, large-scale, prospective studies are needed.

In conclusion, our study results suggest that the presence of symptomatic CPAM and age younger than three months may increase the risk of complications related to CPAM surgery. Taken together, we recommend that surgery in asymptomatic CPAM should be performed before symptoms occur.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

Author Contributions: Idea/concept, design, data collection and/or processing, references, and fundings, materials, literature review, writing the article: A.P.; Control/supervision, critical review: A.N.G.; analysis and/or interpretation: A.P., A.N.G.

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