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Evaluation of Children with Congenital Lung Malformations Who Were Diagnosed in The Prenatal and Postnatal Period

Prenatal ve Postnatal Dönemde Tanı Alan Konjenital Akciğer Malformasyonu Olan Çocukların Değerlendirilmesi

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ABSTRACT

Objective: We aimed to compare clinical features of children with congenital lung malformations (CLM) who were diagnosed in prenatal and postnatal period.

Material and Methods: Children with CLM followed in our pediatric pulmonology department between 2007-2021 were evaluated in terms of sex, age, complaints at presentation, time of onset of symptoms, age at diagnosis, diagnostic methods, gestational ages, birth weights, parental consanguinity, presence of any operations, age and indications of operations and long-term complications. Children who were diagnosed in prenatal period and those in postnatal period were compared in terms of their clinical features.

Results: The mean age of 37 children with CLM was 6.7±5.8 years, and seventeen (45.9%) of the children were girls. Children who were diagnosed during the prenatal period (n:18) had no complaints, whereas cough and recurrent pneumonia were the most common reasons at admission in others. Median age at diagnosis of children who were postnatally diagnosed (n:19) was 30 days (10-1080). Eighteen (48.6%) children were diagnosed by prenatal ultrasonography, 14 (37.8%) by computed tomography, and five (13.6%) by chest x-ray. During follow-up, malformations of two children regressed spontaneously. Twelve children were operated while others were followed up with their anomalies. Asymptomatic follow-up duration of children who were prenatally diagnosed was significantly different than the children who were diagnosed in the postnatal period (36.5±4.7 vs 24.0±12.7 months) (p:0.004).

Conclusion: Children with CLM who were diagnosed in the prenatal period were found to remain asymptomatic for longer. Prenatal diagnosis enables them to live longer without symptoms with appropriate surgical timing.

Key Words: Child, Respiratory System Abnormalities, Prenatal diagnosis

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ÖZ

Amaç: Çalışmamızda prenatal ve postnatal dönemde tanı alan konjenital akciğer malformasyonu olan hastaların klinik özelliklerini karşılaştırmayı amaçladık.

Gereç ve Yöntemler: 2007-2021 yılları arasında Çocuk Göğüs Hastalıkları Bölümü'nde izlenen konjenital akciğer malformasyonu olan hastalar cinsiyetleri, yaşları, başvuru yakınmaları, ilk semptom zamanı, tanı yaşları, tanı yöntemleri, doğum haftaları, doğum ağırlıkları, ebeveyn akrabalığı, operasyon varlığı, operasyon yaşı ve endikasyonları, uzun dönem komplikasyonları açısından değerlendirildi. Prenatal dönemde tanı alan hastalar ile postnatal dönemde tanı alanlar klinik özellikleri açısından karşılaştırıldı.

Bulgular: Konjenital akciğer malformasyonu nedeniyle izlenen 37 hastanın ortalama yaşları 6.7±5.8 yıldı ve 17'si (%45.9) kızdı. Prenatal dönemde tanı alan hastalar (n:18) yakınması olmadan başvururken diğer hastaların en sık başvuru nedenleri öksürük ve tekrarlayan akciğer enfeksiyonuydu. Postnatal dönemde tanı alan hastaların (n:19) ortanca tanı yaşları 30 (en küçük:10; en büyük:1080) gündü. Hastaların 18'i (%48.6) prenatal ultrasonografi, 14'ü (%37.8) bilgisayarlı tomografi, beşi (%13.6) akciğer grafisi ile tanı aldı. İzlemde iki hastanın malformasyonu kendiliğinden geriledi. Oniki hasta opere olurken diğer hastalar mevcut anomalileri ile takip edilmektedir. Prenatal dönemde tanı alan hastaların asemptomatik izlem süresi, postnatal tanı alan hastalardan istatistiksel olarak anlamlı farklıydı (36.5±4.7 ve 24.0±12.7 ay) (p:0.004).

Sonuç: Çalışmamızda prenatal dönemde tanı alan konjenital akciğer malformasyonu olan hastaların daha uzun süre semptomsuz seyrettiği görülmüştür. Prenatal tanı uygun cerrahi zamanlama ile daha uzun süre semptomsuz yaşamalarını sağlamaktadır.

Anahtar Sözcükler: Çocuk, Konjenital akciğer malformasyonu, Prenatal tanı

INTRODUCTION

Congenital malformations of the lung constitute a heterogeneous, rare disease group that include numerous differences in parenchyma, airway, arterial and venous structures resulting from abnormalities during embryological development, and also have many common features (1). Congenital lung malformations are divided into four main groups as congenital pulmonary airway malformation (CPAM), congenital lobar emphysema (CLE), pulmonary sequestration (PS) and bronchogenic cyst, and the prevalence is estimated to be 4 in 10,000 live births. (2).

Many cases can be diagnosed with congenital lung malformations in the intrauterine period by prenatal ultrasonography. Congenital lung malformations can be diagnosed with respiratory distress in the postnatal period or can be detected incidentally in asymptomatic children (3). Computed tomography (CT), which is the gold standard diagnostic method in the postnatal period, is recommended for patients before the operation (4-6). The treatment of children with symptomatic congenital lung malformations is surgical resection. Management of asymptomatic children is controversial. Uncertainty remains about the elective surgical resection of lesions diagnosed in the prenatal period to prevent complications (mainly infection and malignancy), as well as the timing of surgery, and whether resection is necessary for lesions that regress during pregnancy (7).

Accurate diagnosis of congenital lung malformations in the prenatal period enables the prediction of complications such as respiratory distress, recurrent pneumonia, mass effect in the mediastinum, pneumothorax, high-output heart failure and malignancy, and a safe surgical planning (8). In this study, we aimed to evaluate of children with congenital lung malformation, detect of long-term complications and compare the clinical features of patients with congenital lung malformations who were diagnosed in the pre and postnatal period.

MATERIAL and METHODS

All children followed up with congenital lung malformations in the pediatric pulmonology department between 2007 and 2021 were reviewed. The study was conducted in accordance with the principles of the Declaration of Helsinki 2008 and with the approval of Gazi University ethics committee (08.06.2020-372). This study was a retrospective descriptive study. Since the study was conducted retrospectively, patient consent was not obtained.

Children's diagnosis, sex, gestational age, birth weight, place of birth, parental consanguinity, complaints at admission, age at diagnosis, diagnosis in the pre/postnatal period, diagnostic methods, presence of operation, age and indications of operation, duration of hospitalization after operation, accompanying diseases, follow-up duration, asymptomatic follow-up duration were recorded. Preoperative weight, height, body mass index (BMI), z-scores of weight, height and BMI, post-operative and long-term complications of the operated children were also recorded. Children's current age, weight, height, BMI, z-scores of weight, height and BMI of all patients at the last control, number of pneumonia, chest x-ray findings, the percentages of FEV_1 (forced expiratory volume in the 1st second), FVC (forced vital capacity), FEV_1/FVC and FEF_{25-75} (25-75% of forced expiratory flow) in pulmonary function tests (PFT) of patients who complied were recorded. The PFTs were performed according to the American Thoracic Society-European Respiratory Society ATS-ERS guidelines (9). Growth was assessed by weight and height z-scores in children under two years of age, and BMI z-scores in children over two years of age, as recommended by the World Health Organization. A z-score of <-2 Standard Deviation (SD) was considered as growth retardation (10). Pneumonia was defined as an acute respiratory infection affecting the alveoli of the lungs and the distal bronchial tree (11). Patients who had two pneumonia attacks within a year or had at least three pneumonia attacks during their lifetime and were clinically and radiographically normal between attacks were considered to have recurrent pneumonia (12). The lateral curvature of the spine over 10 degrees

to the right or left, which was detected radiologically in the coronal plane, was evaluated as scoliosis (13). Partial or complete fusion of the anterior or posterior ribs was considered to be rib fusion (14). Pectus excavatum was defined as a congenital chest wall deformity in which several ribs and sternum enlarge abnormally, creating a concave or collapsed appearance in the anterior chest wall (15). Children who were diagnosed in the pre and postnatal period were compared in terms of their clinical characteristics.

IBM SPSS Statistics version 22.0 (IBM, Armonk, NY, USA) was used for the statistical analyses. In the statistical analysis of all data obtained, descriptive data were presented as frequency, percentage, mean, ± standard deviation, median, minimum and maximum. The conformity to normal distribution of numerical data was tested with the Shapiro-Wilk test. In the analysis of continuous variables, t-test was used for independent samples when parametric assumptions were met, Mann-Whitney U test was used when they were not met, chi-square test was used in the analysis of nominal variables, and Fisher exact test was used when the distribution was not suitable for chi-square in the analysis of nominal variables. $p<0.05$ was considered significant for all tests.

RESULTS

Over a thirteen-year period, of the 37 children with congenital lung malformations followed up in the pediatric pulmonology department, 16 (43.2%) had PS, eight (21.6%) had CPAM, seven (18.9%) had CLE, and four (4%) 10.8% had bronchogenic cysts and two (5.5%) had coexistence of CPAM and PS. Seventeen children (45.9%) were female. The mean birth weight of the children was 3056±513 grams and 33 (89.1%) were born at term. Eight children (21.6%) had parental consanguinity. Four children had congenital heart disease, one had asthma and one had lignous conjunctivitis. The clinical features of children with congenital lung malformations are given in Table I. Eighteen (48.6%) children were diagnosed prenatally and 19 (51.4%) in the postnatal period. There was no significant difference between the children who were diagnosed in the pre and postnatal period in terms of sex, gestational age, birth weight, and parental consanguinity ($p>0.050$). The comparison of the children who were diagnosed in the pre and postnatal period is given in Table II.

All of the patients who were diagnosed prenatally were born in a tertiary center and evaluated by the pediatric pulmonology department during the neonatal period. Of the patients diagnosed in the postnatal period, 14 (73.6%) were born in a tertiary center. One (5.5%) of the children who were diagnosed in the prenatal period was premature, one (5.5%) had neonatal pneumonia, one (5.5%) was followed in the neonatal intensive care unit (NICU) for clinical follow-up and none of them received respiratory support. Of the children who were postnatally diagnosed, five (26.3%) were hospitalized for neonatal pneumonia, three (15.7%) for transient tachypnea of the newborn, and one (5.2%) for congenital heart disease. There was no statistically significant difference in terms of hospitalization in the NICU of the children who were diagnosed in the pre and postnatal period (p:0.078). One child received nasal continuous positive airway pressure therapy, and two children received incubator oxygen support. While the children who were diagnosed in the prenatal period had no symptoms at the time of admission, the most common reasons for admission were cough and recurrent pneumonia in the children who were postnatally diagnosed. Eighteen of the children (48.6%) had a history of pneumonia.

Eighteen (48.6%) children were diagnosed by prenatal ultrasonography, 14 (37.8%) by CT, and five (13.6%) by chest x-ray. Prenatal ultrasonography were normal in 14 (73.6%) children who were diagnosed in the postnatal period. Malformations were unilateral in all children. Thirteen (35.1%) children's malformations were in the upper lobe of the right lung, eight (21.6%) were in the lower lobe of the left lung, six (16.2%) were in the upper lobe of the left lung, six (16.2%) were in the lower lobe of the right lung, and four (10.9%) were in the middle lobe of the right lung. Right lower lobe was the most common (60.0%) involvement in CPAM, left upper lobe in CLE (85.7%), and left lower lobe in PS (44.4%). Bronchogenic cysts were located in the right middle lobe in all

Table I: Clinical features of children with congenital lung malformations (n:37).

	n (%)
Congenital lung malformations*	
PS	16 (43.2)
CPAM	8 (21.6)
CLE	7 (18.9)
Bronchogenic cyst	4 (10.8)
CPAM+PS	2 (5.5)
Sex*	
Female	17 (45.9)
Male	20 (54.1)
Current age of children (years)†	6.7±5.8
Age at diagnosis of children who were diagnosed in the postnatal period (days) (median, min-max)†	30 (10-1080)
Duration of follow-up (months)†	48.8±26.4
Parental consanguinity*	8 (21.6)
Born at term*	33 (89.1)
Diagnostic methods*	
Prenatal ultrasonography	18 (48.6)
CT	14 (37.8)
Chest x-ray	5 (13.6)
Age at operation (months) (n:12)†	16.3±14.2
Long term complications (n:11)*	
Recurrent pneumonia	4 (36.4)
Rib fusion	4 (36.4)
Scoliosis	2 (18.1)
Pectus excavatum	1 (9.1)

:** *n(%),* **†:** *mean*±SD, ***CPAM: *Congenital Pulmonary Airway Malformation,* ***PS:*** *Pulmonary Sequestration,* ***CLE:*** *Congenital Lobar Emphysema,* ***CT:*** *Computed Tomography,* ***SD:*** *Standard Deviation*

Table II: Comparison of the children with congenital lung malformations who were diagnosed in the prenatal and postnatal period.

	Prenatal (n:18) n (%)	Postnatal (n:19) n (%)	p
Congenital lung malformations			
PS	8 (44.4)	8 (42.1)	
CPAM	7 (38.8)	1 (5.2)	
CLE	0 (0)	7 (36.8)	
Bronchogenic cyst	2 (11.1)	2 (10.5)	
CPAM+PS	1 (5.7)	1 (5.4)	
Sex			
Female	8 (44.4)	9 (47.3)	0.858[a]
Male	10 (55.6)	10 (52.7)	
Parental consanguinity	3 (16.6)	5 (26.3)	0.476[b]
Born at term	17 (94.4)	16 (84.2)	0.217[a]
History of hospitalization in NICU	3 (16.6)	9 (47.3)	0.078[b]
Birth weight (gram) (mean±SD)	2800±456	2960±329	0.227[d]
Current ages of children (years) (mean±SD)	5.6±4.2	7.3±5.6	0.305[d]
Children below 2 years old (n:23)			
Weight z-score (median, min-max)	+1.0 (- 0.8 - + 1.8)	+0.8 (-2.4 - +1.6)	0.097[c]
Height z-score (median, min-max)	+1.4 (- 0.6 - + 2.3)	+0. 9 (-2.6- +2.2)	0.141[c]
Children above 2 years old (n:14)			
BMI z-score (median, min-max)	+ 0.6 (- 0.8 - + 2.8)	+ 0.9 (-2. - +1.8)	0.089[c]
Duration of follow-up (months) (mean±SD)	47.2±20.4	50.4±32.4	0.723[d]
Duration of asymptomatic follow-up (months) (mean±SD)	36.5±4.7	24.0±12.7	0.004[d*]
Age at operation (months) (mean±SD)	17.1±9.1	12.6±10.2	0.166[d]
Post-operative duration of hospitalization (days) (mean±SD)	6.6±5.4	7.9±6.8	0.525[d]
Long term complications	(n:4)	(n:7)	
Recurrent pneumonia	1 (25.0)	3 (43.0)	
Rib fusion	2 (50.0)	2 (28.5)	
Scoliosis	0 (0)	2 (28.5)	
Pectus excavatum	1 (25.0)	0 (0)	

CPAM: *Congenital Pulmonary Airway Malformation,* ***PS:*** *Pulmonary Sequestration,* ***CLE:*** *Congenital Lobar Emphysema,* ***CT:*** *Computed Tomography,* ***NICU:*** *Newborn Intensive Care Unit,* ***BMI:*** *Body Mass Index,* ***SD:*** *Standard Deviation,* [a] *Chi-square test,* [b] *Fisher exact test,* [c] *Mann Whitney U test,* [d] *Independent samples t test,* ******* *Statistically significant*

patients. The median age at diagnosis of those diagnosed in the postnatal period was 30 days (10-1080 days).

The mean FEV_1 was 65.8±19.0%, FVC was 64.8±17.1%, FEV_1/FVC was 95.2±10.8, FEF_{25-75} was 54.5±28.4% of 5 (13.5%) children who were able to perform PFT. All of these children were operated and two had congenital heart disease, one had asthma.

In the chest x-rays of the children, 43.2% had cyst formation, 27% air trapping, 24.3% interstitial thickening, 21.6% mediastinal shift, 13.5% infiltration and 10.8% atelectasis. Thirty-five patients had thorax CT. In the thorax CT, cysts (100%), mediastinal shift (20%), consolidation (50.0%) were detected in children with CPAM, solid lesion unrelated to the tracheobronchial tree (100%), consolidation (37.5%) and atelectasis (37.5%) in children with PS, air trapping (100%), consolidation (42.9%), mediastinal shift (28.6%) in children with CLE, cyst (100%), and consolidation (50%) in children with bronchogenic cysts.

Twelve children (32.4%) were operated. The mean operation age of the children was 16.3±14.2 months. Malformations (CPAM and bronchogenic cyst) of one child (5.4%) who were diagnosed in the prenatal period regressed spontaneously. Four with CLE were operated, two with CPAM, five with PS, and one with bronchogenic cyst. One child with CPAM, one with PS, and one with bronchogenic cyst who were diagnosed in the prenatal period were operated for recurrent pneumonia, while one with CPAM and one with PS were operated under elective conditions. Four children with CLE and three children with PS who were diagnosed in the postnatal period were operated. While these children were operated for recurrent pneumonia, growth retardation developed in three of them during the follow-up. The pathology of a child who was diagnosed as a bronchogenic cyst in the prenatal period was compatible with CPAM. Four children had elective operation plan, but they were postponed due to the concerns of the families about the coronavirus disease 2019 (COVID-19). Other children are followed up with their existing anomalies asymptomatically.

The mean operation age of the patients who were prenatally diagnosed was 17.1±9.1 months, and was 12.6±10.2 months for the children who were diagnosed in the postnatal period (p:0.166). Chylothorax was observed in one child who was operated for PS, while no early complications were observed in other patients. The

mean duration of hospitalization after the operation for the children who were diagnosed in the prenatal period was 6.6±5.4 days, and 7.9±6.8 days for the children who were postnatally diagnosed (p:0.525).

The mean duration of follow-up of the children was 48.8±26.4 months. In the long-term follow-up of the operated children, recurrent pneumonia was observed in four (36.3%), rib fusion in four (36.3%), scoliosis in two (18.1%), and pectus excavatum in one (9%).

The mean duration of follow-up of the children who were prenatally diagnosed was 47.2±20.4 months, whereas it was 50.4±32.4 months for the children who were diagnosed in the postnatal period (p:0.723). In the follow-up, 18 (48.6%) children had pneumonia, 11 (29.7%) children had only cough. Eight (21.7%) children were asymptomatic. The mean asymptomatic duration of follow-up of the children who were diagnosed in the prenatal period was significantly longer (36.5±4.7 months), than the children who were diagnosed postnatally (24.0±12.7 months) (p:0.004). The mean age of the children at the last control who were prenatally diagnosed was 5.6±4.2 years, and it was 7.3±5.6 years for the children who were diagnosed in the postnatal period (p:0.305). Twenty-three (62.1%) children were aged under two. The median weight z-score of these children was +0.9 (min:-2.4; max:+1.8), and the height z-score was +1.2 (min:-2.6; max:+2.3). The median BMI z-score of children aged over two was +0.7 (min:-2.2; max: +2.8). There was no significant difference between the children diagnosed in the pre and postnatal period in terms of current age, weight, height and BMI z-scores (p>0.050).

DISCUSSION

In our study, all children who were prenatally diagnosed were evaluated in the neonatal period before the symptoms started. The most common reasons for admission in the children who were diagnosed in the postnatal period were cough and recurrent pneumonia. The asymptomatic duration of follow-up of the children who were diagnosed in the prenatal period was longer than the patients who were postnatally diagnosed.

Lesions in CPAM are mostly unilateral and do not dominate the lobes, but they are rarely seen in the middle lobe (16). The left upper lobe is most commonly involved in CLE. Multiple lobe involvement is also rare (17). In PS, involvement is mostly found in the left lower lobe, and in bronchogenic cysts, involvement is found in the lower lobes without any side difference (18,19). In our study, similar to the literature, mostly CPAMs were unilateral, CLEs were in the left upper lobe, and PSs were in the left lower lobe. However, different from the literature, bronchogenic cysts were located in the right middle lobe.

In a study conducted in Japan, approximately 65% of 428 patients with congenital lung malformations were symptomatic before the age of three (20). In a meta-analysis, 505 patients who were diagnosed in the prenatal period and followed up with CPAM and PS were evaluated of whom 16 (3.2%) became symptomatic at a median age of 7 months (21). It has been shown that 13% of 154 patients with congenital lung malformations who were diagnosed in the prenatal period have symptoms at a median age of 2 years, and all patients have symptoms at the age of 6 years (22). In our study, children who were diagnosed in the prenatal period became symptomatic approximately at three years of age, whereas it was at two years of age in those diagnosed postnatally. Prenatal diagnosis of children with CPAM allows elective resection in the asymptomatic period, resulting in shorter hospital stay, lower major complications and less medical cost. Early diagnosis allows monitoring in a tertiary center for prenatal counseling, possible fetal intervention, birth planning, experienced NICU and surgery (23). In our study, the delivery of children who were diagnosed in the prenatal period in appropriate centers, as well as the fact that families were well informed about the disease itself and protective measures from the infections may have caused the children to be symptomatic later.

The optimal timing for surgery of patients with asymptomatic congenital lung malformations has not been established. The main rationale for the advocates of the observation strategy is that surgery can be totally avoided for some patients (24). In a study evaluating 61 patients with congenital lung malformations, 48% of the patients had a median operation age at 108 (interquartile range: 8-828) days, while 52% were followed without surgery. It has been reported that 62% of the patients who were diagnosed in the prenatal period were followed up without surgery (25). While less than 1/3 of the children in our study required surgery, 72.2% of the children who were diagnosed in the prenatal period are managed with an observation strategy. Some congenital lung malformations can regress spontaneously in the prenatal or postnatal period. In a study, the incidence of spontaneous regression of congenital lung malformations detected in the prenatal period was 14% (2). In our study, this rate was approximately 5%. Fetal hydrops and pleural effusion may develop in the prenatal period in patients with congenital lung malformations (26). Due to the differences in the approach to congenital lung malformations, possible spontaneous regression and intrauterine complications, it is important that the malformations are detected in the prenatal period, the families are informed about the process in detail, and the surgery or observation decision is determined together with families.

One of the reasons for those who advocate early resection in patients with asymptomatic congenital lung malformations is that it has a positive effect on compensatory lung growth. It has been reported that resection performed in patients younger than 4 years of age is associated with improvement in the lung functions during follow-up (27). On the contrary, Keijzer et al. (28) evaluated patients who were operated before or after the age of 2 and showed that there was no significant relationship between the age of resection and FVC and FEV_1 values at mean age of 10 years. Naito et al. (29) evaluated the pulmonary function and exercise tests of patients who were operated before or after the age of 2 years in their prospective study, and found that the age of resection was not associated with

any abnormal respiratory function or exercise test parameters. They reported that although their total lung capacity was preserved at the ages of 8 and 23, their FEV_1 was decreased. In our study, the mean age at operation was approximately 16 months. Five children who were able to perform PFT were operated patients and two had congenital heart disease and one had asthma. The low average lung function of these children may be related to their comorbidities.

Prenatal diagnosis of congenital lung malformations is difficult due to overlapping findings between different lesions or the presence of complex, hybrid lesions with combined vascular and bronchopulmonary abnormalities (6). In our study, prenatal ultrasonography was normal in approximately 75% of the children who were diagnosed in the postnatal period. The pathology of the operation material was compatible with CPAM in one of the children who was thought to have a bronchogenic cyst on prenatal ultrasonography. Although the diagnosis of congenital lung malformations with prenatal ultrasonography is difficult, the correct diagnosis warrants to better follow-up of the patients.

The prenatal ultrasonographic features of CLE are not well characterized. Patients with CLE may be diagnosed less frequently in the prenatal period than patients with other congenital lung malformations. It has been reported that 73.1% of patients with congenital lung malformations and 24.5% of patients with CLE were detected by prenatal ultrasonography. CLE may be misdiagnosed as CPAM or PS in the prenatal period (30, 31). CLE detected in the prenatal period may disappear and become evident again in the postnatal period (32). In our study, similarly, children with CLE could not be diagnosed in the prenatal period.

In a study in which 61 children with congenital lung malformations were evaluated, it has been shown that 4% of the patients at the age of 1, 8% at two, 12% at five, 5% at 8 had pneumonia of whom 23% had an attack at least once. Due to recurrent pneumonia, 6% of the patients were operated (25). In our study, approximately 85% of the operated children had recurrent pneumonia and this was the reason for the operation. Approximately 30% of the children who were not operated had a history of pneumonia, but they did not have any complaints in the follow-up and their growth was normal. In the long-term follow-up of 119 patients with congenital lung malformations, the risk of recurrent pneumonia was below 10% and decreased after the second year (33). Close follow-up of children with congenital lung malformations, especially those with pneumonia, and monitoring their growth could lead to operation decision of the children.

In patients who have been operated for congenital lung malformations, cutting of the latissimus dorsi and/or serratus anterior muscle during posterolateral thoracotomy may result in muscular atrophy in the postoperative period and may lead to the development of chest wall and/or spinal deformities. Chest wall deformity was detected in 7% of the operated children 5 years after the operation (34). In a study evaluating 74 patients who underwent surgery, scoliosis and chest wall deformity were found in 37% of the patients. Of them, one patient was operated for scoliosis, while one patient was treated using a corset (35). In our study, in the long-term follow-up of the operated children, rib fusion were observed approximately in 33%, scoliosis in 16%, and pectus excavatum in 8% of the patients. Close follow-up of patients who have been operated for congenital lung malformations in terms of musculoskeletal complications that may occur during follow-up enables noninvasive treatment approaches to be applied.

One of the limitation of our study is its retrospective nature. In addition, because of our center is a tertiary care center, symptomatic patients who were diagnosed in the postnatal period may have been referred to us.

Prenatal diagnosis of congenital lung malformations provides an opportunity to predict the complications that may be caused by the malformation, to monitor the growth and to plan the surgery at the most appropriate time for the patient. It is possible to monitor patients who were diagnosed in the prenatal period without symptoms for a longer period of time. Due to very different clinical features and different treatment approaches, it is important to diagnose children with congenital lung malformations during the prenatal period and decide on the timing of surgery together with the families, and to follow up the operated patients in terms of musculoskeletal complications.

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