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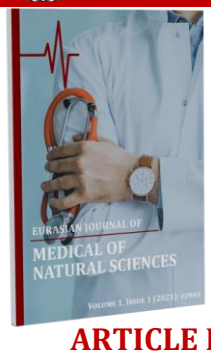

TREATMENT OUTCOMES OF CYSTIC FORMS OF BRONCHOPULMONARY MALFORMATIONS IN CHILDREN

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ABSTRACT

This study, conducted at Tashkent Pediatric Medical Institute, analyzes treatment outcomes for cystic forms of bronchopulmonary malformations (CFBPM) in 106 children from 2006 to 2022. Surgical correction, the primary treatment, was performed in 86.8% of cases, with outcomes assessed as good (56.5%), satisfactory (32.6%), or unsatisfactory (10.9%). Two groups were compared: a comparison group (2006–2017) using conventional methods and a main group (2018–2022) with advanced diagnostics and thoracoscopic techniques. The main group showed improved outcomes, reducing unsatisfactory results from 20.5% to 9%, attributed to refined surgical and perioperative strategies. Watchful waiting was applied in asymptomatic cases, though optimal surgical timing remains debated, balancing early intervention with lung development.

Introduction. According to the World Health Organization (WHO), congenital malformations, such as cardiac defects and lung pathologies, constitute a significant proportion of mortality causes among newborns and children under 5 years of age [18]. Surgical correction of cystic forms of bronchopulmonary malformations (CFBPM) remains the only radical treatment method. The approach of pediatric surgeons in managing various types of lung and mediastinal anomalies is determined by the extent of the lesion and the clinical presentation of the disease.

To date, there is no consensus regarding the optimal age for performing surgical treatment in children with congenital lung and mediastinal anomalies. This issue is extensively discussed in both domestic and international medical literature. The majority of these malformations manifest late in the postnatal period, frequently as respiratory distress, which develops in the context of complications. These complications are typically associated with mass effect on mediastinal organs and the superimposition of an infectious process.

CFBPM is characterized by a heterogeneous clinical course, ranging from regression of paucisymptomatic forms to progression with the development of an overt clinical picture and worsening of symptoms. Asymptomatic cysts pose a challenge in terms of selecting treatment strategy: surgical intervention versus conservative observation. A watchful waiting approach is warranted in cases of paucisymptomatic forms [1, 4, 8, 9, 15]. It is crucial to consider that

lung tissue in children continues to grow through alveolar formation until the age of 5-8 years, making early surgical intervention potentially beneficial for complete restoration of lung volume and function [1, 5, 10, 16].

The aim of this study was to analyze the immediate and long-term outcomes of treatment for cystic forms of bronchopulmonary malformations in children.

Materials and Methods. This work is based on the analysis of the diagnosis and treatment outcomes of 106 children with CFBPM, who were observed at the clinical facilities of the Department of Hospital Pediatric Surgery, Tashkent Pediatric Medical Institute (City Clinical Children's Surgical Hospital No. 2 of Tashkent, Department of Neonatal Surgery of the Republican Perinatal Center of the Ministry of Health of the Republic of Uzbekistan, Department of Minimally Invasive Pediatric Surgery of the National Children's Medical Center) between 2006 and 2022. The age of patients ranged from 1 day to 16 years, and the distribution by age category was as follows: newborns – 31 (29.2%), 1 month to 1 year – 25 (23.6%), 1 year to 3 years – 19 (17.9%), 3 to 7 years – 13 (12.3%), 7 to 15 years – 13 (12.3%), and 15 to 16 years – 5 (4.7%).

The distribution of pathology varied depending on the age of the children; a decrease in the number of cases and a change in the structure of nosological forms were observed with increasing age. Among the 106 patients, 66 were boys (62.3%) and 40 were girls (37.7%).

The distribution of patients by nosological forms of CFBPM is presented in Table 1.

Patients were divided into two groups: 52 children (49.1%) who underwent treatment at the clinic between 2006 and 2017 using conventional diagnostic and surgical approaches (comparison group), and 54 children (50.9%) who were treated at the clinics between 2018 and 2022, where an expanded panel of diagnostic methods was employed. This panel included prenatal screening of pregnant women and fetuses, as well as video-assisted thoracoscopic interventions (main group).

Table 1

Nosological forms and number of patients with CFBPM in the study groups (n=106)

Nosological gorms of pathologies	Main group		Comparison group	
	abs.	%	abs.	%
Congenital cystic adenomatoid malformation (CCAM)	26	24,6	20	18,8
Bronchial cysts (BC)	8	7,5	14	13,3
Bronchogenic cysts (BGC)	7	6,6	9	8,5
Congenital lobar emphysema (CLE)	10	9,4	9	8,5
Bronchopulmonary sequestration (BPS)	3	2,8	-	-
Total:	**54**	**50,9**	**52**	**49,1**

Results and Discussion. The presence of pronounced clinical signs of CFBPM, confirmed by auxiliary diagnostic methods during the neonatal period, and their

manifestation in the context of aggravating factors, indicated a severe disease course requiring surgical intervention. Surgical treatment was performed in 92 (86.8%) patients on an emergency, urgent, or elective basis. A total of 94 operations were performed, including two repeat procedures in patients with congenital congenital cystic adenomatoid malformation (CCAM) (Table 2).

In cases of asymptomatic or compensated disease, a watchful waiting approach was adopted for 5 patients (4.7%), which involved serial functional studies and preventive measures to mitigate potential complications. In 9 cases (8.5%), watchful waiting for paucisymptomatic forms was attributed to the parents' temporary refusal of surgical treatment.

Table 2

Choice of treatment strategy for CFBPM in children (n=106)

Form of pathology	Surgical treatment (n=92)			Wait and see tactics (n=14)	
	urgently	urgently planned	in a planned manner	stably compensated state	based on parents' motives
CCAM (n=46)					
– Type I (n=32)	5	4	19	2	2
– Type II (n=13)	-	2	6	3	2
– Type III (n=1)	-	-	-	-	1
CLE (n=19)					
– compensated (n=1)	-	-	1	-	-
– subcompensated (n=7)	1	3	3	-	-
– decompensated (n=11)	8	1	2	-	-
BGC (n=16)	2	7	5	-	2
BC (n=22)	3	4	14		1
BPS (n=3)					
– intralobar form (n=2)	1	-	1	-	-
– extralobar form (n=1)	-	-	-	-	1
Total (n=106)	**20**	**21**	**51**	**5**	**9**

The optimal timing for surgical correction of CFBPM remains a topic of debate in the medical literature. Many authors advocate for surgical intervention regardless of the severity of clinical manifestations to prevent potential complications. In our experience, the indications and timing of surgery were determined individually for each patient, considering the severity and dynamics of clinical symptoms, as well as the specific anatomical changes within the bronchopulmonary structures.

In the majority of patients (72; 76.6%), the postoperative period was uneventful. However, 22 (23.4%) patients experienced various complications, categorized as general surgical, specific, somatic, and mixed complications (a combination of multiple complication types) (Table 3).

General surgical complications were observed in 2 (2.2%) cases. Specifically, wound dehiscence in the early postoperative period was noted in 1 (1.1%) patient in the comparison group, and a suture sinus in 1 (1.2%) patient in the main group.

Table 3

Nature of postoperative complications in the study groups (n=22)

Nosological gorms	Types of complications							
	General surgical		Specific		Somatic		Mixed	
	Abs.	%	Abs.	%	Abs.	%	Abs.	%
CCAM (n=7)	-	-	4	18,2	2	9,1	1	4,5
CLE (n=3)	-	-	1	4,5	2	9,1	-	-
BGC (n=3)	1	4,5	1	4,5	1	4,5	-	-
BC (n=8)	1	4,5	4	18,2	3	13,6	-	-
BPS (n=1)	-		-	-	1	4,5	-	-
Total (n=22)	**2**	**9**	**10**	**45,5**	**9**	**41**	**1**	**4,5**

General surgical complications were observed in 2 (2.2%) cases. Specifically, wound dehiscence in the early postoperative period was noted in 1 (1.1%) patient in the comparison group, and a suture sinus in 1 (1.2%) patient in the main group.

Specific complications were identified in 10 (10.6%) patients, including pneumothorax (5 cases), persistent air leak (2 cases), and lobar atelectasis (3 cases). In the comparison group, specific complications were recorded in 7 (7.4%) patients, while in the main group, they were recorded in 3 (3.2%) patients.

Somatic complications were recorded in 9 patients (9.6%), of which 3 (33.3%) were in the main group and 6 (66.7%) in the comparison group. Somatic complications manifested as pneumonia (8 cases) and multiple organ failure (1 case). These complications more frequently developed in newborns from mothers with an unfavorable obstetric and gynecological history, as well as due to perinatal and intranatal factors leading to fetal hypoxia. In patients who underwent emergency surgery in the early neonatal period, somatic complications were noted in 4 cases (44.4%). In children up to 3 months of age, somatic complications were recorded in 3 cases (3.2%), and in children up to 3 years old, in 2 cases (2.1%).

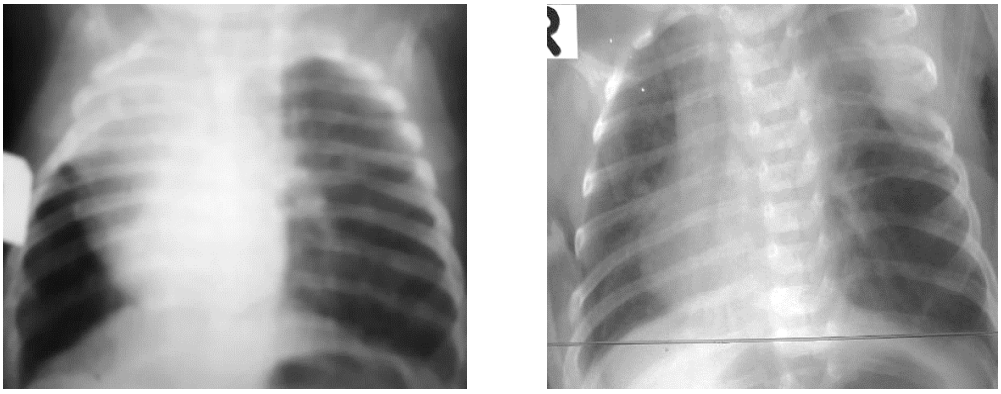
Among the complications identified during treatment were: insufficient lung expansion (aerostasis); atelectasis of the remaining lung lobe; and multiple organ failure. In one case (1.1%) in the main group, a complication associated with COVID-19 pneumonia, complicated by multiple organ failure, was observed, leading to a fatal outcome. This case exemplifies a severe disease course exacerbated by comorbidities. Following resection of the lingular segments of the left lung for CCAM in a child aged 1.5 months, recurrent cystic changes in other areas of the lung, complicated by intrathoracic tension syndrome, were detected within three months. This prompted a repeat surgery – pneumonectomy on the affected side. Against this background, the child contracted a COVID-19 infection, which was complicated by the

development of multiple organ failure, ultimately resulting in a fatal outcome. The fatal outcome was attributed to the severity of the malformation, the presence of comorbidities and anatomical anomalies, as well as tactical and technical errors made at various stages of treatment.

Out of 106 patients with CFBPM, 105 patients (99.1%) were discharged from the hospital, with the exception of one patient who experienced a fatal outcome. Among those discharged, 91 (86.7%) underwent surgical treatment, and 14 (13.3%) received conservative therapy.

Long-term treatment outcomes were studied in 92 (87.6%) of the 105 discharged patients within a period of 6 months to 5 years. Among them, 53 (57.6%) were patients from the main group and 39 (42.4%) from the comparison group. Follow-up examinations were conducted in 82 (89.1%) patients who had undergone surgical treatment and in 10 (10.9%) patients who were managed with a watchful waiting approach.

Patient complaints, objective examination data, and results from ancillary diagnostic methods varied in nature and severity. 11 patients (13.1%) reported complaints of cough of varying periodicity (episodic or persistent) and character (productive or non-productive), which was not clearly associated with a specific nosological form of pathology. In 8 cases (9.5%), emphysematous changes in the remaining lung areas on the operated side were detected (Fig. 1, a and b) to varying degrees, however, clinical manifestations were absent. These patients showed a predisposition to frequent colds.



a **b**

Fig. 1. a) Emphysematous changes in the remaining lower lobe after right bilobectomy for lobar emphysema, patient U.Sh., 2 months old, medical record number 67/47; b) Emphysematous changes in the remaining lower lobe after left upper lobectomy, patient G.D., 1 month old, medical record number 98/63.

During dynamic follow-up, no progression of clinico-radiological changes was observed. These emphysematous changes in the remaining areas of the lung post-surgery can be considered compensatory alterations, although the risk of latent manifestations of dysplasia cannot be ruled out. Further dynamic observation of these patients is necessary to determine the most appropriate treatment strategy.

In the long term, 3 patients (3.6%) exhibited chest wall retraction of varying degrees of severity, ranging from subtle to pronounced. The development of this condition can be attributed to the removal of a portion of the lung, the formation of atelectasis adjacent to the

resection margins, and the development of adhesions and fibrous bands leading to compression of the lung parenchyma.

The outcomes of surgical treatment were assessed using a scoring system, categorizing outcomes as good, satisfactory, and unsatisfactory.

A good outcome was recorded in 52 patients (56.5%). Characteristics of this category included: the absence of complaints and signs of respiratory failure, as well as the absence of residual effects related to the surgery; a symmetrical chest condition and a good cosmetic appearance of the postoperative scar; normal chest X-ray or MSCT findings; and pulmonary function test results within the normal range (Fig. 2).

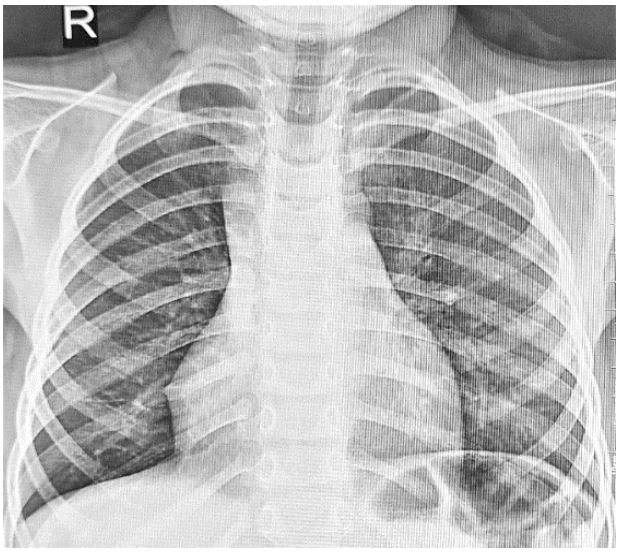


Fig. 2. Patient I.S., age: 2 years 7 months, post-lobectomy status for right CCAM, medical record number 3224-213. Plain chest radiograph – complete expansion of the remaining lung.

A satisfactory outcome was noted in 30 patients (32.6%). The main features of this category included: complaints of cough without overt respiratory distress, as well as moderate residual effects post-surgery requiring additional courses of conservative treatment; moderate chest asymmetry with retraction of certain areas and a satisfactory cosmetic appearance of the postoperative scar; partial lung expansion or limited atelectasis on the operated side according to chest X-ray and MSCT; and pulmonary function test results reduced by 15-30% compared to normal (Fig. 3).

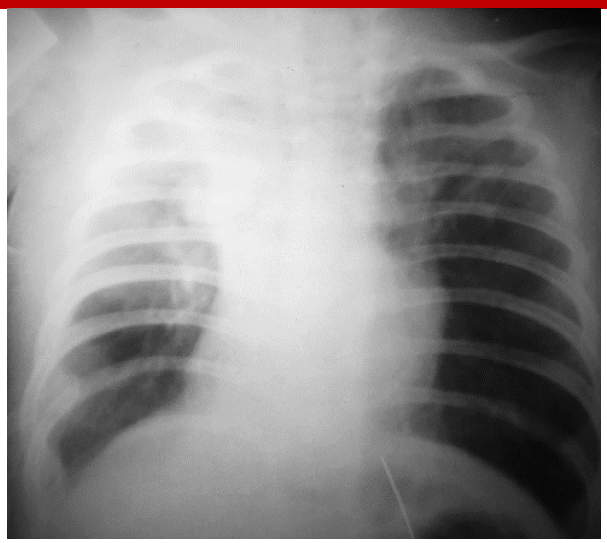


Fig. 3. Patient F.Sh., 1 year 1 month, medical record number 614-52. Post-lobectomy status with partial atypical resection for right CCAM. Plain chest radiograph – partial expansion of the remaining lung.

An unsatisfactory outcome was identified in 10 patients (10.9%). Criteria included: complaints of frequent cough, frequent episodes of cold-related inflammatory diseases, manifestations of respiratory distress exacerbated by physical exertion, as well as the presence of residual effects requiring repeat surgical intervention; pronounced chest asymmetry (retraction or bulging) and an unsatisfactory or keloid condition of the postoperative scar; partial lung expansion, marked emphysema, signs of atelectasis or pleural complications on the operated side, identified on chest X-ray or MSCT; and pulmonary function test results reduced by more than 30% compared to normal (Fig. 4).

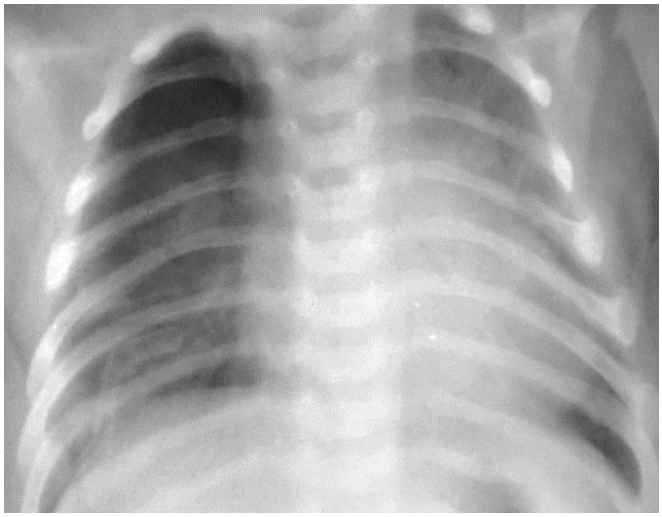


Fig. 4. Patient Z., 2 years 7 months, medical record number 6231-1189. Post-lobectomy status with partial atypical resection for right CCAM. Plain chest radiograph – marked emphysematous distension of the remaining lung.

The examination results of 14 patients (16.7%) with cystic forms of bronchopulmonary anomalies (CCAM – 10 patients, BGC – 2 patients, BC – 1 patient, BPS – 1 patient), characterized by a mildly symptomatic and stably compensated clinical course, confirmed the rationale for a watchful waiting approach. This strategy was adopted, partly due to the parents' temporary refusal of surgical treatment. Over time, this patient group maintained,

and in some instances, developed an increased susceptibility to upper respiratory infections. In 5 of these patients (35.7%), the compensated state progressed to subcompensation, which indicated surgical intervention within a period of 8 months to 2 years.

In the control group (7 out of 38 patients; 18.4%) who underwent surgery at an age older than 10 years (CCAM – 3 patients, BGC – 1 patient, BC – 3 patients) due to late diagnosis, recurrent episodes of tracheobronchial inflammatory diseases were noted in the long-term follow-up. These episodes were associated with the chronicity of the condition, which had a negative impact on the patients' general health and well-being. Based on the evaluation criteria, the long-term treatment outcomes in this group were deemed satisfactory in 4 patients (4.3%) and unsatisfactory in 3 patients (3.3%) out of the 92 examined.

Long-term treatment outcomes, categorized by the nosological form of the pathology and the surgical approach, are detailed in Tables 4 and 5.

Table 4

Long-term outcomes of CFBPM treatment in the study groups (n=92)

Type of pathology	Main group (n=53)						Comparison group (n=39)					
	Good		Satisfac-tory		Unsatisfac-tory		Good		Satisfac-tory		Unsatis-factory	
	abs	%	abs	%	abs	%	abs	%	abs	%	abs	%
CCAM (n=38)	15	28,30	8	15,09	2	3,77	5	12,82	5	12,82	3	7,69
CLE (n=19)	6	11,32	3	5,66	1	1,89	3	7,69	3	7,69	2	5,13
BC (n=20)	5	9,43	2	3,77	1	1,89	7	17,95	3	7,69	2	5,13
BGC (n=13)	4	7,55	2	3,77	1	1,89	3	7,69	2	5,13	1	2,56
BPS (n=2)	2	3,77	0	0,00	0	0,00	0	0,00	0	0,00	0	0,00
P	Pearson's chi-squared = 1,505; p = 0,993						Pearson's chi-squared = 2,747; p = 0,840					
P	Pearson's chi-squared = 3,965; p = 0,860											
Total (n=92)	**32**	**60,38**	**16**	**30,19**	**5**	**9,43**	**18**	**46,15**	**13**	**33,33**	**8**	**20,51**
P	**Chi-squared = 19,111; p = 0,000**						**Chi-squared = 7,538; p = 0,023**					

Table 5

Long-term outcomes of CFBPM treatment based on treatment strategy and surgical approach in the study groups (n=92)

Type of treatment	Main group (n=53)						Comparison group (n=39)					
	Good		Satisfac-tory		Unsatisfac-tory		Good		Satisfac-tory		Unsatisfac-tory	
	abs	%	abs	%	abs	%	abs	%	abs	%	abs	%
Watchful waiting (n=10)	1	1,89	3	5,66	2	3,77	0	0,00	2	5,13	2	5,13
Lobectomy (n=44)	18	33,96	6	11,32	1	1,89	12	30,77	4	10,26	3	7,69

Thoracoscopic lobectomy (n=2)	1	1,89	1	1,89	0	0,00	0	0,00	0	0,00	0	0,00
VATS lobectomy (n=6)	3	5,66	3	5,66	0	0,00	0	0,00	0	0,00	0	0,00
Pulmonectomy (n=2)	0	0,00	0	0,00	0	0,00	0	0,00	1	2,56	1	2,56
VATS pneumonectomy (n=1)	0	0,00	0	0,00	1	1,89	0	0,00	0	0,00	0	0,00
Cystectomy (n=15)	4	7,55	2	3,77	1	1,89	5	12,82	2	5,13	1	2,56
Thoracoscopic cystectomy (n=1)	1	1,89	0	0,00	0	0,00	0	0,00	0	0,00	0	0,00
Bilobectomy (n=3)	1	1,89	1	1,89	0	0,00	0	0,00	1	2,56	0	0,00
Lobectomy with atypical resections (n=3)	1	1,89	0	0,00	0	0,00	1	2,56	1	2,56	0	0,00
Removal of mediastinal cyst (n=4)	1	1,89	0	0,00	0	0,00	0	0,00	2	5,13	1	2,56
Removal of sequestrum (n=1)	1	1,89	0	0,00	0	0,00	0	0,00	0	0,00	0	0,00
P	Pearson's chi-squared = 22,810; p = 0,298						Pearson's chi-squared = 14,158; p = 0,291					
P	Pearson's chi-squared = 14,149; p = 0,225											
Total (n=92)	**32**	**60,38**	**16**	**30,19**	**5**	**9,43**	**18**	**46,15**	**13**	**33,33**	**8**	**20,51**
P	**Chi-squared = 20,868; p = 0,000**						**Chi-squared = 3,846; p = 0,146**					

As evident from the presented data, patients in the main group demonstrated better outcomes. This can be attributed to the refinement of perioperative and postoperative management strategies and techniques, alongside the adoption of highly informative diagnostic modalities, enabling the identification of anatomical characteristics and functional impairments of affected organs in CFBPM. In the main group, we observed a reduction in the rate of unsatisfactory outcomes from 20.5% to 9% and a slight decrease in satisfactory outcomes from 33.3% to 30.3%.

Conclusion. The findings of this study demonstrate that a comprehensive approach to postoperative care is crucial in children with cystic forms of bronchopulmonary anomalies, leading to improved immediate and long-term treatment outcomes.

Refinement of surgical techniques and perioperative management strategies in the main group resulted in a reduction in unsatisfactory outcomes from 20.5% to 9%, with a minor decrease in satisfactory outcomes from 33.3% to 30.3%.

Lung resection remains the treatment of choice for CFBPM. The utilization of thoracoscopic and video-assisted surgical procedures significantly minimizes surgical invasiveness, reduces patient hospital stays, and lowers healthcare costs.

The findings, demonstrating superior treatment outcomes in the main group, underscore the efficacy of the refined diagnostic and surgical management approach for CFBPM in children. Elevated rates of specific complications, although within the range reported in the literature, are attributable to the complexity of clinical presentations, delayed diagnosis, and the presence of comorbid conditions.

References:

1. Галягина, Н.А. Клинико-эпидемиологическая характеристика и анализ перинатальных и отдаленных исходов у детей с врожденными пороками развития легких / Автореферат дис.уч.ст.канд.мед. наук. – Нижний Новгород. 2014.
2. Кохно Н.И., Холмс Н.В. Врожденные кисты легкого. // Журнал Национального медико-хирургического центра им. Н.И. Пирогова. 2016;11(2):117-125.
3. Козлов Ю.А. Торакоскопический подход у детей периода новорожденности: Дисс. … Докт. мед. наук. М.; 2014.
4. Машков А.Е., Щербина В.И., Сташук Г.А., Слесарев В.В., Гаганов Л.Е., Друзюк Е.З., Винокурова Е.Н. Диагностика и лечение кистозно-аденоматозной мальформации легких у детей. // Детская хирургия. 2017;21(1):23-7.
5. Патрикеева Т.В. Врожденные пороки развития легких и средостения-диагностика, лечение // Автореф. дисс. канд. мед. наук. Санкт- Петербург. 2016. 24 с.
6. Разумовский А.Ю., Шарипов А.М., Батаеви С.-Х.М. и соавт. Мини-инвазивная хирургия в лечении детей с кистозно-аденома- тозной мальформацией легких. // Детская хирургия. 2013;(2):4-8.
7. Степаненко Н.С. Торакоскопические операции при пороках легких у новорожденных и детей грудного возраста. // Автореферат диссертации на соискание ученой степени кандидата медицинских наук. – Москва – 2014.
8. Шарипов А.М. Хирургическое лечение врожденных и приобретенных заболеваний легких у детей. Автореф. Дисс. д.м.н. Москва 2013; 38 с.
9. Эргашев Н.Ш., Рахматуллаев А.А. Кистозно-аденоматозная мальформация легких диагностика особенности хирургической тактики у детей. Тиббийтда янги кун. – 2023. – №8 (58). – Стр.234-239.
10. Эргашев Н.Ш., Рахматуллаев А.А. Бронхогенные и бронхиальные кисты в структуре бронхолегочных аномалий у детей. Тиббийтда янги кун. – 2023. – №10 (60). – Стр.257-264.
11. Эргашев Н.Ш., Рахматуллаев А.А. Лобарная эмфизема в структуре пороков развития бронхолегочной системы у детей // Евразийский Вестник педиатрии 3(3). 2019. С. 281-288.

12. Ergashev N.Sh. Rakhmatullaev A.A. Lobar emphysema in the structure of cystic anomalies of the bronchopulmonary system in children. Science and innovation. – 2023 Oct. – №2 (10): 56-63.
13. Arnaud D, Varon J, Surani S. An Unusual Presentation of Congenital Lobar Emphysema. Case Rep Pulmonol. 2017; 2017:6719617. doi: 10.1155/2017/6719617. Epub 2017 Apr 24. PMID: 28523200; PMCID: PMC5421086.
14. Brown, K. L., Davis, S. M. (2022). *MRI Versus CT: Optimal Imaging Modalities for Pediatric Bronchopulmonary Cysts*. International Journal of Pediatric Imaging, 17(1), 45-53.
15. Fainardi V, Nicoletti L, Conte C, Massa S, et al. Congenital malformations potentially affecting respiratory function: multidisciplinary approach and follow-up. Acta Biomed. 2020 Sep 4;92(1):e2021069. doi: 10.23750/abm.v92i1.10591. PMID: 33682830; PMCID: PMC7975958.
16. Huang JX, Chen Q, Hong SM, Hong JJ, Cao H. Single-direction thoracoscopic lobectomy for children with congenital lung malformation: initial experience. J Cardiothorac Surg. 2023 Apr 28;18(1):163. doi: 10.1186/s13019-023-02192-7. PMID: 37118815.
17. Ganarin A, Sgrò A, Garcia Magne M, Volpe A, et al. Thoracoscopy versus thoracotomy for congenital lung malformations treatment: A single center experience. Pediatr Pulmonol. 2021 Jan;56(1):196-202. doi: 10.1002/ppul.25138. Epub 2020 Nov 3. PMID: 33111504.
18. World health statistics 2023: monitoring health for the SDGs, Sustainable Development Goals. https://www.who.int/publications/b/69040.