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CONGENITAL BRONCHIAL MALFORMATIONS ASSOCIATED WITH CONGENITAL HEART DEFECTS

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Abstract. The association of congenital bronchial anomalies with congenital heart defects is linked to disruptions in embryonic development, which necessitates careful monitoring and early intervention for patients suspected of having these anomalies. Pathophysiological processes can significantly vary depending on the type of anomaly, its severity, and the involvement of other organs. Therefore, it is crucial to conduct thorough examinations of patients with suspected congenital bronchial anomalies to identify any accompanying disorders early and prevent life-threatening risks.

Key words: Congenital bronchial anomalies, congenital heart defects, atrial septal defect, patent ductus arteriosus, hypoplasia of the left ventricle, total pulmonary venous anomaly, pulmonary hypertension, right atrium, hypertrophy of the right ventricle, diagnosis, treatment, comprehensive approach, surgical intervention, pathophysiology, embryonic development, monitoring, early intervention, examinations, accompanying disorders, life-threatening risks.

Introduction. The association between congenital bronchial anomalies and congenital heart defects originates from disruptions in embryonic development. These anomalies are not only anatomically interconnected but also share common pathophysiological mechanisms, clinical manifestations, and treatment strategies. The coexistence of bronchial anomalies and congenital heart defects often leads to significant impairments in both the respiratory and circulatory systems, complicating disease progression and management.

Since the formation of the respiratory and cardiovascular systems is closely linked during embryogenesis, even minor developmental disturbances can result in various pathological conditions. Such anomalies frequently coexist with atrial septal defect, patent ductus arteriosus, left ventricular hypoplasia, and total pulmonary venous anomaly. These conditions contribute to pulmonary hypertension and respiratory insufficiency, manifesting as breathing difficulties, hypoxia, and heart failure in affected patients.

Therefore, early detection of congenital bronchial anomalies and their associated heart defects, along with comprehensive diagnostic and targeted therapeutic approaches, is crucial.

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Studying these pathologies not only enhances diagnostic and treatment capabilities but also plays a vital role in preventing severe complications and improving patient outcomes.

Relevance and necessity of the study. The relevance and necessity of this topic align with the findings in the scientific literature (G.I. Kolpinskiy, S.N. Burdin, A.S. Shkaraburov, 2016; I.I. Zakirov et al., 2014), which indicate that a significant proportion of children born with congenital bronchial anomalies do not have other congenital defects. However, research has revealed that in cases where congenital bronchial anomalies are present, congenital heart defects are frequently identified as well. The consistent co-occurrence of congenital heart defects in all observed cases highlights the significance of this topic.

Studies conducted in Uzbekistan confirm that congenital bronchial anomalies are often associated with the following congenital heart defects: atrial septal defect, patent ductus arteriosus, left ventricular hypoplasia, total anomalous pulmonary venous return, pulmonary artery hypertension, pulmonary veins draining into the right atrium, and hypertrophy of the right atrium and right ventricle. The correlation between specific types of congenital bronchial anomalies and congenital heart defects has been established.

Congenital lung diseases are rare, but their clinical presentation and progression vary significantly, ranging from large masses to minor asymptomatic lesions. Some cases require urgent surgical intervention due to their severity. The consequences of congenital lung anomalies can be critical, making their timely diagnosis and treatment highly relevant.

Understanding lung development and anatomy serves as a fundamental basis for assessing these anomalies, helping to comprehend the pathophysiology of each condition and analyze various diagnostic approaches. Research in this area holds particular scientific and practical significance, contributing to the advancement of diagnostic and therapeutic strategies.

Objective. Quantitative indicators were processed using descriptive and variation statistical methods. The obtained micropreparations were analyzed at 200x magnification, and cellular structures within the field of view were mathematically calculated using the NanoZoomer (REF C13140-21, S/N000198, HAMAMATSU PHOTONICS, 431-3196, JAPAN). Morphometric parameters were measured in micrometers (μm) using the Hamamatsu (QuPath-0.4.0, NanoZoomer Digital Pathology Image) software, and statistical analysis was performed to determine the average values and overall statistical significance.

Materials and Methods. To study the fundamental changes occurring in the lung and bronchial tissues of children who died from pulmonary edema due to congenital bronchial anomalies, immunohistochemical (IHC) analysis was conducted using the markers CD-3, CD-20, CD-34, and VGFR-1. These markers were used to assess pathological changes in lung tissue, bronchial, and bronchiolar walls, examining each marker's specific pathway.

Paraffin-embedded biopsy samples were subjected to immunohistochemical examination following standard protocols using monoclonal antibodies. Sections of 4 μm thickness were prepared from paraffin blocks, mounted on glass slides, and dried at room temperature for 24 hours. Before staining, the sections were incubated at 55°C for 60 minutes in a thermostat in a vertical position. Deparaffinization was performed in two xylene baths (10 minutes in each), followed by rehydration through a decreasing ethanol concentration series (three ethanol baths, 3 minutes each), and then rinsed in distilled water.

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For antigen retrieval, the slides were heated in a demasking buffer at 98°C in a water bath for 30–40 minutes. After cooling to room temperature, the samples were washed in Tris-buffered saline (pH=7.5). To block endogenous peroxidase activity, the sections were treated with 3% hydrogen peroxide (H_2O_2) for 15 minutes. To reduce nonspecific binding and limit background staining, the samples were incubated with Protein Block (X0909, DAKO) for 10 minutes. Before the addition of primary antibodies, a special delimiting medium was applied to prevent reagent waste and sample overflow. The slides were then incubated with primary antibodies at room temperature for 60–120 minutes.

For visualization, the Universal LSAB2 KIT (DAKO) detection system was used, with a minimal exposure time of 40 minutes.

Research results and discussion. Our study identified that congenital bronchial anomalies were frequently associated with the following congenital heart defects: atrial septal defect, patent ductus arteriosus, left ventricular hypoplasia, total anomalous pulmonary venous return, pulmonary artery hypertension, pulmonary veins draining into the right atrium, and hypertrophy of the right atrium and right ventricle. Specific congenital bronchial anomalies were correlated with particular congenital heart defects.

Congenital lung diseases are rare; however, their presentation and progression vary significantly. These conditions range from large masses to minor, asymptomatic lesions, some of which necessitate urgent surgical intervention. The consequences of congenital lung defects can be severe, often leading to critical complications.

In this study, pulmonary edema was found to be the primary cause of death. In most cases, congenital bronchial anomalies were identified as the primary underlying pathology (see Table 1).

The incidence of specific morphological types of congenital bronchial anomalies is presented in Table 1 and Figure 1.

1-table.

Incidence of Morphological Types of Congenital Bronchial Anomalies (Based on Clinical Diagnosis)

№	Type of Defect	Number of Cases	Proportion of Total Cases (%)
1	Bronchial agenesis	7	6,0
2	Bronchial aplasia	7	6,0
3	Bronchial hypoplasia	10	8,6
4	Cystic bronchial hypoplasia	12	10,3
5	Tracheobronchomegaly (Mounier-Kuhn syndrome)	12	10,3
6	Tracheobronchomalacia	10	8,6
7	Williams-Campbell syndrome	5	4,4
8	Leschke's bronchiolectatic emphysema	7	6,0
9	Tracheal and bronchial stenosis	7	6,0
10	Tracheoesophageal fistula	10	8,6

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11	Bronchial and tracheal diverticula	7	6,0
12	Parabronchial lung cysts	5	4,4
13	Hamartomas	5	4,4
14	Kartagener syndrome	5	4,4
15	Tracheal bronchus	7	6,0
	Total	116	100

Based on the analysis of clinical diagnoses recorded in medical histories, bronchial agenesis was identified in 7 cases, bronchial aplasia in 7 cases, bronchial hypoplasia in 10 cases, cystic bronchial hypoplasia in 12 cases, tracheobronchomegaly (Mounier-Kuhn syndrome) in 12 cases, tracheobronchomalacia in 10 cases, Williams-Campbell syndrome in 5 cases, Leschke's bronchiolectatic emphysema in 7 cases, tracheal and bronchial stenosis in 7 cases, tracheoesophageal fistula in 10 cases, bronchial and tracheal diverticula in 7 cases, parabronchial lung cysts in 5 cases, hamartomas in 5 cases, Kartagener syndrome in 5 cases, and tracheal bronchus in 7 cases. This data confirms the varying prevalence of different congenital bronchial anomalies in the studied population

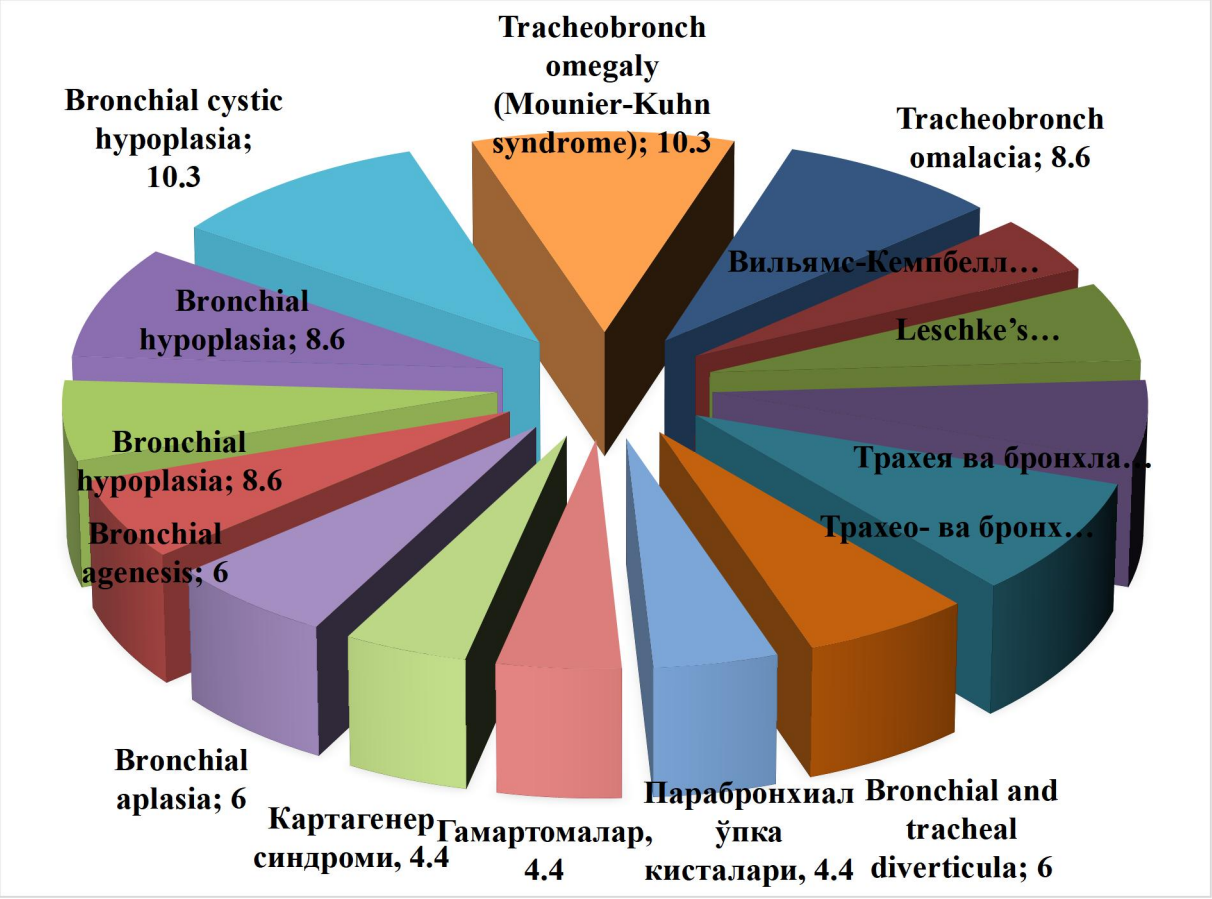

Figure 1. Distribution of Morphological Types of Congenital Bronchial Anomalies (Based on Clinical Diagnosis)

In conclusion, based on the analysis results, Group 1—newborns aged 0-7 days—was the most affected, with 75 cases (64.6%). The second most affected group was Group 2, consisting of newborns aged 8-19 days, with 32 cases (27.6%). Groups 4 and 5, representing

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older children, had the lowest occurrence, with only 2 cases each (1.7%). In Group 3—newborns aged 20-29 days—there were 5 cases (4.4%).

The aim of this study was to determine the prevalence of congenital bronchial anomalies in children and to identify the early morphological changes leading to mortality. The primary cause of death in this study was pulmonary edema. In most cases, congenital bronchial anomalies were identified as the underlying disease.

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