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# MORPHO-FUNCTIONAL ANALYSIS OF LUNG HYPOPLASIA IN CHILDREN.

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According to the literature, there is a continuous increase in the number of children suffering from chronic pneumonia, the pathogenesis and pathomorphology of which are based on congenital anomalies and malformations of the bronchopulmonary structures [1,3]. Among the malformations, pulmonary hypoplasia is the most common, accounting for 75-80% and characterized by a steady increase in morbidity among children, an increase in the incidence of extensive, advanced and severe forms of damage [5,6].

The difficulties of early detection of HL are associated with the fact that the disease occurs under the guise of a chronic suppurative process in the lung. Recognition of pulmonary hypoplasia at the stages of primary diagnosis is difficult due to the lack of clear clinical and morphological changes that are smoothed out against the background of a long-term inflammatory process, in connection with which the number of erroneous diagnoses ranges from 73 to 94.5%. [4, 6 ]

The purpose of this work was to study the morpho-functional changes in various forms of pulmonary hypoplasia in children. The work is based on the results of examination and treatment of 93 children with pulmonary hypoplasia aged from 3 months to 15 years, who were hospitalized for the period from 2013 to 2023 in the clinic of TashPMI and TMA.

The distribution of patients by age was as follows: up to 1 year - 12 children, from 1 year to 5 years - 2 children, from 6 to 10 years - 36, from 11 to 15 years - 24 patients. There were 55 boys, 33 girls. Lesions of 1 lobe on one side were observed in 53 (56.9%) patients, lesions of 2 lobes on one side - in 14 (15%) patients, lesions of individual segments in 10 (10.7%) patients, bilateral lesions in 16 (172%) patients.

The examined groups included children with pulmonary hypoplasia: severe form (SF), simple form (SF), cystic form (CF), congenital lobar emphysema (CLE), postnatal hypoplastic bronchiectasis (PHB). Congenital lobar emphysema and postnatal hypoplastic bronchiectasis are described in textbooks and manuals on pediatric surgery as separate nosological entities. We included them in the pulmonary hypoplasia group. According to the classification of the Research Institute of

Pulmonology of the Russian Federation (2000), this is justified by the fact that all forms of lung underdevelopment are based on hypoplasia of the bronchial wall, bronchioles and alveoli as a whole or in a limited area of the lung [7,8].

The morphological picture of HL was studied in 39 patients, including 18 (46.1%) with CF, 3 (7.6%) with VLE, 11 (28.2%) with PF, and 7 (18%) with GSD.

The functional characteristics of external respiration and hemodynamics in the pulmonary circulation in HL were studied using spirography, echocardiography, and ECG in 58 (62.3%) children with HL aged 5 to 15 years; the examined children were divided into 2 groups[9,10]..

*The first group* (41 children) included children with antenatal HL (RF, PF, CF, VLE). The duration of the disease in these children was more than 4 years, the course was characterized by frequent relapses, occurring with the phenomena of severe intoxication and respiratory failure. *The second* group (GB, 17 (18.2%) children) included children whose disease progressed more favorably than in group 1, although these children suffered from colds quite often. The disease duration exceeded 2 years, relapses were rare.

**Results and discussion**: In cystic hypoplasia of the lungs, the results of microscopic studies conventionally divided the results into two morphological types:

impaired development of the proximal sections of the bronchial tree and 2) impaired development as a result of reduction of its distal sections. The first morphological type of CF could be evidence of an ontogenesis disorder that occurs as a result of pneumomeres, "amputation" of the bronchial tree, which ceases to divide in the first generations (the first type of bronchographic variants of cystic hypoplasia). Histologically, this type was characterized by the formation of large cystic cavities, which were undivided pneumomeres immersed in a stroma poor in vessels. At the same time, an unordered type of branching was revealed with the transition of large, sharply dilated and deformed bronchi into cystic cavities. Cystic-altered large-caliber bronchi are closely located among the fields of fibrous, hyalinized connective tissue, areas of proliferation of adenomatous structures and muscular sclerosis. Segmental, as well as interlobular and intralobular bronchi, were not expressed. The cartilaginous framework of the bronchi was sharply reduced (Fig. 1). In the presence of large cysts, the above-mentioned signs of cystic hypoplasia are determined in the tissue surrounding them. Mucous glands, a lining of cylindrical or cubic epithelium, clusters of cartilaginous islands of various sizes, as well as daughter cysts are constantly visible in the walls of such cysts. Also characteristic of the first type of CF was the absence of formed alveolar tissue (Fig. 2).

The second type was characterized by a simplification of the bronchial tree structure due to the reduction of small bronchi and partial underdevelopment of the alveolar tissue. Histologically, cystic cavities were determined, located among the alveolar parenchyma, which was reduced in quantity. The bronchi were cystically dilated, their wall was represented by hyalinized connective tissue. The cystic cavities were lined with columnar epithelium, the flask-shaped cells were atrophied, the walls of the cavities contained bundles of smooth muscles and fibrous structures without elements of the bronchial wall. Areas of atelectasis with adenomatous structures and randomly located fields of muscular sclerosis alternated with foci of emphysema, sharply dilated respiratory bronchioles and alveoli. A mosaic histological picture was noted - underdeveloped and cystically altered areas of the lung tissue were presented along with normally developed bronchi and respiratory sections. The presence of alveolar structures in areas of cystic hypoplasia can be explained by the fact that when some bronchial buds stopped developing, others developed up to the respiratory sections. In this case, the developmental defect of a certain area of the lung was not continuous, but mosaic in nature. Respiratory sections among bronchial cysts arose not only when the development of the bronchial tree stopped, but also when the development processes were distorted, which we have repeatedly seen in histological studies, when cystically altered bronchi did not end blindly, but continued to divide, resulting in a branched system of interconnected cysts related to several successive generations of bronchi. Thus, the presence of respiratory structures, including alveoli, does not contradict the idea that this defect occurs in the early antenatal period.

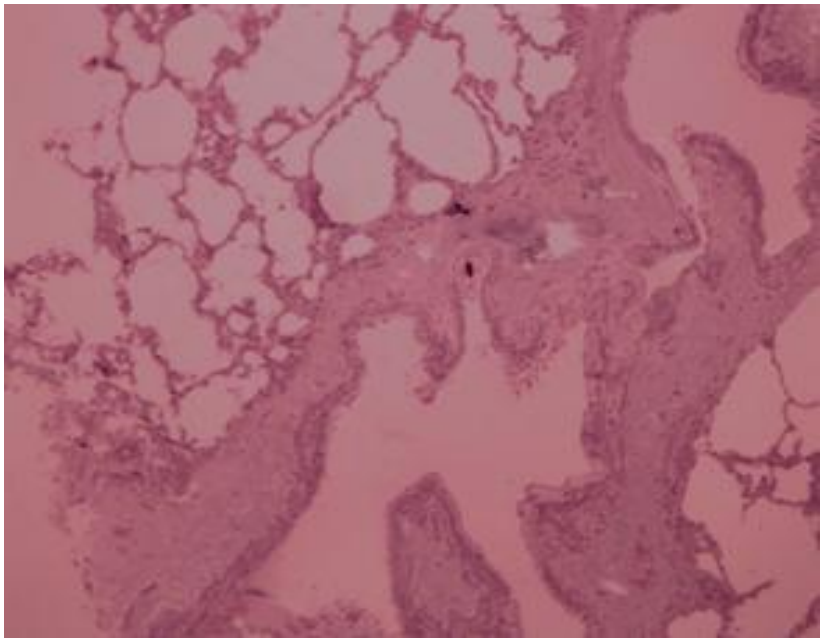

Fig. 1. The cystically dilated bronchus has a lumen of a bizarre shape. Its inner surface is lined with high cylindrical epithelium lying on the basement membrane. Stained with hematoxylin and eosin. Magnification x 120

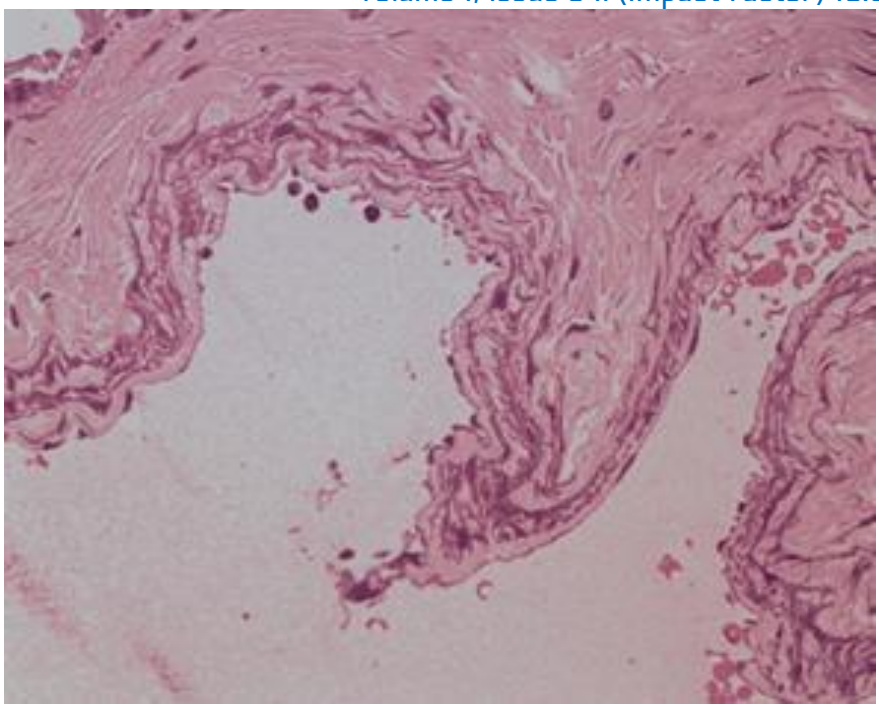

Fig. 2. Small cysts are represented by terminal bronchioles with pronounced lymphoid peribronchiolar infiltration. Hematoxylin and eosin staining. Magnification x 12

In congenital lobar emphysema, macroscopic examination did not reveal any violations of the anatomical patency of the lobar and segmental bronchi. The removed lobes were sharply swollen, pale pink formations of a consistency tightly packed with feathers, without individual emphysematous bubbles. After cutting off from the root, the lobe did not subside. Histological studies revealed sharply swollen respiratory bronchioles and alveolar ducts with a noticeable decrease and underdevelopment of the elastic framework in the respiratory sections (Fig. 3), uneven expansion of the alveoli, without ruptures of the interalveolar septa, and parenchyma sclerosis. In none of our observations were signs of malformation of the leading large bronchi found, which indicates the localization of the main pathology in the respiratory sections (Fig. 4). Macroscopic examination of PF hypoplasia shows that the lung volume is reduced, the visceral pleura is thickened, the interlobar fissure is obliterated, and the normal lobar anatomical structure of the lung is absent. The section shows dilated and deformed segmental and subsegmental bronchi that end blindly with the formation of multiple bronchiectasis. The microscopic examination allowed us to divide them into 2 conditional types: the first type included morphological changes around large bronchi - pronounced fibrous changes. The lumen of small bronchi is narrowed, the mucous and muscular layers are thickened, in some places small bronchi are dilated with the formation of bronchiectasis. In the defect area, the absence of twisted bundles of elastin and collagen was found, which gives grounds to speak of the absence of alveolar tissue. The pulmonary vessels are deformed. Arteriovenous shunts are detected in the root area. In atelectatic lung tissue, small vessels are narrowed and obliterated due to the proliferation of connective tissue (Fig. 5).

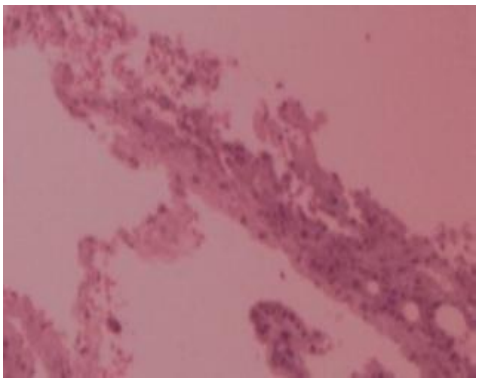

Fig. 3. Elements of the bronchiole wall are hypoplastic and dilated, interalveolar septa are thinned. Hematoxylin and eosin staining. Magnification x 200

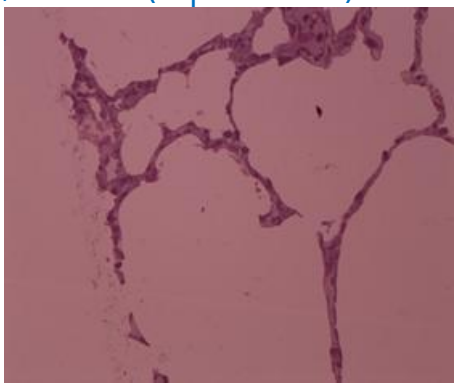

Fig. 4. Enlarged alveoli, thinned interalveolar septa, sclerosis and fibrosis in places. Hematoxylin and eosin staining. Magnification x 200

In the second type, narrow bronchi continue to the subpleural sections, where they are arranged in spirals and disintegrate into small trunks; a decrease in the number of bronchial generations and hypoplasia of the respiratory section were detected in them. In general, these structures resemble randomly located small bronchi and bronchioles. Their walls are thinned, the mucous membrane is smoothed, fibrously changed, the number of mucous glands is reduced. Alveoli are found near them, and it is possible to find communications between them. Signs of inflammation, expressed to varying degrees, and small bronchioloectasias are also noted (Fig. 6).

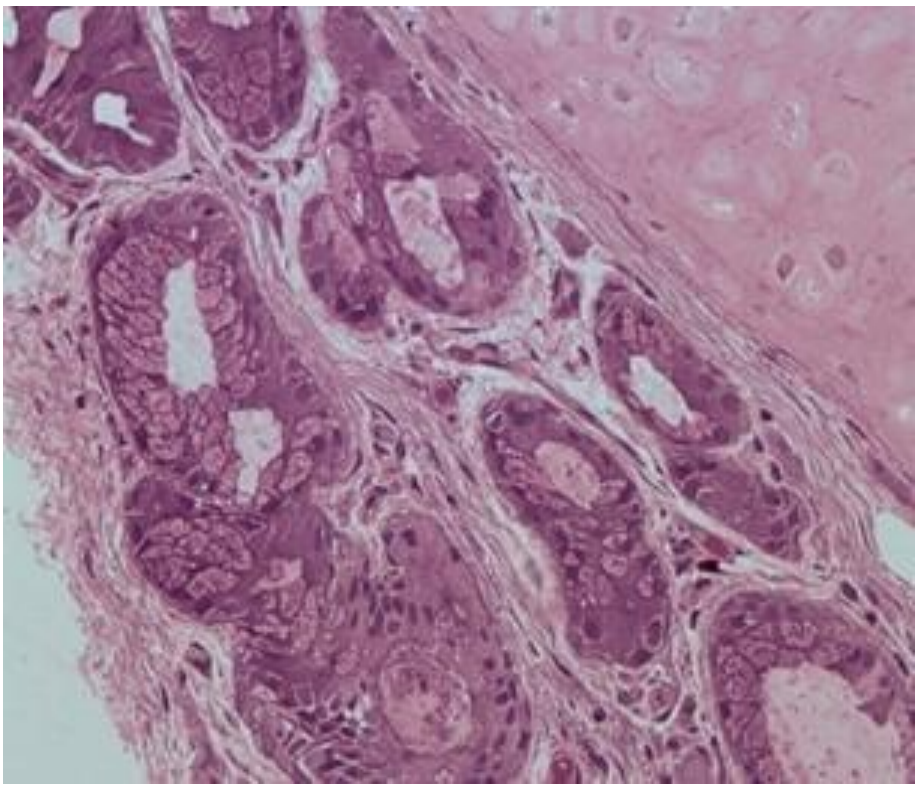

Fig. 5. Bronchial wall with dilated bronchial glands, plethora of vessels of the proper plate of the mucous membrane. Hematoxylin and eosin staining. Enlargement x 200

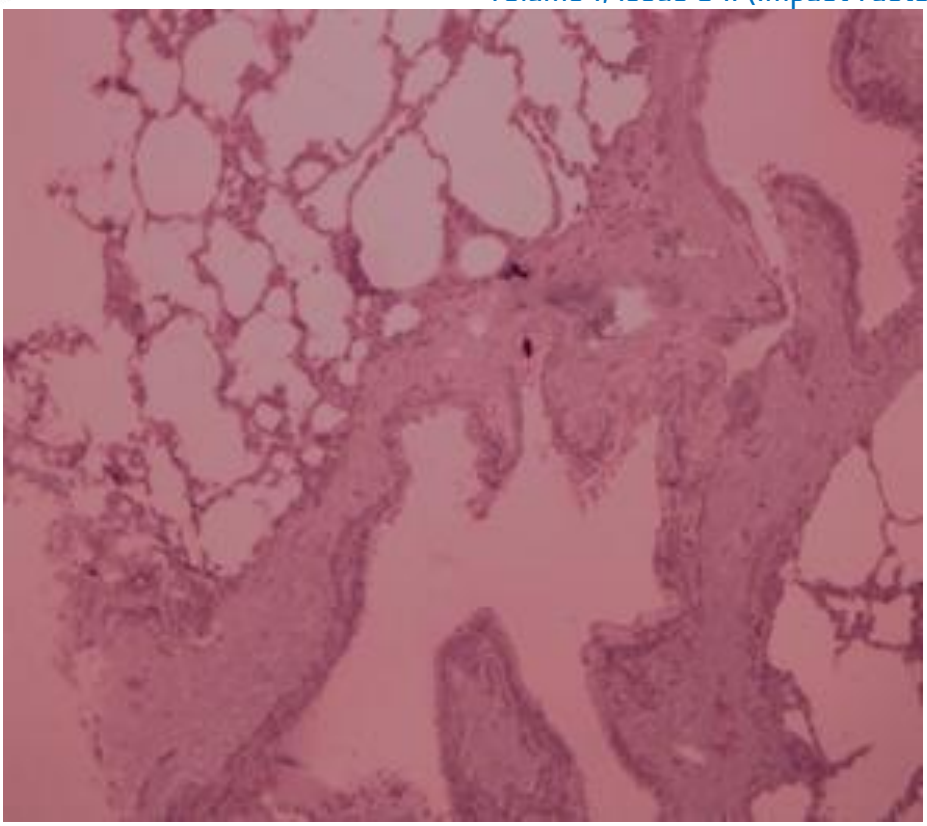

Fig. 6. Inflammatory infiltration of the bronchiole wall, in the lumen - secretion with a large number of neutrophils, cellular detritus. Hematoxylin and eosin staining. Enlargement x 200

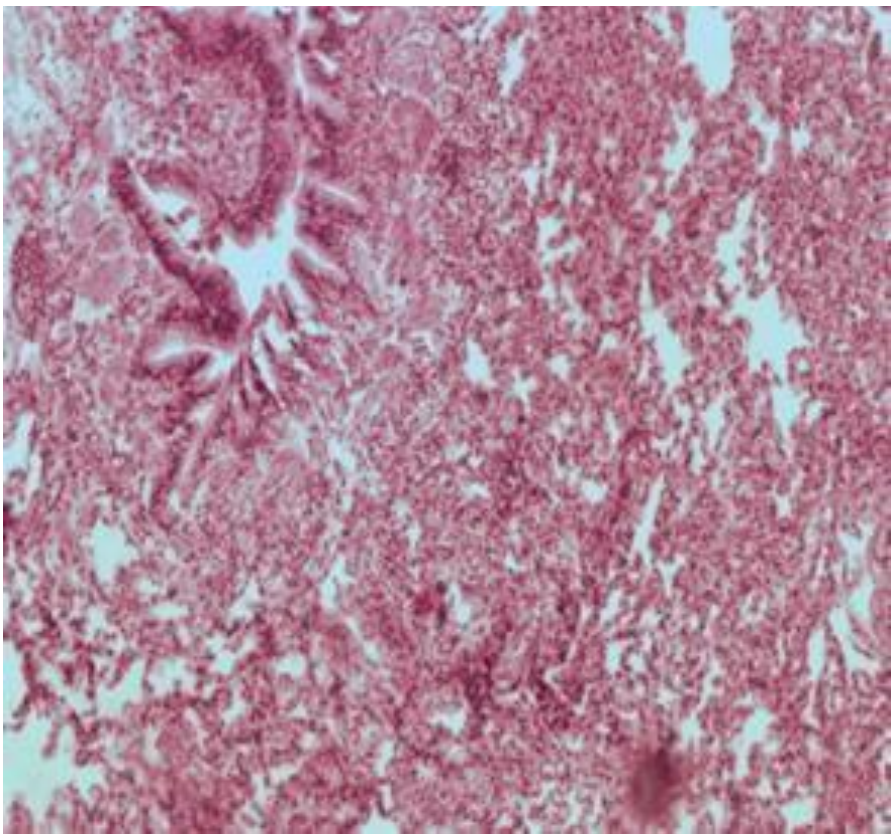

Fig. 7. Obliterating bronchiolitis with adenomatous structures in the bronchial lumen. Hematoxylin and eosin staining. Magnification x 80

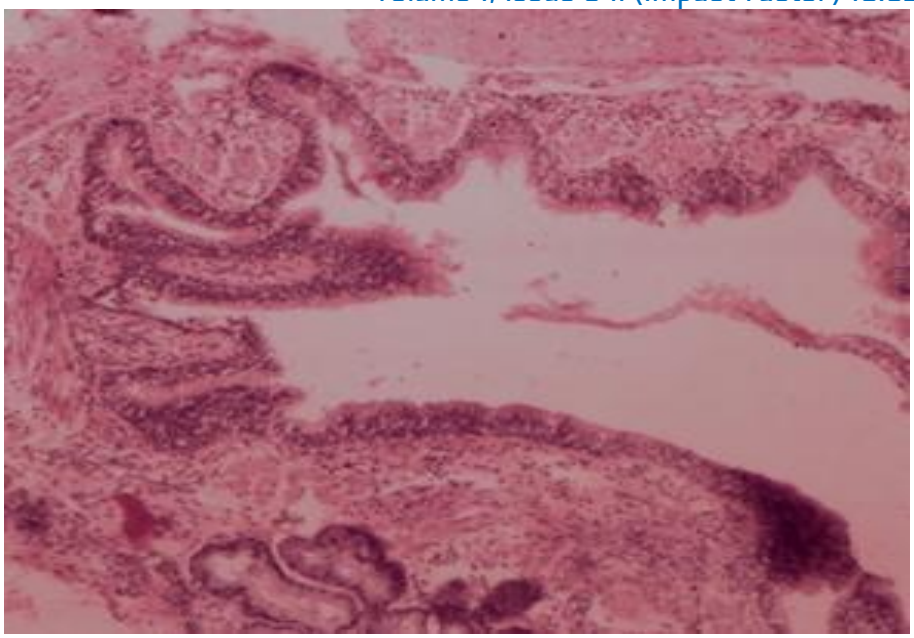

Fig.8.Bronchioloectasis with pronounced lymphoid peribronchiolar infiltration. Hematoxylin and eosin staining. Magnification x 120

## Conclusions.

•**Types of Morphological Changes in Cystic Hypoplasia of the Lungs**: The results are divided into two types: impaired development of the proximal sections of the bronchial tree (Type 1) and reduction of the distal sections (Type 2). Type 1 is characterized by large cystic cavities and the absence of alveolar tissue, while Type 2 involves a simplified bronchial tree structure and partial underdevelopment of alveolar tissue.

•**Histological Features of Type 1 Cystic Hypoplasia**: This type shows large cystic cavities lacking alveolar tissue, thickened bronchial walls, and disordered branching of the bronchial tree. Microscopically, cyst walls consist of hyalinized connective tissue with smooth muscle bundles and fibrous structures, but no typical bronchial wall elements.

•**Macroscopic and Microscopic Characteristics of Congenital Lobar Emphysema**:
Macroscopically, lobar and segmental bronchi remain patent, while the affected lobes are overinflated. Histologically, there is swelling of respiratory bronchioles and alveolar ducts, underdevelopment of the elastic framework, and areas of emphysema and parenchymal sclerosis.

•**Pathology of Pulmonary Vessels and the Bronchial Tree**: Pulmonary vessels are deformed, with arteriovenous shunts in the root zone and obliteration of small vessels due to connective tissue proliferation. Large cystically

altered bronchi are surrounded by adenomatous structures, hyalinized connective tissue, and areas of muscular sclerosis.

- **Mosaic Histological Pattern**:

Underdeveloped and cystically altered lung tissue alternates with normally developed bronchi and respiratory sections. This mosaic development suggests that certain areas of the lung stopped developing early in the antenatal period, with the presence of branched cystic structures linked to multiple bronchial generations.

Thus, morphological studies indicate that various forms of pulmonary hypoplasia are characterized by a common feature: underdevelopment, insufficient differentiation and various disorders of all structural elements of the lung. In the second type and the third "A" type of HL, morphological changes in the form of alternation of sections occur, there is a reduction in the branching of the bronchial tree, agenesis of the respiratory sections and preservation of the alveolar structure of the lung parenchyma, due to a developmental defect of the organ in the early stages of embryogenesis. In the third "B" type of HL, the basis of the developmental defect of the lung is a decrease in the branching of the bronchial tree with preservation of the pulmonary parenchyma, which can be repatriated.

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