LOBAR EMPHYSEMA IN THE STRUCTURE OF CYSTIC ANOMALIES OF THE BRONCHOPULMONARY SYSTEM IN CHILDREN

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Abstract. *Congenital lobar emphysema constitutes a significant part of the malformations of the bronchopulmonary system. The severity of clinical signs and the course of the disease correlate with the volume of damage to the pulmonary parenchyma. A subcompensated course is more often observed. The radical method of treatment is surgical correction, depending on the volume and location of the lesion. After resection, dynamic monitoring of the condition of the remaining areas of the lungs is necessary.*
Keywords: *malformations of the bronchopulmonary system, lobar emphysema, diagnosis, treatment, children.*

Introduction. Congenital lobar emphysema (CLE) is a malformation of the parenchyma and small bronchi, characterized by stretching of the parenchyma of one lobe of the lung or its segments, and makes up a significant part of cystic lung anomalies. The affected lobe increases in volume several times and contains non-collapsing bullae of varying sizes. Pathology often manifests itself in the neonatal period and in infants. The literature presents cases of diagnosing the disease in adolescents 16 years old [10, 11]. The anomaly is registered in one case per 70-100 thousand children born and accounts for approximately 14% of all congenital lung anomalies. It is more common in boys than in girls [1, 2, 4, 6, 12, and 16]. VLE is classified as a correctable defect with an ambiguous prognosis; there are known cases of antenatal fetal death, disability from childhood, and asymptomatic course in the postnatal period. Good immediate and long-term postoperative results are possible. Currently, endosurgical methods for correcting defects of the bronchopulmonary system are widely used in practice. They have an advantage over open operations due to their low morbidity [8, 9, 12, 14, 16, 18]. Methods of pre- and postnatal diagnostics are being improved using ultrasound and computerized tomography [2, 4, 7, 11].

The purpose of the study is to analyze diagnostic methods, clinical and anatomical features of congenital lobar emphysema and the results of surgical correction in children based on clinical data of patients.

Materials and methods. In the Hospital Pediatric Surgery Department of the Tashkent Pediatric Medical Institute, were examined and treated 115 children with bronchopulmonary anomalies, 19 (16.5%) patients were diagnosed with CLE. Patients underwent comprehensive clinical studies to assess the anatomical and functional state of the lungs and identify concomitant diseases; Ultrasound, X-ray, MSCT examinations of the chest organs. Collected results were verified by using surgical data and the results of morphological studies of resected areas of the lung.

Results and discussion CLE in all 19 observations occurred in the form of a symptoms complex with corresponding clinical manifestations; various forms of hyperextension. With damage to one lobe or several segments of the lung due to congenital anomalies; accompanied by

interstitial emphysema and bronchopulmonary dysplasia. Acquired forms that arise in children as a result of prolonged artificial ventilation or previous diseases of the bronchopulmonary system were not encountered in our observations. The patients were aged from 1 day to 3 years which were hospitalized in a surgical hospital with characteristic clinical signs of the disease. There were 7 newborns (36.8%); 1-3 months - 4 (21.1%); 4-12 months – 6 (31.6%); 1 - 3 - 2 (10.5%). There were 14 boys (73.7%), 5 girls (26.3%). According to the type of course, there were 8 patients with decompensated defect, 10 subcompensated and 1 compensated.

The distribution of patients by age depending on the time of admission was uneven. This indicates undiagnosed cases of the disease in newborns due to insufficient examinations and poor clinical symptoms in children in the postnatal and subsequent period of the child's life. Undiagnosed VLE can cause fatal complications.

The introduction of antenatal examination of pregnant women to identify congenital defects in the fetus has significantly increased the detection of abnormalities in lung development. When performing antenatal ultrasound in 8 fetuses, specific (macro- or microcystic changes, signal hyperintensity of the parenchyma of the vicious lobe) and indirect (polyhydramnios, fetal hydrops and symptoms of displacement or compression of other organs due to increased pulmonary volumes) characteristic ultrasound signs of bronchopulmonary anomalies were revealed. In 4 (50%) cases, the diagnosis was confirmed postnatally by the results of a comprehensive examination. Our data coincide with the literature that prenatal studies for congenital malformations of the bronchopulmonary system are crucial in the final diagnosis. However, the results may be inconsistent and level off by 30-33 weeks of pregnancy. Thus, out of 8 antenatally diagnosed cases, 4 (50%) turned out to be false positive.

Clinical signs of the disease in 5 (26.3%) patients were detected in the first days or weeks of life; in 9 (47.4%) - during the neonatal period; in 3 (15.8%) in infancy; in 2 (10.5%) children of older age groups. Before admission to the clinic, the course of the disease was asymptomatic in 1 (5.3%), fewer symptomatic in 7 (36.8%) children. Early clinical manifestation with symptoms of respiratory failure and hemodynamic disturbances was observed in 5 (26.3%) children with extremely severe lung damage and 6 (31.6%) patients in combination with anomalies of other organs and systems. And also in 2 () children with pectus excavatum - and 4 with congenital heart disease. The main clinical signs of the disease are symptoms of respiratory failure, which worsen when the child is fed. An objective examination reveals a bulge on the corresponding side of the chest lesion; decreased conductivity of breathing with a boxy tint of percussion sound; displacement of the borders of the mediastinum in the opposite direction (Fig. 1). Data from clinical, radiological and morphological studies indicate the involvement of all three variants of ARF in CLE. This is a hypoxemic ARF (shunt-diffusion), characterized by insufficient blood oxygenation with relatively adequate ventilation. Hypercapnic ARF (ventilation) resulting from primary hyperventilation followed by a sharp decrease in its volume and severe hypercapnia. Mixed ARF is manifested by hyperventilation and an increase in the alveolocapillary gradient. In 4 (28.6%) patients, characteristic changes in concomitant conditions were noted: most often in the cardiovascular system in the form of an open "foramen oval". In the decompensated course, 5 (26.3%) patients had asymmetry of the chest, weakness of heart sounds, and displacement of the percussion boundaries of the liver or spleen due to an increase in the volume of lung tissue in the chest.

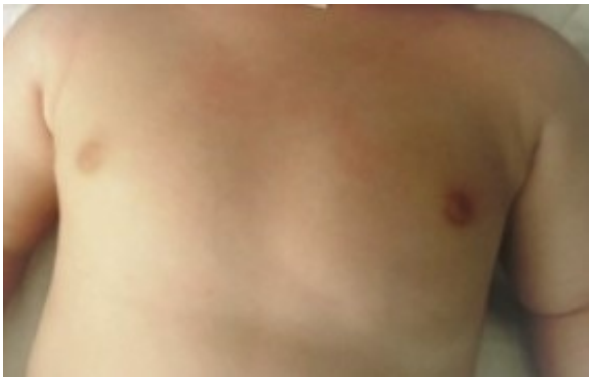


Fig. 1. Asymmetry of the chest in the form of bulging on the left.

In 2 cases, the complicated course of the disease was provoked by inadequate puncture of the pleural cavity and erroneous interpretation of CLE instead of tension pneumothorax. Our data confirm the opinion of many surgeons that intrapulmonary punctures in the treatment of CLE are inappropriate, as they are fraught with serious complications.

Chest X-ray is the main method of examination when a defect is suspected and the initial assessment of the extent of lung damage. Plain radiographs (Fig. 2) reveal sharply increased transparency and hyperairiness, an increase in the volume of the affected lobe, and distortion of the vascular pattern on the affected side. Depending on the severity of emphysema, displacement of the mediastinum is possible, sometimes with compression and atelectasis of the lung on both sides; smoothness and low standing of the dome of the diaphragm on the affected side. Due to the displacement of the organs, the opposite side appears darkened, and the pulmonary pattern in it appears enhanced. Usually the affected lobe is increased in volume several times. Significant stretching causes increasing symptoms of respiratory failure and decompensated condition of the patient.

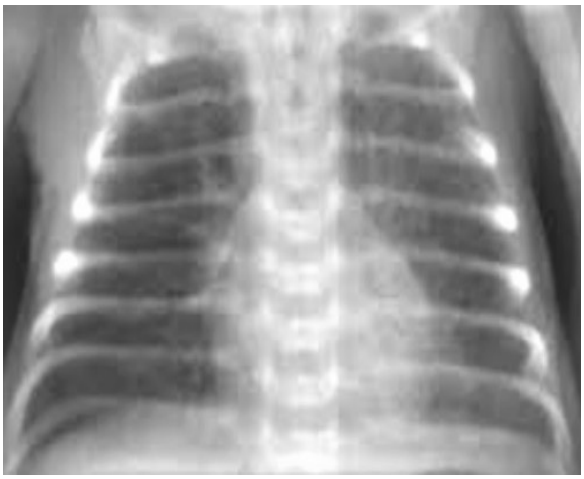


Fig. 2a.

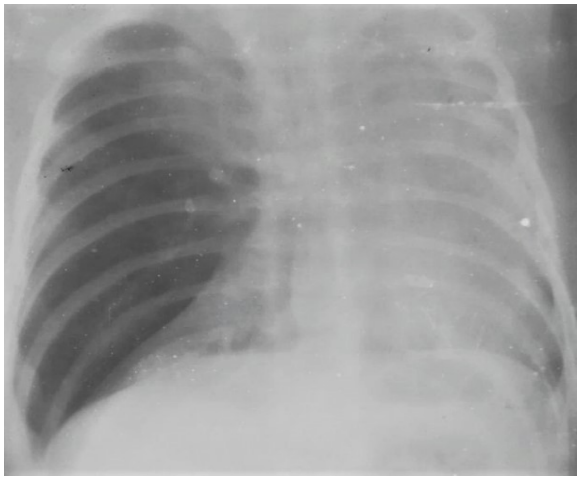


Fig. 2b.

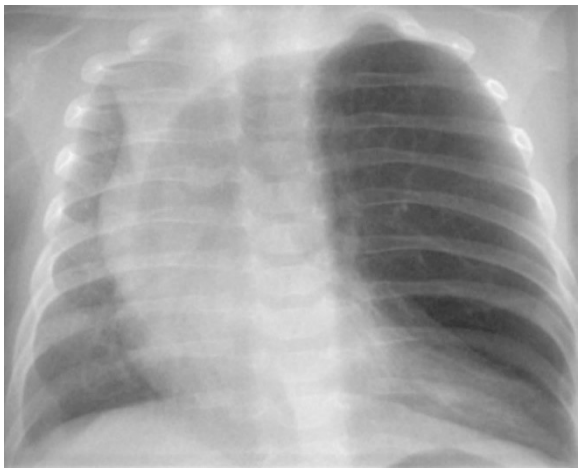
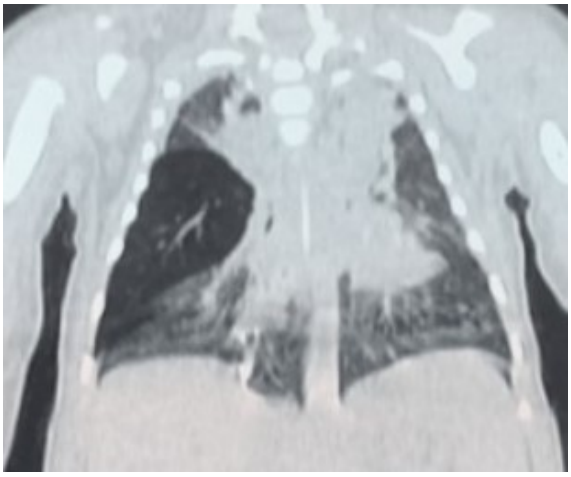
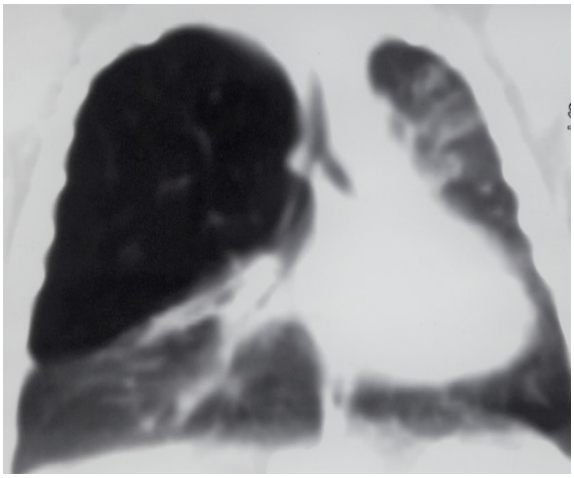


Fig. 2в.

Fig. 2. Radiographic: a) with moderate lung distension; b) with significant stretching of the lung with compression of the pulmonary parenchyma and adjacent organs; c) formation of a mediastinal hernia with compression of organs on the opposite side.





To identify cystic lung lesions, an MSCT examination is required (Fig. 3).

Fig. 3a. ***Fig. 3b.***

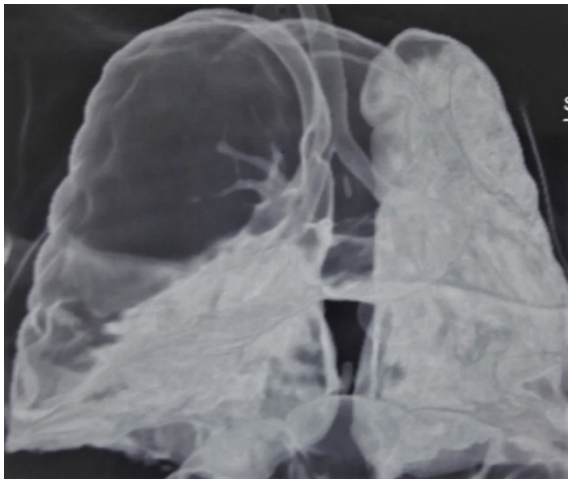


Fig. 3в.

Fig. 3. Computer tomograms: a) with moderate lung distension; b) with significant stretching of the lung with compression of the pulmonary parenchyma and adjacent organs; c) formation of a mediastinal hernia with compression of organs on the opposite side.

Computed tomography reveals an increase in the transparency of the lung tissue, an increase in the volume of the affected lobe, and a narrowing of the bronchi. Axial CT imaging allows for an accurate diagnosis. The use of MSCT angiography makes it possible to identify congenital lung defects associated with abnormalities of blood vessels. This study is the final method of confirming or refuting a suspected lung malformation established by the results of a postnatal X-ray examination. In 15 (78.9%) patients, the pathology was localized in the right lung: with damage to the upper lobe – in 8; middle lobe – in 6; in 1 – covering two lobes (upper and middle). Corresponding changes on the left side were detected in 4 (20.1%) patients: upper lobe – in 2; lower lobe – in 2. No lesions were observed in the two lobes on the left. Echocardiography is an important method for identifying associated cardiovascular abnormalities.

Table 1

Combined anomalies in CLE (n= 19)

Forms of CLE										
	CVS		MPS		ODS		GIT		CNS	
	Abs.	%	A		A		A		Abs.	%
Compensated	-	-	-	-	-	-	-	-	-	-
Subcompensated	1	5,3	-	-	2	10,5	-	-	-	-
Decompensated	3	15,8	-	-			-	-	-	-
Total	4	21,1	-	-	2	10,5	-	-	-	-

Surgical correction is the only radical treatment for CLE. The duration and indications for surgical treatment are determined individually for each patient, taking into account the severity and dynamics of clinical symptoms. Of the 19 patients, 8 - 42.1% had indicators corresponding to the decompensated stage. Increased clinical signs of the disease with increasing respiratory failure in a subcompensated course were noted in 3 (15,8%). A stable condition corresponding to a subcompensated course was observed in 5 (26.3%) patients. In 2 (10.5%) cases, the intensity of respiratory failure decreased with stabilization of the children's condition. In 1 (5.3%) with a compensated course, progression of clinical signs of the disease was not observed. In 5 children diagnosed antenatally with a decompensated form of pathology, surgical intervention was performed in the early neonatal period on an emergency basis. In 3 cases with progression of subcompensated course and in 7 cases with persistent sub compensation, surgical intervention was performed in early infancy (3-4 months). The provoking factors for the increase in clinical symptoms were frequent colds against the background of ongoing bronchitis. With a stable compensated course in 1 observation, surgical intervention was performed as planned 6 months after diagnosis. Surgical intervention was performed in 7 (36.8%) newborns for emergency and urgent indications; up to 3 months of age – 7 (36.8%) patients, 3 (15.8%) patients under 1 year of age and 2 (10.6%) patients under 3 years of age.

The intensity and duration of preoperative preparation of patients with CLE depends on the course of the disease and the nature of the complications that have developed. Preoperative preparation included sanitation of the tracheobronchial tree, antibacterial therapy, inhalation of humidified oxygen, and correction of threatening changes in the CBS. The use of sedatives helped reduce the symptoms of respiratory failure. The choice of surgical treatment method was determined taking into account the volume and location of the lesion. Posterolateral thoracotomy in the IV-VI intercostal space was predominantly used, providing good intraoperative visualization of the anatomical structures to be resected. In 1 observation, the operation was performed with video-assisted (VATS) intervention (Table 2).

Table 2

Surgeries performed due to CLE (n=19)

Forms of CLE	Lobectomy		Bilobectomy		Segmentectomy		Pulmectomy		video-assisted thoracoscopic (VATS) lobectomy	
	Абс.	%	Абс.	%	Абс.	%	Абс.	%	Абс.	%
Compensated	1	5,3	-	-	-	-	-	-	-	-
Subcompensated	10	52,6	-	-	-	-	-	-	-	-
Decompensated	6	31,5	1	5,3	-	-	-	-	1	5,3
Total	17	89,4	1	5,3	-	-	-	-	1	5,3

In case of total lesion of the lung lobe, a typical anatomical lobectomy was performed with separate treatment of the root elements in 18 (94.7%) patients: upper - 11, middle - in 7. In 1 (5.3%) case of multilobar lesion (upper and middle lobe on the right) a bilobectomy (upper and middle) of the right lung was performed. If there was excessive mobility of the remaining sections of the lung after resection, the adjacent sections of the organ were fixed to the parietal pleura to prevent torsion. The operation was completed by drainage of the pleural cavity. The drainage tube was removed after straightening of the lung tissue, confirmed by X-ray examination, and cessation of discharge from the pleural cavity on days 5-6 after surgery. The postoperative period in 14 (73.7%) patients was without complications. In 1 case there was a prolonged release of air. This required extension of drainage to 10 days. In 4 cases, postoperative pneumonia developed, which was relieved by intensive therapy. Long-term results were monitored in 15 (78.9%) of 19 operated patients. Children develop according to their age, the results are considered good. In 2 (10.5%) cases, emphysematousness of varying severity in the remaining areas of the lung was noted, resembling a relapse of the disease without characteristic clinical signs in 1 with a tendency to frequent colds in the second. During dynamic observation, no increase in clinical and radiological data was noted. This allows us to consider this condition a compensatory process that does not require repeated surgical intervention.

As conclusion we can say that PE of congenital origin makes up 16.5% of PBLS in children and manifests itself in the form of a compensated (5%), subcompensated (53%), decompensated (42%) course. Antenatal and postnatal diagnosis of the disease is necessary to identify pathology

in the early, including preclinical stages, which will allow planning adequate treatment. The severity of clinical signs and the degree of intrathoracic tension correlate with the nature and extent of damage to the pulmonary parenchyma, revealed by objective data and radiological diagnostic methods. Concomitant developmental defects aggravate the course of the disease and the severity of clinical manifestations. Surgical correction of the defect is a radical treatment method. The timing of surgical intervention should be determined individually for each patient. Signs characteristic of a decompensated and subcompensated course are an indication for emergency surgery. In case of compensated course – perform a planned operation for urgent indications. The method of choice is to remove the affected areas of the lung in the form of lobectomy or bilobectomy. Anatomical resection of the affected lobe of the lung with separate ligation of vessels and suturing of the stump of the lobar bronchus allows achieving optimal results and avoiding the development of complications in the immediate and late postoperative period.

The immediate and long-term results of timely operations are satisfactory. The phenomena of emphysema in the remaining areas of the lung after surgery can be regarded as compensatory changes. However, this does not exclude the risk of hidden dysplasia that requires dynamic monitoring.

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