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Features Of Pulmonary Function Impairment In Children With Cystic Fibrosis According To Computerized Spirometry Data

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Abstract: Objective. To assess the state of pulmonary function in children with cystic fibrosis and to determine the nature and severity of ventilatory disorders based on computerized spirometry data.

Materials and Methods. Pulmonary function was assessed in 64 children with cystic fibrosis aged 5 to 14 years who were receiving inpatient treatment. The control group included apparently healthy children and children with recurrent obstructive bronchitis. Computerized spirometry was performed using a BTL-08 Spiro Pro spirometer with evaluation of the main

spirometric parameters (FEV_1, vital capacity, Tiffeneau index, MEF50).

Results. All children with cystic fibrosis demonstrated a reduction in pulmonary function parameters. The combined type of ventilatory impairment was the most frequently observed (62.5%), characterized by the presence of both restrictive and bronchial obstructive components. A statistically significant decrease in FEV_1, vital capacity, Tiffeneau index, and MEF50 was noted compared with children with recurrent obstructive bronchitis and apparently healthy peers ($p<0.001$). In the majority of patients, marked and severe deviations of pulmonary function indices were identified, reflecting progression of chronic bronchopulmonary involvement.

Conclusion. Children with cystic fibrosis exhibit a pronounced and statistically significant deterioration of pulmonary function parameters, predominantly of a combined ventilatory impairment type. Computerized spirometry is an informative and accessible method for assessing the severity of bronchopulmonary involvement and for monitoring the effectiveness of therapy in this patient population.

Keywords: Cystic fibrosis, children, pulmonary function, computerized spirometry, ventilatory disorders, FEV_1.

1. Introduction: Among the five thousand orphan diseases described to date, cystic fibrosis is the most common. The number of newly diagnosed patients is increasing annually. The worldwide prevalence ranges from 1:2000 to 1:100,000 [3,4,8,9,10].

Over the past decade, the quality and life expectancy of patients with cystic fibrosis (CF) have significantly improved due to the refinement of therapeutic approaches and the development of new methods targeting the pathogenetic mechanisms of the disease [1,5,6,7].

The chronic inflammatory process leads to sclerosis of the lung parenchyma. It is evident that a comprehensive assessment of pulmonary function in patients with CF is only possible through the combined use of various diagnostic methods. The current "gold standard" for investigation is spirometry, which allows for the assessment of both obstructive and restrictive types of ventilation disorders. This method is highly informative; however, its application in pediatric practice is limited by the necessity for the patient's active participation in the procedure, which becomes feasible in children over 5 years of age. Currently, spirometry is the "gold standard" for assessing pulmonary function. It enables the clinician to detect the presence of ventilatory insufficiency, as well as the type and degree of ventilation impairment [2].

Purpose of the research

To assess the state of pulmonary function in children with cystic fibrosis and to determine the nature and degree of ventilatory disorders according to computerized spirometry data.

2. Methods

We performed computerized spirometry on 64 children with cystic fibrosis aged 5 to 14 years, who were undergoing inpatient treatment in the pulmonology department. The pulmonary function test (PFT) was conducted using a BTL-08 Spiro Pro spirometer (UK) at the Republican Specialized Scientific and Practical Medical Center for Pediatrics of the Ministry of Health of the Republic of Uzbekistan under the guidance of Professor F.M. Shamsiev.

3. Results And Discussion

Computerized spirometry was performed on patients starting from 5 years of age. The severity of the identified lung ventilation disorders was assessed based on FEV1. Analysis of pulmonary function in patients with cystic fibrosis, regardless of age, revealed various PFT impairments. The pulmonary function study was conducted in 64 children over 5 years old. All children with CF had reduced respiratory function.

In children with cystic fibrosis, the combined type of lung ventilation impairment was most frequently detected. This type was characterized by a predominance of the restrictive ventilation disorder pattern, confirmed by a decrease in vital capacity (VC), along with a simultaneous reduction in FEV1, indicating obstruction and the development of inflammation and fibrosis (Table 1).

Table 1

Spirometric Indicators in Patients with CF

Indicators	Recurrent OB (n=25)	CF (n=64)	P
	abs. %	abs. %	
Restrictive type	3 12.0	15 23.45	<0.05
Obstructive type	16 64.0	9 14.0	<0.001
Combined type	8 32.0	40 62.5	<0.01

Note: P - significance of differences in indicators between the recurrent OB and CF patient groups.

In the comparison group, spirometry in children with recurrent OB predominantly showed an obstructive type of lung ventilation impairment, accounting for 64.0% (16 children). The combined type of ventilation impairment was observed in 8 children (32.0%), and the restrictive type in 3 children (12.0%). In most children, indicating impaired bronchial patency, a simultaneous decrease in both FVC and the Tiffeneau index was determined. This most often arises due to inflammatory changes in the bronchi and lungs. In cystic fibrosis, the combined type of ventilation impairment was most common, found in 40 children (62.5%). The restrictive type was found in 15 patients (23.45%), and the obstructive type in 9 pediatric CF patients (14.0%). In the group of children with cystic fibrosis, obstructive and combined types of ventilation impairment were observed significantly more frequently ($P<0.001$ and $P<0.01$, respectively), while differences for the restrictive type were not statistically significant.

A statistically significant decrease in pulmonary function parameters was identified in children with cystic fibrosis (Table 2). In the CF group, FEV1 was within the range of significant deviation, at 42.03±0.9. In the comparison group, this indicator was 58.88±0.86, which is 1.4 times higher compared to CF children. The average vital capacity (VC) in the cystic fibrosis group was 49.52±1.05, which is 1.3 times lower than in the recurrent OB group and 1.6 times lower compared to practically healthy children (83.05±2.0). The average Tiffeneau index (FEV1/VC) in children with CF was 56.18±1.03, which is 1.2 times lower compared to recurrent OB children (68.42±1.12) and 1.4 times lower compared to practically healthy children (80.29±2.1). Moderate deviation of all parameters in the cystic fibrosis group was observed in 15 (23.4%) patients. Significant PFT changes were noted in 28 (43.75%) children, and severe deviations in 21 (32.8%) children.

Table 2.

Characteristics of PFT Parameter Impairments in Children with CF and Recurrent OB (M±m)

Parameter	Practically Healthy Children (n=20) (I)	Recurrent OB (n=25) (II)	Children with CF (5 to 14 yrs) (n=64) (III)	P	P1
FEV1	88.62±1.7	58.88±0.86	42.03±0.9	<0.001	<0.001
VC	83.05±2.0	65.12±1.01	49.52±1.05	<0.001	<0.001
FEV1/VC	80.29±2.1	68.42±1.12	56.18±1.03	<0.001	<0.01
MEF50	92.95±1.6	56.22±0.93	49.13±1.0	<0.001	<0.05
PFT Deviations, n (%)	20 (100)	25 (100)	64 (100)		
Moderate	5 (25.0)	9 (36.0)	15 (23.4)	<0.001	<0.01
Significant	0	11 (44.0)	28 (43.75)	<0.001	<0.001
Severe	0	5 (20.0)	21 (32.8)	<0.001	<0.001

Note: P - significance of differences in indicators between patient groups I and II; P1 - significance of differences in indicators between patient groups II and III.

In children suffering from cystic fibrosis, pulmonary function parameters were significantly lower than in practically healthy peers and patients with recurrent obstructive bronchitis. The most substantial differences were revealed in the values of FEV_1 and MEF50, which were highly statistically significant ($p<0.001$).

Thus, children with cystic fibrosis exhibit a pronounced and significant deterioration in pulmonary function parameters compared to both the comparison and control groups. The maximum reduction is noted in the indicators FEV1, VC, and the Tiffeneau index, indicating the presence of a pronounced broncho-obstructive syndrome. In the majority of examined patients, significant deviations in PFT parameters were recorded, reflecting the progression of chronic bronchopulmonary system involvement and the gradual decline of lung ventilation function.

4. Conclusions

All examined children with cystic fibrosis showed impairments of pulmonary function of varying severity, confirming early and progressive involvement of the bronchopulmonary system in this disease.

The most characteristic finding for children with cystic fibrosis is a combined type of ventilation impairment, combining signs of restriction and bronchial obstruction, which occurred significantly more frequently compared to the group of children with recurrent obstructive bronchitis.

Computerized spirometry is an informative method for monitoring the state of the respiratory system in children with cystic fibrosis and should be used to assess disease severity and the effectiveness of ongoing therapy.

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