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ROLE OF COMPUTED TOMOGRAPHY FOR EVALUATION OF LUNGS CYSTIC FIBROSIS IN CHILDREN

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Abstract

BACKGROUND: *Cystic Fibrosis is a hereditary disease that mainly affects the lungs and it becomes worse with the time leading to chronic respiratory issues and infections. Early diagnosis and management are crucial to slowing disease progression. Computed Tomography Imaging is a valuable tool for assessing lungs involvement, particularly in pediatric patients with cystic fibrosis. It provides detailed visualization of lung structures and airway abnormalities, allowing for early detection of complications like bronchiectasis, hyperinflation, and atelectasis. Despite its potential, the role of CT in early diagnosis and ongoing monitoring in children with cystic fibrosis remains under-explored.*

OBJECTIVE: *To evaluate the role of computed tomography in assessing lung involvement in children with cystic fibrosis.*

METHODOLOGY: *This descriptive cross-sectional study was conducted at Children Hospital Lahore. Data were collected using a high-resolution 640-slice CT scanner over a period of 4 months. A total of 40 pediatric patients were selected through convenient sampling. Inclusion criteria included children aged 1 to 15 years with a confirmed diagnosis of cystic fibrosis and clinical indication for CT imaging. Imaging was performed using a multi-detector CT scanner. Data were analyzed using SPSS version 25.*

RESULTS: *This study conducted at Children Hospital Lahore included 40 children with cystic fibrosis, mainly aged 1–15 years. Computed tomography demonstrated significant lung involvement, with bronchiectasis present in 80% of patients, hyperinflated lungs in 65%, and atelectasis in 47.5%. Additional findings included peri bronchial thickening and air trapping in 42.5% of cases each. The majority of patients also showed evidence of chest infections clinically, correlating with CT abnormalities. These results underscore the vital role of high-resolution CT in early detection, detailed evaluation, and monitoring of pulmonary changes in children with cystic fibrosis.*

CONCLUSION: *The study concluded that computed tomography is an effective and essential tool for assessing lung involvement in children with cystic fibrosis. It detected key pulmonary abnormalities such as bronchiectasis,*

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hyperinflation, atelectasis, and peri bronchial thickening in the majority of patients. These findings helped guide clinical management and improved monitoring of disease progression, contributing to better care for pediatric cystic fibrosis patients.

INTRODUCTION

Cystic fibrosis (mucoviscidosis) is the most common life-shortening multisystem disease with an autosomal recessive inheritance pattern in Germany today, affecting 1 in 3300 to 1 in 4800 neonates and affecting approximately 1 in 4000 newborns in the US. It is caused by dysfunction of the chloride channels of exocrine glands, specifically of the so-called cystic fibrosis transmembrane conductance regulator (CFTR) protein. It mainly involves the lungs and pancreas, but also the upper airways, liver, intestine, and reproductive organs.[1] Cystic fibrosis (CF), a monogenic disease caused by mutations in the CFTR gene on chromosome 7, is complex and greatly variable in clinical expression. The lack of CFTR or its impaired function causes fat malabsorption and chronic pulmonary infections leading to bronchiectasis and progressive lung damage. Previously considered lethal in infancy and childhood.[2] It is characterized by pancreatic insufficiency and chronic endobronchial airway infection. This latter feature results in progressive bronchiectasis and ultimately respiratory failure, which is the leading cause of death in patients with CF. Other complications include sinusitis, diabetes mellitus, bowel obstruction, hepatobiliary disease, hyponatremic dehydration, and infertility.[3] Diagnosis of CF in the non screened population can be challenging because the age of onset and severity of symptoms can differ greatly as a result of highly variable levels of CFTR dysfunction. Presenting manifestations can include pancreatitis, respiratory symptoms, chronic sinusitis, and male infertility.[4] Respiratory disease remains the predominant manifestation of CF with a progressive decrease in lung function, most commonly assessed with the spirometry measurement of forced expiratory volume in the 1st second (FEV_1) standardized for height and sex and expressed as percent predicted (pp) of normal. The major cause of premature death in CF is lung disease progression with the eventual development of respiratory failure.[5] Recent data highlighted a number of defects identifiable at birth with CF. These include congenital airway abnormalities, increased acidity of the airway surface liquid that results in inhibition of the function of antimicrobial peptides, and failure of mucus to detach from submucosal gland ducts. These basic defects lead to the clinical consequences of infection, inflammation, functional abnormalities, and lung structural damage.[6] Computed tomography (CT) is the imaging reference method in the diagnosis, assessment and management of lung disease. In the setting of cystic fibrosis (CF), CT demonstrates increased sensitivity compared with pulmonary function tests and chest radiography, and findings correlate with clinical outcomes. Better understanding of the etiology of CF lung disease indicates that even asymptomatic infants with CF can have irreversible pulmonary pathology. Surveillance and early diagnosis of lung disease in CF are important to preserve lung parenchyma and to optimize long-term outcomes. CF is associated with increased cumulative radiation exposure due to the requirement for repeated imaging from a young age. Despite the radiation doses incurred, CT remains a vital imaging tool in children with CF.[7] Nowadays it is regarded as the gold standard for the identification of airways and lung parenchymal structural changes in CF. Computed tomography can identify a wide range of morphological abnormalities in patients with cystic fibrosis, such as bronchiectasis (which is progressive, irreversible and probably the most relevant structural change in cystic fibrosis) peri bronchial thickening, mucous plugging and many other disorders that occur in the course of the disease. Computed tomography has a crucial role in the assessment of pulmonary damage over time, detecting complications and monitoring treatment effects in patients with cystic fibrosis.[8]

Material and Methods:

This descriptive cross-sectional study was conducted at Children Hospital Lahore. Data were collected using a high-resolution 640-slice CT scanner over a

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period of 4 months. A total of 40 pediatric patients were selected through convenient sampling. Inclusion criteria included children aged 1 to 15 years with a confirmed diagnosis of cystic fibrosis and clinical indication for CT imaging. Patients with contraindications to CT imaging, severe renal impairment, inability to hold breath, or unstable clinical conditions were excluded. Imaging was performed using a multi-detector CT scanner with dedicated lung analysis software, 3D reconstruction tools, and non-ionic iodinated contrast agents. Data were analyzed using SPSS version 25.

Results:

This descriptive cross-sectional study was conducted at Children Hospital Lahore, involving 40 children diagnosed with cystic fibrosis. The age distribution showed that the majority of patients (65%) were in the 1–4 years age group, followed by 25% between 4 and 8 years. The older age groups, 8–12 years and 12–15 years, comprised smaller proportions of 2.5% and 7.5%, respectively. This indicates that the sample mainly consisted of younger children. Regarding gender distribution, males represented 62.5% of the sample, while females accounted for 37.5%, showing a higher male predominance in this cohort. Clinically, the overwhelming majority of children did not exhibit microcephaly (97.5%), indicating it is not a common finding in this population. Similarly, diarrhea was rare, affecting only 5% of participants. Respiratory distress was reported in 17.5% of the children, suggesting that most patients were not in acute respiratory distress during evaluation. Fever was present in 35% of the cases, and cough was noted in 32.5%, indicating common respiratory symptoms in this group. Chest infections were prevalent in 62.5% of the children, highlighting the frequent occurrence of infectious complications in cystic fibrosis. Radiological findings from high-resolution computed tomography (CT) scans revealed that pleural effusion was uncommon, observed in only 2.5% of patients, while the vast majority (97.5%) showed no evidence of pleural fluid accumulation. Hyperinflated lungs, a hallmark of cystic fibrosis-related airway obstruction, were present in 65% of cases, confirming its frequent involvement. Bronchiectasis, another key feature of lung damage in cystic fibrosis, was observed in 80% of patients, making it the most common CT finding in this study. Peri bronchial thickening and air trapping were noted in 42.5% of patients each, suggesting widespread airway inflammation and obstruction. Multifocal pulmonary nodules were less common, present in 17.5%, while atelectasis was seen in nearly half the patients (47.5%), reflecting areas of lung collapse likely secondary to mucus plugging or infection. Cross-tabulation and chi-square tests revealed no statistically significant associations between multiple clinical and radiological variables. For instance, microcephaly showed no significant relationship with hyperinflated lungs ($p > 0.05$), and gender did not correlate significantly with the presence of multifocal pulmonary nodules. Similarly, chest infection was not significantly associated with pleural effusion, nor was peri bronchial thickening linked to respiratory distress. No significant associations were found between cough and air trapping, age and atelectasis, chest infection and bronchiectasis, or fever and air trapping. All these findings indicate that these clinical and radiological features appear to occur independently in children with cystic fibrosis. Overall, the results demonstrate a predominance of younger children affected by cystic fibrosis, with common clinical features such as chest infection, fever, and cough, alongside typical CT findings including bronchiectasis, hyperinflation, and atelectasis. The lack of significant associations between many of the studied variables may reflect the complex and multifactorial nature of lung involvement in pediatric cystic fibrosis patients.

Table 1. Association Between Clinical and CT Findings

No.	Variables Compared	χ^2 (Chi-Square)	df	p-value (Asymp. Sig.)	Fisher's Exact (2-sided)	Significant ($p < 0.05$)
1	Microcephaly × Hyperinflated Lungs	0.552	1	0.457	1.000	NO

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2	Gender × Multifocal Pulmonary Nodules	0.104	1	0.747	1.000	NO
3	Chest Infection × Pleural Effusion	0.615	1	0.433	1.000	NO
4	Peri bronchial Thickness × Respiratory Distress	0.744	1	0.388	0.432	NO
5	Cough × Air Trapping	1.015	3	0.314	0.496	NO
6	Age × Atelectasis	2.254	1	0.521	-	NO
7	Chest Infection × Bronchiectasis	0.667	1	0.414	0.686	NO
8	Fever × Air Trapping	1.710	1	0.191	0.315	NO

Table: The statistical analysis of the dataset showed that there are no significant associations between any of the examined variables, as all p-values from the Chi-Square and Fisher's Exact Tests were greater than 0.05. This indicates that variables such as microcephaly, chest infection, pleural effusion, hyperinflated lungs, bronchiectasis, respiratory distress, air trapping, and atelectasis occur independently of factors like age, gender, or the presence of other clinical conditions. In this sample of 40 individuals, no meaningful patterns of co-occurrence were identified, suggesting that these conditions are not statistically linked to one another within the observed group.

Discussion

Cystic fibrosis (CF) is a progressive genetic disorder that primarily affects the lungs, leading to chronic respiratory infections and structural damage. High-resolution computed tomography (HRCT) has emerged as a vital imaging modality for assessing pulmonary involvement in CF, providing detailed visualization of structural abnormalities that may not be apparent through clinical symptoms or conventional chest radiographs. In this descriptive cross-sectional study conducted at Children's Hospital Lahore, 40 pediatric CF patients aged 1 to 15 years were evaluated using a 640-slice CT scanner over a four-month period. The aim was to explore the role of CT imaging in identifying lung involvement and compare the findings with clinical symptoms and previously published research. Bronchiectasis was the most prevalent CT finding in our cohort, observed in 80% of the children. This is consistent with previous studies, particularly the work of Björkman-Burtscher (2013), which emphasized bronchiectasis as the most diagnostically significant radiological marker in CF scoring systems.[9] Similarly, Sasihuseyinoglu et al. (2018) reported a moderate positive correlation between bronchiectasis and clinical parameters, including sputum cultures and pulmonary function tests.[10] However, our study found no statistically significant association between bronchiectasis and chest infection, suggesting either early-stage structural changes or a limitation of using symptom-based cross-sectional assessment. Hyperinflated lungs, identified in 65% of patients, also showed no significant correlation with microcephaly or fever. This differs from the findings of Burgel and Hubert (2018), who developed automated CT density-based scores and reported significant correlations with FEV1%, indicating that CT-based quantification can effectively predict pulmonary function decline.[11] Atelectasis was seen in nearly half of the sample (47.5%), but again, no statistically significant association was found with age groups. While Sanders et al. (2011) demonstrated that CT scores could predict long-term disease severity more accurately than spirometry at the time of scanning, our results did not reflect such associations, likely due to the cross-sectional design and lack of longitudinal follow-up.[12] Air trapping was another frequent finding, affecting 42.5% of the children. Despite its clinical importance in indicating small airway obstruction, our study did not find a significant link between air trapping and symptoms such as cough or fever, contrasting with the earlier work of Sasihuseyinoglu et al., who found such features to correlate with poorer lung function

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outcomes. Peri bronchial thickening, noted in 42.5% of participants, was not significantly associated with respiratory distress. This contrasts with previous literature where such thickening has been described as an early indicator of CF-related airway changes. Similarly, pleural effusion and multifocal pulmonary nodules were less frequently seen and showed no significant association with chest infections or gender. These findings align with Björkman-Burtscher's (2013) conclusion that pleural changes are less weighted in CT-based scoring systems due to their inconsistent presence across patient populations. Despite a high prevalence of clinical symptoms such as cough (32.5%), fever (35%), and chest infections (62.5%), no significant association was observed between these symptoms and CT findings. This is in line with Sanders et al. (2011), who noted that CT imaging is more predictive of future disease progression than concurrent symptoms or spirometry results. The disparity between clinical presentation and radiological findings in our study highlights the crucial role of CT in detecting early or subclinical lung involvement, reinforcing the necessity of imaging for comprehensive CF management. In conclusion, this study supports the utility of high-resolution CT in identifying structural lung abnormalities in children with cystic fibrosis, especially in early disease stages where clinical symptoms may not fully reflect the severity of pulmonary involvement. The most frequent findings such as bronchiectasis, hyperinflated lungs, atelectasis, and air trapping underscore the chronic and progressive nature of CF-related lung disease. Although no statistically significant associations were established between CT features and clinical symptoms, these results align with prior research suggesting that imaging offers a more sensitive measure of disease progression. Therefore, CT imaging should remain an integral component of CF assessment and follow-up, ideally combined with longitudinal data and pulmonary function testing to enhance clinical decision-making and early intervention strategies.

Conclusion

The study concluded that computed tomography is a valuable and reliable imaging modality for evaluating lung involvement in children with cystic fibrosis. It effectively identified common pulmonary abnormalities such as bronchiectasis, hyperinflated lungs, atelectasis, peri bronchial thickening, and air trapping in a significant number of patients. The high detection rate of these changes through CT imaging provided crucial information for early diagnosis, accurate assessment, and monitoring of disease progression. This enabled more informed clinical decision-making and helped tailor management strategies to improve respiratory outcomes in pediatric patients. Overall, the study highlighted the indispensable role of high-resolution CT in the comprehensive evaluation of lung pathology associated with cystic fibrosis in children.

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