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Chest X-Ray Changes in Cystic Fibrosis Patients in A Tertiary Care Center

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Abstract

Introduction*: Chest radiography has been the most commonly used tool for evaluating pulmonary disease in cystic fibrosis (CF) in all age groups, by using the Brasfield scoring system which was created in 1979 to evaluate the evolution of pulmonary abnormalities in CF. The scoring system has a maximum possible score of 25. Points are subtracted based on radiological findings. A higher overall score indicates greater disease severity. The minimum possible score is 3.*

Objectives: *This study evaluates the radiologic findings in pediatric and adult patients with CF by using the Brasfield scoring model at the time of diagnosis.*

Methodology: *A retrospective chart review to identify the Brasfield score on plain chest x-ray findings in our confirmed CF patients from the period 1989-2018.*

Results: *A total of 229 confirmed CF patients. Applying Brasfield score (BS): Twenty-seven (12%) had normal lungs, 33 (14%) had mild changes with BS of 10 points, 77 (34%) had moderate changes with BS of 15 points, 72 (31%) had severe changes with BS of 20 points, 20 (9%) had very severe changes with BS of >21 points with complications as pneumothorax and empyema. Sixteen out of the 92 patients (17%) with BS >20 required lung transplants, 2 of the recipients died post lung transplant.*

Conclusion: *Chest radiography is an important tool to evaluate the severity of pulmonary involvement and to institute proper treatment early to prevent the progression of the disease. More than 40% of CF patients had severe x-ray changes with BS score >20 points that may require lung transplantation or may have a progressive disease.*

Keywords: *Chest x-ray, Brasfield, CFTR, cystic fibrosis, Arab.*

Introduction:

Cystic fibrosis (CF) lung disease begins in early childhood. Evidence of the presence of potentially irreversible pulmonary findings is detected in the chest radiographs of up to 50% of children by the age of 2 years **[**1–3**]**. The pulmonary evaluation of children younger than 6 years is limited, particularly in low–middle-income settings, because of the unavailability of spirometers and chest CT, the difficulty in gaining the cooperation of young children when performing spirometry and the reluctance to expose young children to the higher doses of ionizing radiation attributed to chest CT.

Chest radiography has been the most commonly used tool for evaluating pulmonary disease in all age groups, together with microbiological monitoring of the airway **[**1–3**]**.

The systematized study of chest radiographs by radiographic scoring allowed for the objective analyses of pulmonary lesions and the comparison of disease severity among patients. Authors have recently shown that quantitative chest radiology is the best procedure for the frequent assessment of bronchopulmonary disease in CF **[**4**]**.

The Brasfield scoring system was created in 1979 and has been extensively used scientifically to evaluate the evolution of pulmonary abnormalities in children and adults with CF.

The Brasfield scoring system is simple, has a high degree of intra- and inter-observer reproducibility, and correlates with the child's clinical status and with pulmonary function tests **[**5**]**. Greater knowledge of the disease, early diagnosis, follow-up of patients to control pulmonary infections, and appropriate weight gain has contributed significantly to increasing patient survival **[**6–15**]**.

Cystic fibrosis patients experience a progression of their disease **[**14**]**. However, while chest x-rays are useful to monitor chronic disease progression, acute exacerbations are not always associated with radiographic changes. Therefore, a lack of radiographic findings does not rule out acute pulmonary exacerbations**[**15**]**.

This study evaluates the radiologic findings in pediatric and adult patients with CF by using the Brasfield scoring model at the time of diagnosis and its evolution through the follow-up period.

Aim of the Study

To identify the Brasfield score on plain chest x-ray findings in our confirmed CF patients during the period 1989-2018.

Methodology

A retrospective chart review of all confirmed CF patients who had chest radiological investigations on their first visit, during the period 1st January 1989- 30 December 2018. After obtaining the ethical approval, charts were reviewed, and the information aiming at answering the above objectives was collected, tables and charts have been used to present the data appropriately. A literature review follows along with an appropriate analysis of the available information.

Inclusion Criteria: All confirmed CF patients of all age groups that had chest x-rays done at diagnosis and last follow-up period during the period 1989 – 2016.

Exclusion Criteria: Clinical symptoms, with normal sweat CL level, in addition to pathologic CFTR mutations on one chromosome only. No clinical symptoms, with borderline or normal sweat CL level, and no CFTR mutations.

Brasfield Scoring System: [5,6,13**]**

The Brasfield scoring system is a scoring system for cystic fibrosis. It is based on plain film radiographic findings and has been reported to have a good correlation with pulmonary function. There can however be some subjectivity in scoring between radiologists.

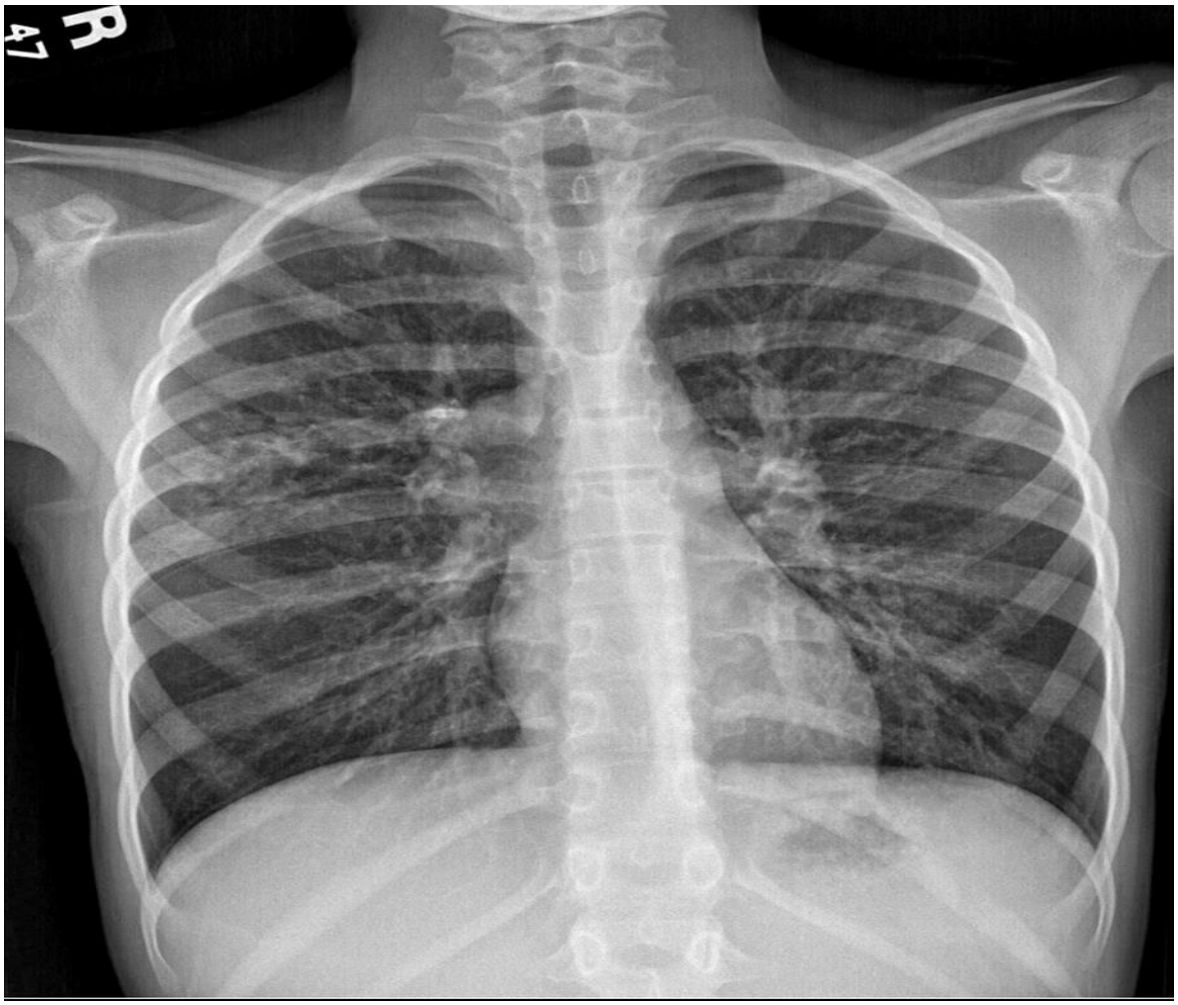

Figure (1): There is bilateral peribronchial wall thickening and branching opacities at the right upper lobe and lingula related to early bronchiectatic changes.

A maximum possible score of 25. Points are subtracted based on the score from each of the following categories:

1. **Air trapping**: 0: absent, 1-3: increasing severity, 4: most severe (figure (1)
2. **Linear markings**: 0: absent, 1-3: increasing severity, 4: most severe (figure (1)
3. **Nodular cystic lesions**: 0: absent, 1-3: increasing severity, 4: most severe
4. **Large lesions**: 0: absent, 3: segmental or lobar atelectasis and pneumonia, 5: multiple atelectasis and pneumonia (figure 2)
5. **General severity:** (impression of overall severity on CXR): 0: absent, 1-3: increasing severity, 4: complications: (cardiac enlargement, pneumothorax), 5: most severe (figure 3 and figure 4).

Overall score: 25 - total demerit points. A higher overall score indicates greater disease severity. The minimum possible score is 3.

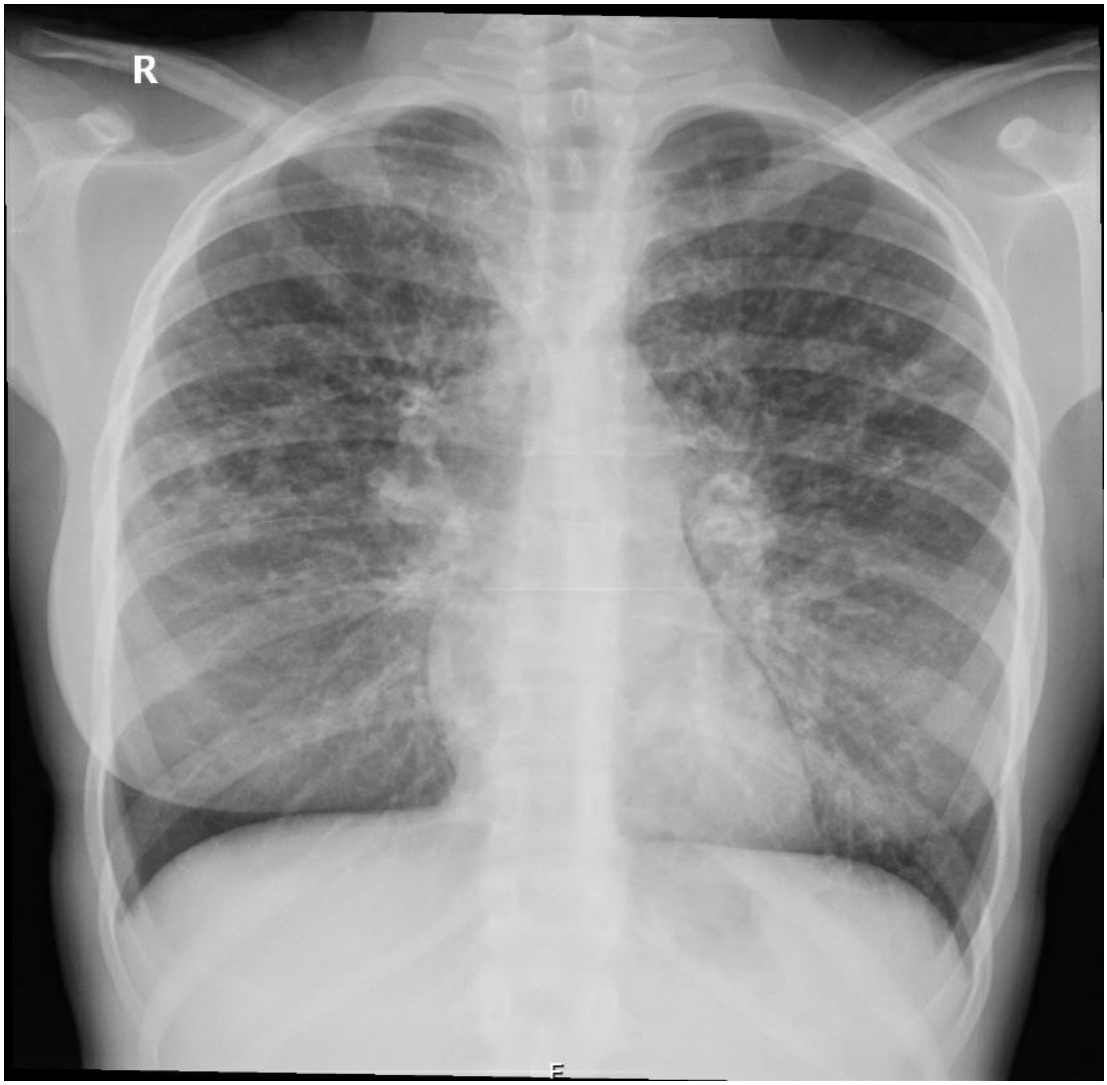

Figure (2): Diffuse bronchiectatic changes throughout both lungs more prominent at the perihilar and upper lobes bilaterally. Peribronchial wall thickening with increased hilar opacities due to underlying enlarged lymphadenopathy. Linear left lower lobe atelectasis.

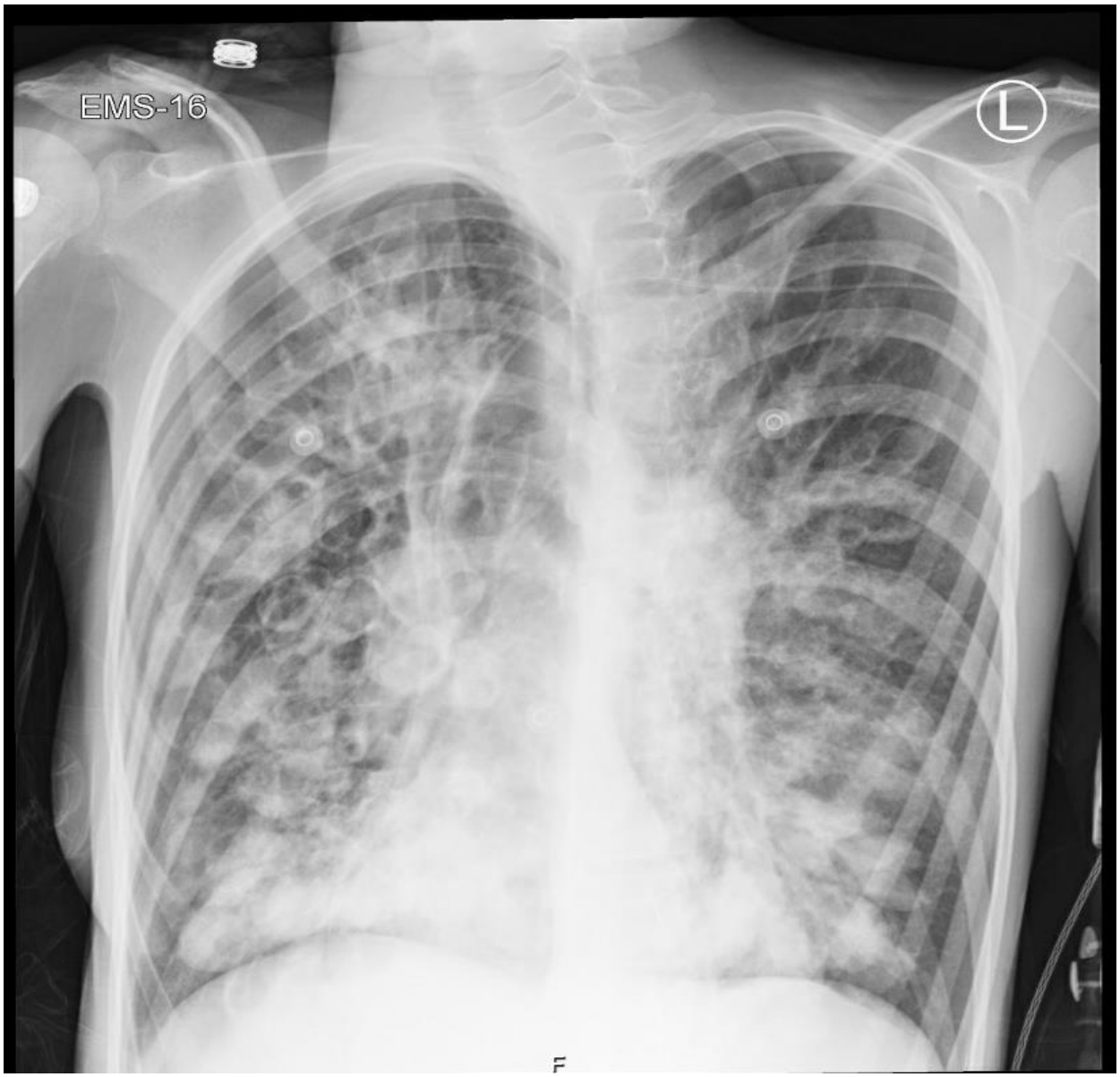

Figure (3): Bilateral diffuse bronchiectatic changes. Right lower lobe cardiophrenic airspace opacity, related to mucous filling the bronchiectatic cavities. Right upper lobe ground-glass patchy opacity due to fibrosis.

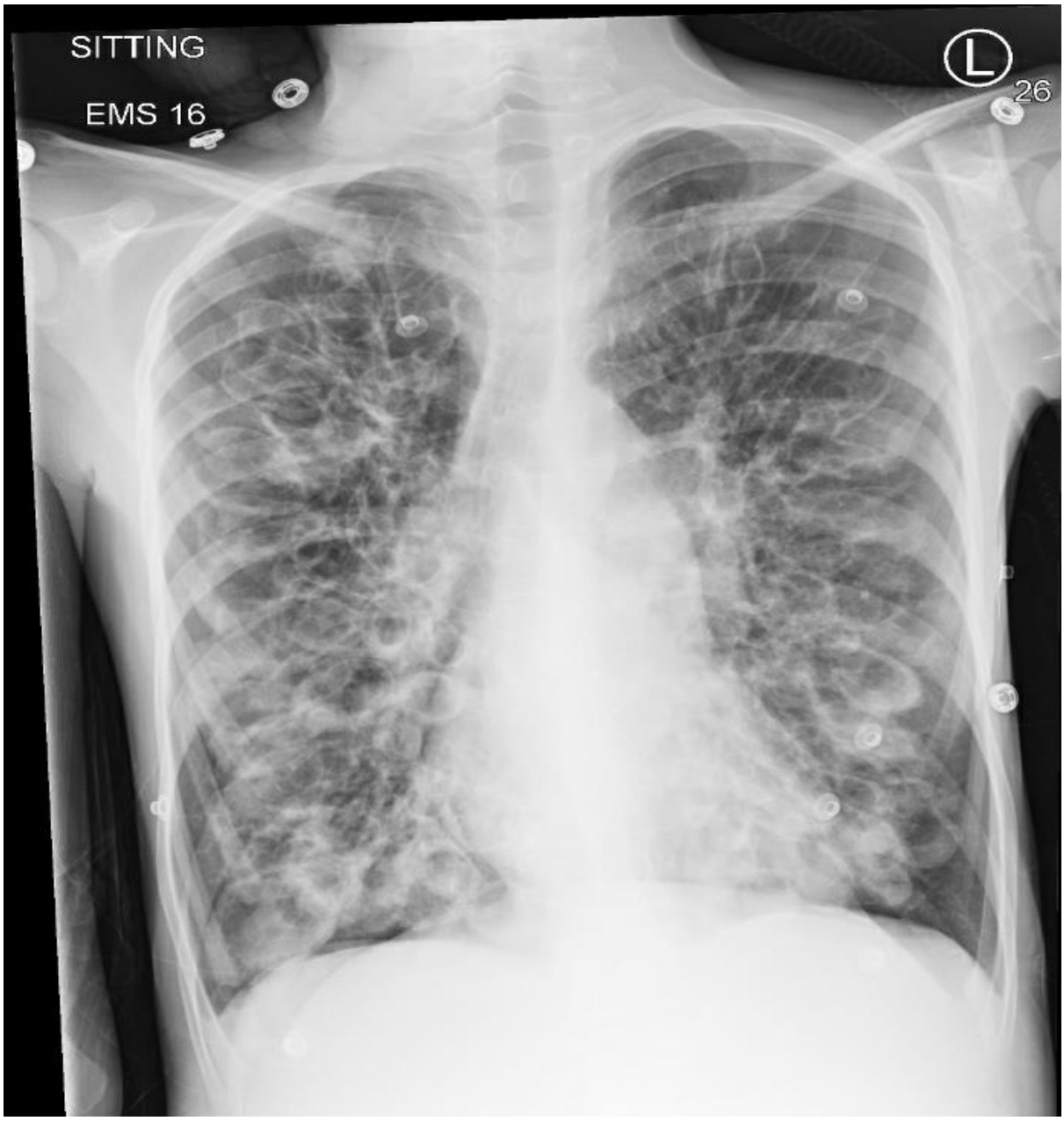

Figure (4): Bilateral diffuse bronchiectasis with fibrotic streaks.

ETHICAL CONSIDERATIONS: All data were stored in the Pediatric Research Unit, accessed only by the Principal Investigator and the assigned Assistant Clinical Research Coordinators. The patient's information was kept strictly confidential. Each patient was given a study number, and all patient data were entered into the designated data sheet (EXCEL) without any patient identifiers. The data that were collected from this study was electronically entered into a database. The Declaration of Helsinki and GCP guidelines have been followed.

STATISTICAL STATEMENT: The Department of Biostatistics, Epidemiology and Scientific Computing (BESC) carried out statistical analysis of the data utilizing the appropriate techniques.

Results

A total of 229 confirmed CF patients. The most common radiological findings at presentation were: Diffuse infiltrate in 70 (30.6%), hyperinflation in 49 (21.4%), bronchiectasis in 44 (19.2%), fibrotic changes in 9 (3.9%), bronchial wall thickening in 136 (59.4%), nodular changes in 7 (3%), cystic changes in 9 (3.9%), interstitial changes in 33 (14.4%), atelectatic changes in 38 (16.6%), and increased vascularity in 10 (4.8%). (Table 1)

Chest X-ray changes	Score # (%)									
Severity	0		1		2		3		4	
	n	%	n	%	n	%	n	%	n	%
Diffuse Infiltrate	159	69.4	6	2.6	28	12.2	8	3.5	28	12.2
Hyperinflation	180	78.6	5	2.2	-	-	2	0.9	42	18.3
Bronchiectasis	185	80.8	4	1.7	18	7.9	4	1.7	18	7.9
Fibrotic changes	220	96.1	3	1.3	2	0.9	-	-	4	1.7
Bronchial wall thickening	93	40.6	13	5.7	55	24.0	13	5.7	55	24.0
Nodular changes	222	96.9	-	-	3	1.3	-	-	4	1.7
Cystic changes	220	96.1	3	1.3	4	1.7	-	-	2	0.9
Interstitial Changes	196	85.6	5	2.2	12	5.2	8	3.5	8	3.5
Atelectatic changes	191	83.4	7	3.1	26	11.4	2	0.9	3	1.3
Increased Vascularity Changes	219	95.6	4	1.7	5	2.2	-	-	1	0.4
Final Score	27	11.8	33	14.4	77	33.6	72	31.4	20	8.7

0 = normal, 1 = mild, 2 = moderate, 3 = severe, 4 = most severe

Table 1: Brasfield scoring of Chest X-ray changes of cystic fibrosis patients (Total= 229 Patients)

Applying Brasfield score (BS): Twenty-seven (12%) had normal lungs, 33 (14%) had mild changes with BS of 10 points, 77 (34%) had moderate changes with BS of 15 points, 72 (31%) had severe changes with BS of 20 points, 20 (9%) had very severe changes with BS of >21 points with complications as pneumothorax and empyema. Sixteen out of the 92 patients (17%) with BS >20 required lung transplants, 2 of the recipients died post lung transplant. (Table 1), (figures 1-4).

Discussion

Previous studies have shown that anatomical and physiological differences result in cystic fibrosis progressing at different rates at different regions of the lungs **[**16**]** The upper lobes and most commonly the right upper lobe being the most severely affected **[**17**]**. In contrast, the proximity of the lower lobes to the diaphragm results in a greater ventilatory excursion which promotes the mobilization of sticky mucus from the bronchi in these lower regions**[**16**]**.

In our study chest x-rays were obtained for 229 cystic fibrosis patients at presentation. After applying the Brasfield score a majority of 155 out of 229 (65%) of patients had moderate or severe changes indicating that they had moderate to severe disease based on radiological findings. 14% had the mild disease while 9% had very severe disease. (Table 1)

Common radiological findings included diffuse infiltrates, hyperinflation, bronchiectasis, bronchial wall thickening and atelectasis – all hallmarks of the CF lung. Hyperinflation of the lungs due to obstruction of the small airways leads to air trapping **[**18**]**. Increased incidence of bacterial infection leads to bronchial wall thickening which then progresses to bronchiectasis**[**18**]**. Since bronchiectasis is considered one of the earliest irreversible structural abnormalities detected by imaging it is even seen in asymptomatic infants identified by newborn screening **[**19**]**.

Our study showed the combination of moderate to severe disease that includes fibrotic changes as a healing process in 6 (2.6%), nodular changes in 7 (3%) and cystic changes in 6 (2.6%). A total of 19 (8.25%) (Table 1). Which are considered permanent changes of CF disease that may progress gradually to end-stage lung disease if not treated properly and at the appropriate time.

Furthermore, our study showed that 77 (34%) already had moderate changes while 72 (31%) already had severe changes on their chest x-rays on the first presentation. This illuminates the issue that many CF patients present to our CF center with already advanced disease due to delayed referral to the CF center and poor knowledge of pulmonary CF disease in the community. This leads to the low median survival of 22 years at our center compared to the 45 years reported in European and North American studies **[**20**]**. Other factors may also have contributed to the progression of their CF diseases such as CFTR mutations and poor compliance to medications **[**21**]**.

Chrispin and Norman [2] described their structured methods of semi-quantifying the morphological features that are commonly seen in CF patients by chest radiograph, but the illustrations for their scoring system are limited [22].

Chest x-ray has often been replaced by computed tomography (CT) at specialized centers, because of its higher sensitivity for early and subtle changes in the CF lung [19]. However, the use of CT for short-term follow-up in infants and preschool children as well as lifelong longitudinal monitoring is accompanied by an accumulation of radiation dose [19]. Most recently, magnetic resonance imaging (MRI) has emerged as a radiation-free technique for assessing the CF lung [19]. Besides morphological information comparable to CT, MRI can depict several components of lung function, i. e. respiratory movements, ventilation and perfusion. CXR, CT and MRI each have intensively studied individual strengths and drawbacks. The risk of sedation in preschool children and allergies against MRI contrast material must be weighed against the risk from radiation exposure [19]. MRI's capability for combined morphological and functional imaging at sufficient spatial and high temporal resolution to obtain information on regional lung function should be taken into account as well. To appreciate its advantages over CT, a perfusion study, which is available on most state-of-the-art MRI scanners already, should be included in the MRI protocol [19].

Our study is limited as it was a retrospective study as we didn't have any control over the internal radiologist's differences and did not establish criteria for chest x-ray interpretation, but it was usually read by 1-2 radiologists that have the internal agreement of similar interpretation criteria.

Conclusion

Chest radiography is an important tool to evaluate the severity of pulmonary involvement and to institute proper treatment early to prevent the progression of the disease. More than 40% of CF patients at our center had severe x-ray changes with Brasfield score >20 points that may require lung transplantation or may have progressive disease.

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