IMPLEMENTING THE BRODY SCORING SYSTEM IN THE EVALUATION OF SEVERITY OF PULMONARY CHANGES IN CYSTIC FIBROSIS ON HIGH RESOLUTION COMPUTED TOMOGRAPHY

Sonja Nikolova
Institute of Radiology, Faculty of Medicine, University "Ss. Cyril and Methodius",
Skopje, North Macedonia, sonikmk@gmail.com

Abstract:.This study aims to perform an in- depth evaluation on High Resolution Computed Tomography (HRCT) exams in patients diagnosed with cystic fibrosis(CF), taking into consideration the gravity and prevalence of lung parenchymal findings by implementing the Brody Scoring System (BSS).
We assessed and scored the HRCT exams of 11 patients diagnosed with cystic fibrosis with the mean age of 13, 5 (4 to 23 years old), by using the Brody Scoring System. All 11 patients underwent a HRCT according to appropriate diagnostic protocol on a 64- slice Somatom Definition AS+ Computed Tomograph (Siemens Healthineers, USA) on which we graded the prevalence, severity and extent of several findings in all 6 lobes in central and peripheral locations, including air trapping, peribronchial wall thickening, mucous plugging, bronchiectasis and the most common parenchymal findings like ground glass opacities, consolidation and cysts.
Considering the habitual nature of the disease, bronchiectasis, mucous plugging and peribronchial wall thickening were the most consistent findings, whereas the others were less frequently encountered. The highest Brody score was affiliated with bronchiectasis and mucus plugging. There was a significant statistical difference between variables and predominant location only with mucous plugging and peripheral lung zones and common parenchymal findings were usually located in the upper right lobe, followed by the lower lobes on both sides.
High Resolution Computed Tomography has an important diagnostic role in the timely assessment of prevalence, severity and extent of a spectrum of lung parenchymal changes in cystic fibrosis, whereas by using the readily available semi- quantitative scoring systems, gives an accurate estimation of the clinical status of the patients and predicts complications and unwanted outcomes.
Keywords: cystic fibrosis(CF); Brody Scoring System(BSS); High Resolution Computed Tomography(HRCT); air trapping; mucus plugging; bronchiectasis.

1. INTRODUCTION

Cystic fibrosis (CF) is a fatal, perennial genetic disorder in children of Caucasian populations, with an incidence of 1/2500 live births, with a still unrevealed specific cause for a genetic mutation of the CFTR gene (1-6). In recent years the clinical spectrum and severity of symptoms is more diverse, ranging from initial malabsorption to recurrent lower respiratory tract infections, fatty diarrhea or failure to thrive. Most patients suffer fatal outcome due to respiratory complications. Although cystic fibrosis continues to be a serious health risk, recent treatment and management breakthrough resulted in prolonged survival of these patients, with a median age close to 50 years. In 1938, when CF was first described by Dorothy H. Anderson, patients didn't survive their first years of life, but recently in developed countries, the fraction of adult patients exceeds that of children, resulting in higher survival rates (7- 13). Chest x- ray and computed tomography are considered the most valuable and frequently used diagnostic modules in the systematic follow- up of patients with cystic fibrosis since early childhood. Chest CT is a diagnostic gold standard for monitoring the progression of bronchiectasis and detecting air trapping which is defined as a regional low attenuation and reduced perfusion. These CT findings have a positive correlation with the severity of the disease and even more so, they appear before the decline of the pulmonary functions, thus proclaiming the CT as a valuable tool for early detection of developing lung damage (14, 15).
There isn't any study alike conducted in North Macedonia, so we aimed to implement a comprehensive assessment of the severity and prevalence of late lung parenchymal changes in patients diagnosed with cystic fibrosis (CF) by administering the Brody Scoring System (BSS) on High Resolution Computed Tomography (HRCT) exams.

2. MATERIALS AND METHODS

We analyzed and scored the HRCT scans of 11 patients diagnosed with cystic fibrosis with the mean age of 13, 5 (4 to 23 years old), by using the Brody Scoring System. All 11 patients underwent a HRCT according to appropriate diagnostic protocol on a 64- slice Somatom Definition AS+ Computed Tomograph (Siemens Healthineers, USA) on which we graded the findings of air trapping, peribronchial wall thickening, mucous plugging, bronchiectasis and the most common parenchymal findings like ground glass opacities, consolidation or cysts, in peripheral and central locations in all 6 lobes of the lungs. HRCT or thin- section CT, refers to a thin slice technique and high frequency

reconstruction algorithms that are used to create comprehensive images of the lung parenchyma with a near isotropic resolution (16, 17). We executed all scans in deep inspiration, including all lung fields from the apex to the base. We didn't perform any expiratory scans due to the ionization exposure as well as the lack of cooperation with most of the young patients.

The 2006- revised Brody Scoring System is a semi- quantitative assessment tool of the prevalence, distribution and extent of bronchiectasis, peribronchial wall thickening, mucous plugging, air trapping and parenchymal involvement in all six lung lobes, on central and peripheral location, by giving them an appropriate score. The BSS score value is ranging from zero to the maximum score value of 243. The HRCT Brody Scoring System (Alen, S. Brody, March 2006) is calculated for all six lobes, using the following prototype (18):

1. Bronchiectasis= (extent in central lung+ extent in peripheral lung)x average size multiplier (range 0- 12); where the average bronchiectasis size is calculated by the sum of the size of largest dilated bronchus+ the average size of dilated bronchi+ 2.
2. Peribronchial wall thickening= (extent in central lung+ extent in peripheral lung)x severity (range 0- 9);
3. Mucous plugging= extent in central lung + extent in peripheral lung (range 0- 6);
4. Air trapping= extent of air trapping x appearance of air trapping (range 0- 4.5);
5. Parenchymal involvement score= extent of dense opacity+ extent of ground glass opacity+ extent of cysts or bulla (range 0- 9).

The extent of all the variables is calculated by giving them an appropriate score: 0= none; 1= 1/3 of a lobe; 2= 1/3 to 2/3 of a lobe and 3= more than 2/3 of a lobe.

3. RESULTS

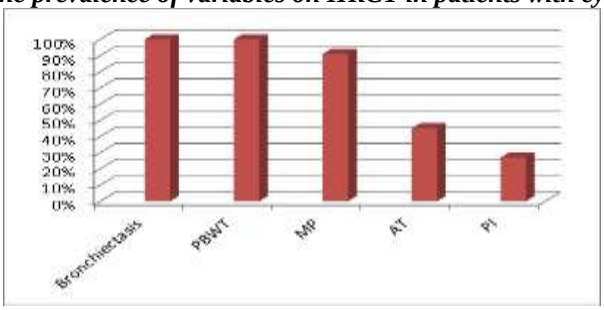
Out of 11 patients that we analyzed, eight (72.7%) were male and three (27.3%) female. The mean CT score for all 11 patients was 75.5± 54.6. Bronchiectasis and peribronchial wall thickening were the most prevalent with 100%, whereas mucous plugging was found in 91%, air trapping in 45% and parenchymal changes in only 27% of all the patients (Table 1, Figure 1). From the total Brody Severity Score of 820 calculated in all 11 patients, bronchiectasis were the most prevalent with a maximum score of 298, peribronchial wall thickening with a score of 241.5, mucous plugging of 196, air trapping 70.5 and the parenchymal involvement significantly less in comparison to the previous, with the score of only 14 (Table 1, Figure 2, 4a-f).

Table 1. Brody Scores of different variables in all six lobes of patients with cystic fibrosis

	URL	ULL	MRL	LINGULA	LRL	LLL	TOTAL SCORE
BRONCHIECTASIS	67.5	64.5	64.5	43.5	29	29	298
PERIBRONCHIAL WALL THICKENING	42.75	42.75	46	46	32	32	241.5
MUCUS PLUGGING	37	34	37	38	25	25	196
AIR TRAPPING	12.5	11	10.5	9.5	13.5	13.5	70.5
CONSOLIDATION OR GROUND GLASS OPACITY	2	3	2	3	2	2	14
TOTAL SCORE	161.75	155.25	160	140	101.5	101.5	820

URL- upper right lobe, ULL- upper left lobe, MRL- medial right lobe, LRL- lower right lobe, LLL- lower left lobe.

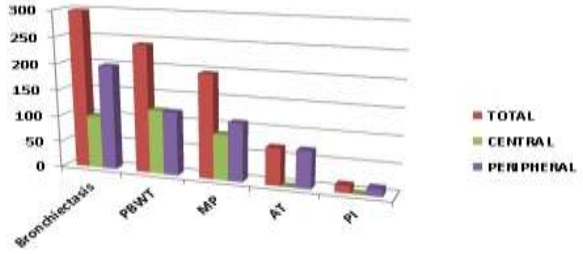
Figure 1. The prevalence of variables on HRCT in patients with cystic fibrosis



PBWT- peribronchial wall thickening, MP- mucous plugging, AT- air trapping, PI- parenchymal involvement.

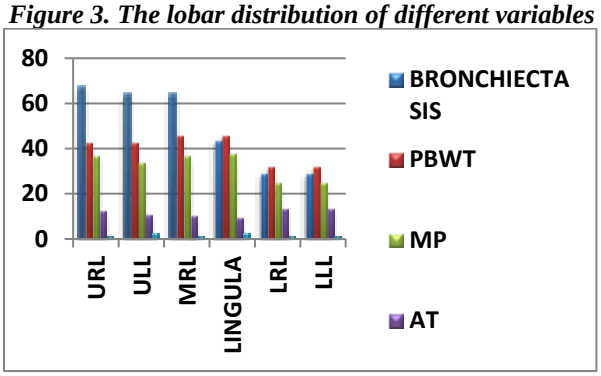
Considering the total score of 820 and subscores by lobar distribution, the procession is as follows, upper right lobe(URL): 67.5> upper left lobe and middle right lobe(URL and MRL): 64.5> lingula: 43> and lower right and left lobes(LRL and LLL):29, which didn't exhibit a significant difference. Bronchiectasis, mucous plugging and air trapping were more frequent in peripheral regions of the lungs (Figure 2 and 3).

Figure 2. The zonal distribution of different HRCT findings



PBWT- peribronchial wall thickening, MP- mucous plugging, AT- air trapping, PI- parenchymal involvement**.**

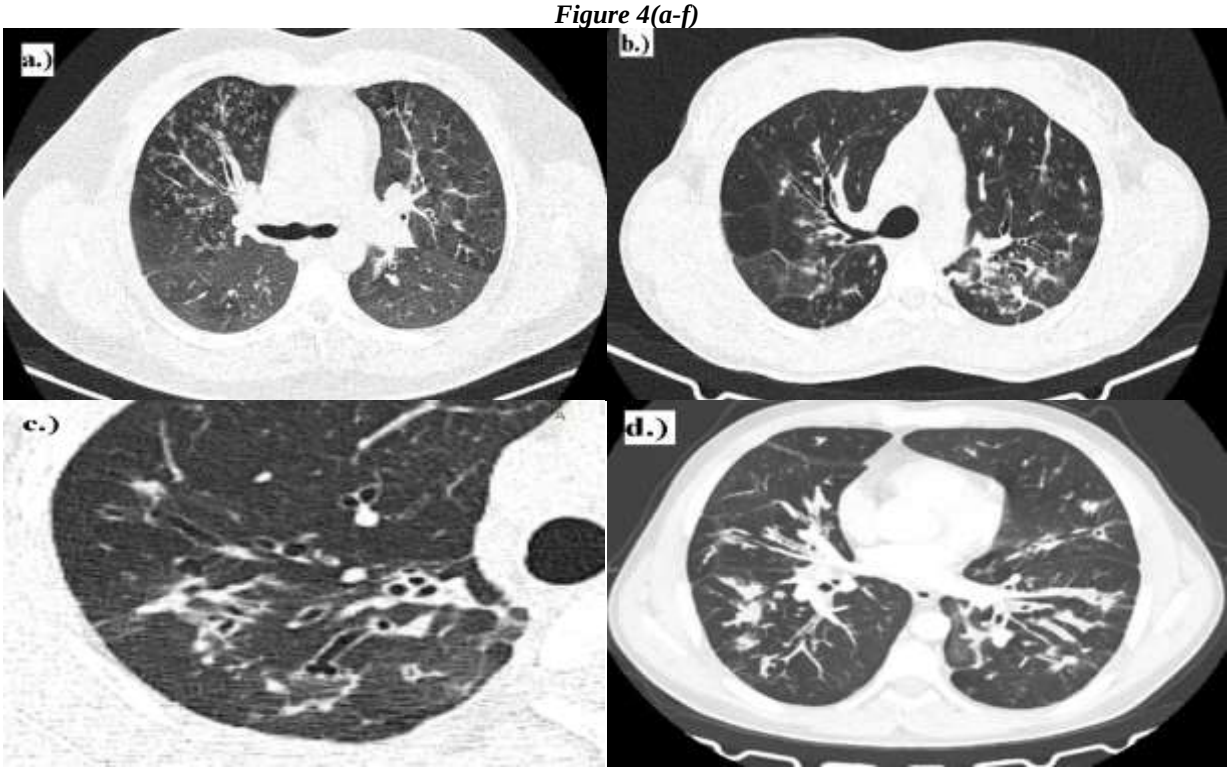
Figure 3. The lobar distribution of different variables

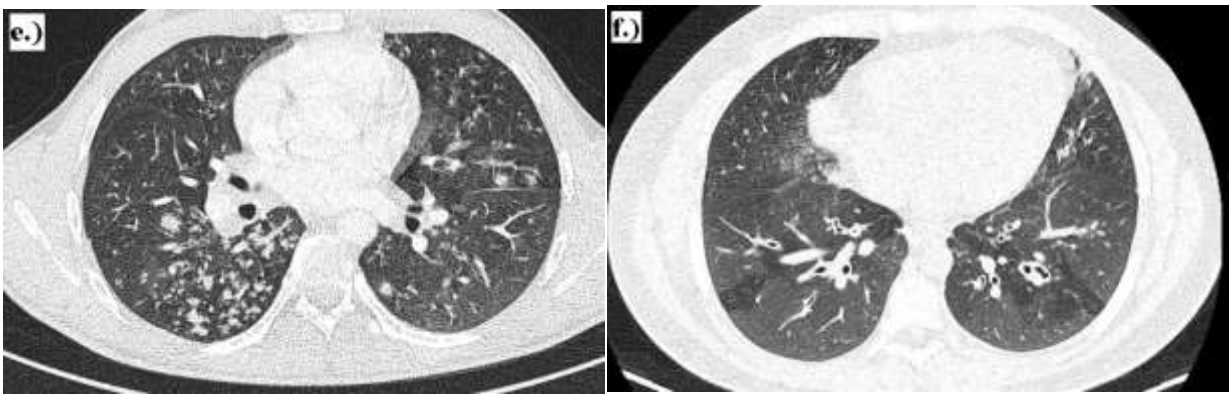


PBWT- peribronchial wall thickening, MP- mucous plugging, AT- air trapping, PI- parenchymal involvement, URL- upper right lobe, ULL- upper left lobe, MRL- medial right lobe, LRL- lower right lobe, LLL- lower left lobe.

Bronchiectasis, mucous plugging and peribronchial wall thickening were most frequent in the upper and middle lobes on both sides, whereas the air trapping was commonly found in the lower lobes on both sides as well as the upper right lobe (Table 1, Figure 2).

Figure 4(a-f)





a) peribronchial wall thickening, mucous plugging and air trapping in the upper right and left lobes; b) extensive air trapping due to bronchiectasis and peribronchial wall thickening in the upper right and left lobes; c) magnified image of bronchiectasis and peribronchial wall thickening; d) bronchiectasis and mucous plugging in the middle and inferior lobes on both sides; e) mucous plugging and air trapping in the middle and inferior lobes on both sides, more accentuated on the right side; f) air trapping and peribronchial wall thickening in the lower lobes on both sides.

The summary of all HRCT findings by lobar distribution and their scores is given in detail in Table 1 and Figure 3.

4. DISCUSSION

CF is recognized as common and fatal genetic disease among Caucasians (1-4). Since there are no early screening initiatives in most of the developing countries, the diagnosis is delayed until older age, when patients are referred to hospitals for recurrent pneumonias that serve as the main reason for extended parenchymal involvement and irreversible lung lesions such as bronchiectasis. Parenchymal involvement is most frequently noted in central and upper lung regions. In this study of 11 patients diagnosed with cystic fibrosis, there was no statistically significant zonal or lobar involvement. The bronchiectasis and peribronchial wall thickening were most prevalent in the upper and middle lobes, whereas the mucous plugging in lingula, upper and middle lobes. The air trapping was most commonly encountered in the lower lobes, and parenchymal involvement presented by consolidation and ground glass opacity in the left upper lobe. All variables were more common in the peripheral regions of the lungs, except the peribronchial wall thickening which was equally distributed in central and peripheral lung regions.

Almost identical results like ours, concerning the gravity and prevalence of bronchiectasis and PBWT in all 6 lobes, were published in a study by Bhalla et al. that included 14 patients with CF between 4 – 10 years old (19). There was no HRCT finding suggesting an exacerbation by atypical pulmonary infection in all 11 patients, and parenchymal involvement consisted of mostly cystic lesions and ground glass opacities, which didn't necessarily exclude it. The air trapping as a finding didn't offer authentic results since all the exams were performed in deep inspiration only, due to the lack of cooperation with the younger patients and the inability to perform a comparative expiratory exams.

5. CONCLUSION

High Resolution Computed Tomography is an important diagnostic tool that aids the timely assessment of prevalence and gravity of a spectrum of common, late lung parenchymal changes in patients diagnosed with cystic fibrosis, whereas by using the readily available, semi- quantitative scoring system of Brody, we can achieve accurate estimation of patient clinical status and predict any complications and unwanted outcomes.

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