CASE REPORT

Critical analysis of scoring systems used in the assessment of cystic fibrosis severity: state of the art*

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This study carries out a descriptive and comparative analysis of the various types of cystic fibrosis severity scores described in the literature and contextualizes the origin and objective of each. A total of 16 scoring systems were found: 8 are used predominantly for clinical evaluation, 5 for radiographic findings, 2 for tomographic findings and 1 for scintigraphic findings. Despite the criticism and controversy regarding these instruments of assessment, they have contributed to a better understanding of the disease and to the development of more effective therapeutic procedures.

Key words: Severity of illness index. Cystic fibrosis. Review literature.

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Abbreviations used in this paper:
CF – Cystic fibrosis
CFSS – Cystic fibrosis severity scoring
CT – Computed tomography
FEV_1 – Forced expiratory volume in one second
HRCT – High-resolution computed tomography

INTRODUCTION

Cystic fibrosis (CF), sometimes referred to as mucoviscidosis, is a lethal, genetic, autosomal recessive disorder. It is more prevalent in Caucasians, in whom approximately 1000 mutations have been identified. The defect affects a protein known as cystic fibrosis transmembrane conductance regulator, causing an increase in the viscosity of gland fluids and a decrease in mucociliary clearance[1]. Clinical manifestation of the disease varies among patients, depending on the severity of pulmonary and gastrointestinal involvement. Pulmonary involvement is the most common cause of mortality[2]. Progressive pulmonary deterioration may be caused by inflammation, mucus accumulation, chronic infection by *Staphylococcus aureus* and *Pseudomonas*, bronchial hyperresponsiveness or hyperinflation, as well as obstruction or instability of the airways[3]. There is no consensus on the ideal antibiotic therapy for the treatment[4], but controlling pulmonary infections is important in the management of the disease, as well as for improving bronchial fluid clearance, reversing poor absorption of nutrients and providing nutritional support[1]. Individual characteristics of each patient are also taken into consideration when devising therapeutic strategies because, as has been reported since the first studies on the disease, treatment adherence and disease severity are important prognostic factors[5,6,7,8].

In recognition of the fact that CF is so frequently seen and its clinical manifestation so variable, scoring systems designed to assess CF severity have been developed. These systems have contributed to the characterization and evaluation of the course of the disease, the portrayal of the history of the disease and the determination of phenotypic differences among populations. These scores have been elaborated through the analysis and comparison of signs and symptoms found in patients[9], and the development of each system was directly related to the scientific understanding of the disease. Cystic fibrosis severity scoring (CFSS) has been used to evaluate the extension of lung damage, compare clinical severity, evaluate the effects of therapeutic interventions and estimate prognosis. However, there is no consensus as to the ideal score[10,11,12]. Since clinical, radiography and computed tomography (CT) scores are all valued and widely used in CF health care centers, the present study was designed to analyze these various scoring systems (Chart 1).

This study carried out a critical, descriptive and comparative analysis of the various types of CFSS through a review of the literature using the MEDLINE database.

SCORING SYSTEMS

Through a review of the literature, we identified 16 CFSS systems: 8 are used for clinical evaluation, 5 for radiographic findings, 2 for tomographic findings and 1 for scintigraphic findings. Each CFSS system was related to the historical moment of scientific understanding of the disease and aimed to assess CF severity and evaluate therapeutic interventions.

The first case reports and studies on CF identified mortality in newborns[13,14], the idea of which was modified by evidence from later studies. McIntosh[15] carried out the first prospective study, observing patients until they were 10 years old in order to relate disease evolution to disease severity. Subsequently, Royce[16] elaborated the first set of disease assessment clinical parameters, which included only digital clubbing, cough, respiratory frequency and exercise tolerance. The author emphasized only the symptoms and did not related them to disease severity or to the extent of lung damage[5].

The question of disease severity and the need for a system of clinical evaluation was emphasized by Shwachman[5] in a longitudinal study comprising 105 CF patients who were monitored for 5 years after diagnosis. The author reported that, in these patients, regardless of whether diagnosis was early or delayed, there was wide variation in degree of disease severity. The same author also stressed the importance of pulmonary function testing in the assessment of the disease. This clinical evaluation system (Shwachman score) was a milestone in the scientific history of CF and is still respected and used as a classical tool for the assessment of CF disease severity[7,8,11,12]. The scoring system was created in

order to compare clinical manifestations among patients, detect treatment effects and aid in the determination of diagnostic criteria. To that end, the system evaluates 4 major parameters: general activity, nutrition, chest radiographic findings and physical examination (Chart 2). The score for each parameter ranges from 5 to 25. The lower the score, the more severe the disease.

The Shwachman score received some criticism due to its subjectivity and, in 1964, Doershuk et al. modified it and proposed a new score that would be more objective and could be also used for adolescents and adults[6]. The authors still used the 4 criteria recommended by Shwachman, adding only some aspects related to adolescents and adults. A 5-point range was suggested for the final score, and the authors recommended that double-blind trials be conducted and that the examiners discuss controversial cases. The authors also monitored clinical manifestations in patients under treatment for 5 years. Since there was no improvement in the scores of patients who presented with poor radiographic findings, even after the introduction of a medication regimen, a new issue was identified: the importance of early treatment of pulmonary manifestations.

In 1971, inspired by criticism of the existing scores, Cooperman prepared a simplified scoring system based on the Apgar test model and known as the Simplified (Cystic Fibrosis) Scoring Scale (SCS). The objective of the score is to show the effects of medication and treatment on CF management (Chart 3). The score comprises 5 parameters: growth and development, general activity, chest radiographic findings, digital clubbing and complications. Patients receive 0,1, or 2 points depending on clinical profile. Patients suffering from the severe form of the disease score 0, whereas those who present with no involvement may score as many as 2 points in each category, up to a maximum of 10 points[7]. The authors periodically evaluated 45 patients for a year and a half and admitted having limited experience with the use of this scoring system.

In 1973, Taussig created the National Institutes of Health (NIH) score, a clinical scoring system for the assessment of CF severity which was intended to be objective and was defined by the authors as simple, easily implemented and immediate. In order to test this scoring system, the authors evaluated 73 patients, ranging from 5 to 30 years of age, for a period between 3 and 6 years. Common complications such as pneumothorax and hemoptysis, as well as surgical complications, were considered in the prognosis. The scoring system included general and pulmonary evaluations. Sub-categories with specific scores were defined, so that a patient suffering from severe CF could score up to 100 points. Detailed clinical data, such as blood gas analysis results, gastrointestinal involvement and arthropathy were also considered during the evaluation process but did not affect the score (Chart 4). Adherence to treatment was also related to the severity of the disease so that physicians and family could have a numeric value predictive of life expectancy, improving their participation in multiprofessional care[8].

A year later, Chrispin and Norman created the first radiographic scoring system for the assessment of CF severity, considering the fact that changes in radiographic findings portray the progression of the pulmonary disease and may be related to the clinical condition of patients (Chart 5). The scoring system was based on the systematic analysis of radiological scans, and the authors created uniform terms, describing 5 radiographic characteristics. Evaluation was based on the division of the chest X-ray into 4 zones and on the classification of images into 3 categories depending on the presence and severity of alterations[17]. The validation of this scoring system was made by 2 examiners who evaluated chest X-rays of 30 pediatric CF patients and compared those to chest X-rays of the same children 20 months later.

Although there were several CFSS systems in use, there was still some controversy and debate regarding the variability of the severity of clinical manifestations, the relationship between radiologic changes and pulmonary disease, and the efficacy and effectiveness of the existing scoring systems. Therefore, in 1979, Brasfield et al. created a new, radiographic scoring system, the Brasfield score[10]. Three experts, a pediatrician and two general physicians, each separately evaluating 643 radiographs from 118 patients, demonstrated the efficacy of this scoring system. This study was also designed to confirm

the system reproducibility and its relationship to clinical parameters and pulmonary function test results. The Brasfield score comprises 5 categories, scored from 0 to 5. Therefore, the most severe radiographic alterations receive a maximum score of 25 points (Chart 6).

In 1980, Piepszi et al., motivated by the creation of seven scoring systems for clinical evaluation and radiographic findings, as well as their interest in early diagnosis of, and increasing survival for, CF patients, suggested the creation of a scoring system based on scintigraphic findings[18]. Two examiners evaluated 285 scintigraphs from 111 patients ranging from 2 months to 20 years of age. Each lung was considered in 3 sections and each section received from 0 to 2 points depending on the severity of changes. A total score of 12 points signified extensive involvement in the perfusion. Although this study highlighted the importance of scintigraphic evaluation, a detailed system of quantification was not described.

In the following year, due to the fact that great attention was being given to clinical parameters in CF, Huang et al.[19] created a scoring system that took into consideration the failures of previous CFSS systems[5,8]. The system evaluated the therapeutic response of various antibiotic regimens used at that time[20,21]. Two of the authors tested the scoring system by using it to evaluate 22 CF patients. The study was proven to be statistically consistent and reliable. The Huang score includes 20 items (worth 5 points each). There are 10 clinical, 5 radiographic and 5 pulmonary function parameters (Chart 7). The lower the score, the more severe the disease. The system also takes into consideration 5 typical complications of the disease, graded according to their severity (5 points for mild and 10 for severe complications). What makes this scoring system unique is the fact that it is applied prior to and after therapy in order to identify improvement or worsening of the clinical profile in relation to specific therapeutic interventions[19].

In 1982, Van der Put et al. [22] proposed a modification to the Chrispin & Norman[17] scoring system, adding 4 extra categories to the assessment of images of hilar enlargement. This scoring system, known as the CN score[22], was created to relate age and the onset of typical CF radiographic signs, to assess the disease, and to compare scores between CF patients and patients with obstructive pulmonary disease or children with probable cardiopathy. This comparison between scores did not demonstrate that characteristics such as hyperinflation and linear images were specific for CF, as demonstrated in the case of annular images and condensations.

In 1987, Lewiston et al.[23] proposed a new adaptation based on the pre-existing scoring systems. This new scoring system included the Brasfield[10] radiographic scoring system as a substitute for that of the Shwachman score[5] and was called the S-B score. It was developed at a time when studies, drug trials and specialized institutions employed the existing scoring systems and the variations in the evaluation of these scoring systems among specialists were being questioned. This new CFSS system was developed for use in multicenter studies, and the reproducibility of its criteria was confirmed by the small variation in the scores given by the 5 physicians who evaluated the 41 CF patients[23].

Despite the systematic use of clinical[5,6,8] and radiographic[17,10] scoring systems, as well as pulmonary function tests and sputum cultures, the first scoring system using findings from high-resolution computed tomography (HRCT) was developed. It was called the Nathanson scoring system and was created in order to provide more precise data in pulmonary evaluation and to determine the condition and site of bronchiectasis and mucoid impaction in the airways of CF patients[24]. The system was developed by two radiologists and a pediatric pulmonologist through analysis of the HRCT scans of 28 CF patients in a blind trial. The examiners also made use of the Shwachman[5] and Brasfield[10] scoring systems, as well as the results of pulmonary function tests of 19 patients. In this scoring system, chest HRCT scans are scored through dividing the lungs into 12 zones and scoring each zone separately. Bronchiectasis is scored between 0 and 5, according to its severity, and the presence of impacted mucus adds another point to the score. High scores correspond to high severity.

Bhalla et. al.[25] delved further into the use of CT in the assessment of this pulmonary disease. Because the authors considered radiographic scoring systems to be imprecise and subjective, they also developed a tomographic scoring system. Their system was designed to assess pulmonary involvement, determine therapeutic effects and aid selection of patients for transplants. The CT scans of 14 patients, between 5 and 42 years of age, were retrospectively studied by 3 radiologists using the morphologic quantification technique proposed by the authors. This scoring system, called the Bhalla[25] score, presented significant and positive results, evidenced by the small variation in the scores given by the various examiners. In addition, the system proved to have good reproducibility and high correlation with pulmonary function test results. A total of 9 categories, worth 3 points each, are scored, and a maximal score equals a high degree of severity. The final score must be subtracted from 25. The lower the result, the more severe the condition (Chart 8).

Advances in the diagnosis and treatment of CF and the lack of a truly sensitive and reproducible radiographic evaluation system for the quantification of the pulmonary disease led to the development of another scoring system, called the Wisconsin scoring system[26]. This radiographic scoring system was structured into 3 phases: attribute and scoring system elaboration; comparison with the Brasfield scoring system[10]; and validation. It comprises 6 main attributes and a total of 24 individual components with specific scores, ranging from 0 to 100 points (Chart 9). Patients with more severe manifestations of the disease have higher scores[26].

In 1994, Conway et al. proposed a system that can be performed by a single examiner, the Northern scoring system[27]. Previous radiographic scoring systems required a full medical team in order to score the pulmonary involvement of patients. In the Conway et al. study, 10 clinical physicians evaluated 45 X-rays and compared the new system to other radiographic scoring systems[10,17]. Using the Northern scoring system, there was better agreement among examiners, regardless of the evaluation of X-rays in profile (Chart 10).

Matouk et. al.[29] modified Huang[19] scoring system, detailing some of its items, but maintaining the original structure, because it was necessary to assess the disease in adults and scoring criteria had to be clearer. A study comprising 109 adults diagnosed with CF revealed high consistency and statistically significant correlation between this system and force expiratory volume in one second (FEV_1) [28]. Two years later, this system was shown to be useful in the evaluation of the effects of therapeutic interventions[29].

The most recently developed clinical scoring system is the Cystic Fibrosis clinical score created by Kanga et al.[12], who subsequently conducted a study correlating signs and symptoms of pulmonary function damage in CF patients[30]. According to the authors, this is a system designed to assess acute exacerbations of the disease, to predict improvement or worsening of pulmonary function of patients and to evaluate therapeutic effects. The scoring system is simple, inexpensive and easily applied. This was a prospective, multicenter study comprising 130 patients between 5 and 17 years of age with acute pulmonary exacerbations. It included results of routine evaluation of patients and common clinical variables. The authors demonstrated that the system presented little variability among examiners and correlated significantly with pulmonary function (FEV_1 and forced vital capacity) test results. The system helps identify daily clinical changes and includes 5 common symptoms (cough, fluid secretion, appetite loss, dyspnea and frailty) and 5 physical signs (temperature, weight, respiratory frequency, wheezing and breath sounds). Each criterion is worth 1 to 5 points. The higher the score, the more severe the condition. A drop of 15 points in the score suggests clinical improvement, and the authors recommend hospitalization if there is an increase of 10 to 15 points.

CRITICAL ANALYSIS

Since it is very difficult to establish degree of CF severity, the scientific community must be attentive to CFSS objectives, applications and proposals. Scoring systems are an important tool in the evaluation and management of CF.

Each CFSS system was developed within a specific historical context and in an attempt to fill the needs of that time.

In the 1950s, researchers discussed the need for a clinical and prognostic instrument that would allow comparison among patients, define disease severity and aid in devising therapeutic strategies. McIntosh proposed a simple scoring system based on clinical evaluation[15], but Shwachman[5] created the first elaborate clinical scoring system. The Shwachman score, although the most frequently used in the medical community, is often criticized, mainly because it is subjective, has global criteria and was based on clinical evidence in children. It is also criticized because it does not highlight the respiratory system evaluation, excluding pulmonary function test results and complications due to disease progression from consideration[6-8,11,12]. However, the Shwachman score is still considered important because of its contribution to the understanding of the disease[15,5]. Doershuk adapted the Shwachman score, aiming at higher objectivity and the inclusion of teenagers and adults in the evaluation[6]. However, the Doershuk score is still considered very subjective[7,8], even though it is widely used. The Cooperman score, which is a simplification of the pre-existing scoring systems at that time[7], is seldom used.

Although the subjectivity of these various systems was frequently questioned, and there were calls for a comprehensive and objective clinical scoring system that would bring light to evolution and early treatment modalities[7], survival among CF patients increased, leading to the emergence of new complications. The scoring system developed by Taussig, the NIH score[8], was the only system taking into account these clinical complications and has been recognized as useful in prognosis assessment and disease evolution[32]. It was also an attempt to address the shortcomings of the Shwachman score. However, the NIH score has been criticized for its complexity[33], since it depends on cooperation from families of patients, overestimates some rare clinical elements and ignores others[31], does not include patients younger than 5 years old and does not assess daily changes in the clinical profile of patients[12]. In addition, the NIH scoring system recommends that patients be informed of their prognosis, lacks specificity in the gradation of variables[33,31] and presents high variability in pulmonary evaluation[34], all of which has also been criticized. Nevertheless, Huang[19] considered the NIH score more comprehensive than the Shwachman score.

Huang reported the scoring system used in their practice[19,20,21]. It is considered quite complete and addresses a frequent medical concern: the quantification of responses to various treatments[19,20,21]. In addition, this system evaluates parameters, such as pulmonary function test results, hypercapnic and hypoxic respiratory failure and other pulmonary complications, not taken into account in other scoring systems and has been used in other studies[28,29]. Likewise, Kanga et al.[12] developed a score that takes a preventative approach. The Kanga system is based on the daily observation of signs and symptoms in order to detect acute pulmonary exacerbations in patients, compare therapeutic responses, differentiate disease severity and control respiratory function. Although no studies have been published using this scoring system, it can already be considered very important since it portrays the current state of CF, the scoring systems for which are being reevaluated[11]. In addition, important therapeutic discoveries have been made, and there is a general concern regarding treatment adherence, as well as early diagnosis and intervention.

Some studies have shown that radiographic findings correlate strongly with clinical evidence and pulmonary function test results in CF patients[36,37,38,39]. Therefore, radiographic scoring systems are considered important in the assessment of the disease[35].

Chrispin & Norman[17] created the first specific radiographic scoring system from studies that had already identified the various degrees of pulmonary involvement in CF patients. The authors recommended radiographic testing as an important indicator of evolution of the disease[40,41]. The authors also attempted to simplify the organization of specific, previously defined alterations so that these alterations were compatible with severity and correlated to clinical aspects. However, the system has been criticized since there are no data available on its reproducibility[10,26]. On the other hand, the Brasfield[10] radiographic scoring system has proven reproducible and presented high correlation with pulmonary function test results and prognosis. It has been considered a good resource in CF

management[38]. Both scoring systems have been used in large medical centers and in studies. However, the Brasfield[10] scoring system has been criticized by its lack of flexibility, whereas the severity discrimination of the Chrispin & Norman[27] score has been questioned. Weatherly et. al. [26] considered the Wisconsin score better in the assessment of mild CF than the Brasfield score. The Northern radiographic scoring system is considered simple and practical[35], allowing chest X-ray evaluation to be conducted by a single examiner[27].

Just as Doershuk[6] modified the Shwachman[5] clinical scoring system, some adaptations have also been made to the radiographic scoring systems. Van der Put[22] modified the Chrispin & Norman [17] scoring system for use in their study. Lewiston[23] replaced the radiographic evaluation component of the Shwachman scoring system with that proposed by Brasfield[10].

Although X-rays are inexpensive, are easily performed and expose the patient to a minimum of radiation, the development of CT has allowed better viewing of pulmonary structures and, more recently, early detection of airway alterations[24]. In addition, CT scans are less subjective and imprecise[25]. Two tomographic scoring systems, the Bhalla score[25] and the Nathanson score[24], have been developed in order to achieve detailed analyses of distinct pathological elements that are important in determining the prognosis of the disease. They differ in that the latter involves HRCT, but both are aimed at therapeutic regimens and selection of patients for surgical intervention or transplant.

Piepsz et al. [18] proposed a system based on scintigraphic findings. However, they neither clearly defined scintigraphic criteria nor developed a scoring system. In accordance with other clinical, radiographic and tomographic scoring systems, the study highlighted the importance of analyzing test results together with technological resources in order to characterize respiratory involvement[35,39]. Such analysis allows better understanding of the evolution, control, prognosis and treatment of the disease, elements vital to any CF care center.

Unlike the Huang [19] and Wisconsin [26] scoring systems, most CFSS systems lack guidelines for the application of the system and do not provide scoring grids, important features for reducing variability and increasing reproducibility. Although some CFSS systems seem to be “self-explanatory”, their correct execution seems to be restricted to the centers where they were developed, usually applied by their authors. Recent studies have discussed, studied and confirmed the standardization and validation of the existing CFSS systems.

Despite all the criticism, concern and controversy, CFSS systems are accessible tools that can be easily applied and play an important role in the understanding of CF and its evolution. Scoring systems are predictive of the evolution of the disease and are used in order to establish the speed of progression, identify the clinical profile, estimate intervention needs, detect therapeutic responses and select patients for special, immediate procedures. However, CFSS systems still need to be more precisely defined and more objective, especially in relation to follow-up evaluation of pulmonary function, the deterioration of which is the leading cause of mortality in CF.

CONCLUSION

Although various national and international studies have shown that there is no relationship between genotypic and phenotypic characteristics in CF, clinical characteristics and mutations have been related to disease severity. As we have described herein, the search for clinical and laboratory severity markers has been a constant concern for the past 100 years, advancing in parallel with increased knowledge of CF pathogenesis. This influenced the development of methods for scoring the multisystemic effects of the disease. These methods have been used in the assessment of CF severity. Today, there is better understanding of the disease thanks to half a century spent on study and research. The CFSS systems have been part of this process, characterizing the evolution of the disease, defining severity, indicating the prognosis and measuring the efficacy and the

effectiveness of interventions. The CFSS systems are useful tools in the qualification and quantification of various therapies and represent another available resource for multiprofessional teams in CF therapy centers, allowing them to choose, from among these various systems, the most appropriate system and use it in their routine. In addition, the increased knowledge of the genotypic and phenotypic characteristics of the more than 1000 CF-related mutations, as well as the technological advances in testing (CT, scintigraphy, magnetic resonance, etc.) warrant changes in the existing CFSS systems, as well as the development of new systems in the near future.

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CHART 1

Cystic fibrosis severity scoring systems

	SCORING SYSTEM	YEAR	EVALUATION	REFERENCE
1	Shwachman Score	1958	clinical	
2	Doershuk Score *	1964	clinical	6
3	Simplified Cystic Fibrosis Scale - SCS	1971	clinical	7
4	Taussig Score - NIH	1973	clinical	8
5	Chrispin Norman Score	1974	radiographic	17
6	Brasfield Score	1979	radiographic	10
7	Scintigraphic score	1980	scintigraphic	18
8	Huang Score	1981	clinical	19
9	CN Score *	1982	radiographic	22
10	SB Score *	1987	clinical	23
11	Nathanson Score	1991	tomographic	24
12	Bhalla Score	1991	tomographic	25
13	Wisconsin Score	1993	radiographic	26
14	Northern Score	1994	radiographic	27
15	Matouk Score*	1997	clinical	29
16	Kanga Score - CFCS	1999	clinical	12

* = ADAPTED OR MODIFIED SCORING SYSTEMS

CHART 2
Shwachman score

Score	General activity	Radiographic findings
25	completely normal activity; practices sports; regularly goes to school	clear lung fields
20	lacks resistance and is tired at the end of the day; good school attendance	minimal signs of bronchovascular markings; primary emphysema
15	voluntarily rests during the day; easily tires from exercise	mild emphysema; signs of atelectasis; increased bronchovascular markings
10	satisfactory school attendance; private teacher; rests often; dyspnea after short walks	moderate emphysema; simultaneous diffuse atelectasis and infected areas; minimal bronchiectasis
05	orthopnea; bedridden	extensive obstructive pulmonary alterations and infection; lobular atelectasis and bronchiectasis
Score	**Nutrition**	**Physical examination**
25	Normal weight and height (25^{th} percentile); good tonus and body mass; normal well-formed stools	no cough; normal HR and RR; clear lungs; good posture
20	Weight and height between the 15^{th} and 20^{th} percentile; slightly abnormal stools; satisfactory tonus and body mass	Occasional cough; normal HR and RR at rest; minimal emphysema; clear lungs; no digital clubbing
15	Weight and height above 3^{rd} percentile; frequently abnormal stools – poorly formed; weak tonus and reduced body mass; slight or no abdominal distention	Occasional cough (in the morning); slightly high RR; mild emphysema; rare crackling sounds; early clubbing
10	Weight and height below 3^{rd} percentile; abnormal, greasy, poorly-formed stools; weak tonus and reduced body mass; mild to moderate abdominal distention	Frequent cough, productive in general; chest retraction; moderate emphysema; frequent crackling sounds; digital clubbing (2/3)
05	Pronounced malnutrition; severe abdominal distention; frequent, voluminous, foul-smelling, greasy stools; frequent rectal prolapse	Severe paroxysmal cough, tachypnea and tachycardia; extensive pulmonary alterations; signs of right heart failure; clubbing (3/4)

Classification	Score
severe	< 40
moderate	55-41
mild	70-56
good	85-71
excellent	100-86

HR: heart rate; RR: respiratory rate
(Charts organized from the original article by Shwachman, 1958)

CHART 3

Cooperman score

CATEGORIES	2	1	0
Activities	Normal upon physical exertion; Engages in typical physical activities	Regular school attendance (maximum, 2 absences/month)	
Chest X-ray	Normal	Slightly increased markings; emphysema	
Digital clubbing	0 to 1+	1 to 2+ (no cyanosis)	2+ (extensive)
Growth and development	height and weight above the 25th percentile	height and weight above the 3rd percentile	height and weight below the 3rd percentile
Complications	none	transitory	perpetual

(Chart organized from the original article by Cooperman, 1971)

CHART 4

Taussig (NIH) score

LUNG	SCORE
X-ray	01–17
Pulmonary function	01–17
Pulmonary exacerbations	03–05
Pneumothorax	03–05
Hemoptysis	04–07
Pulmonary surgery	02–07
cor pulmonale	03–05
Lung auscultation	01–09
Cough and expectoration	01–03
TOTAL	**75**
GENERAL	SCORE
Weight	01–06
Activity	01–10
Attitude	01–09
TOTAL	**25**
COMPLICATION	CHARACTERISTIC
Blood gas analysis	PO_2
	PCO_2
gastrointestinal	obstruction
	poor absorption
	abnormalities
	nasal polyps
	sinusitis
other	male infertility
	osteoarthropathy
	salt depletion

(Chart organized from the original article by Taussig, 1973)

CHART 5

Chrispin & Norman scoring system

Characteristic	Absent	Mild	Severe
Thorax			
Prominent sternum	0	1	2
Diaphragm flattening	0	1	2
Kyphosis	0	1	2
Bronchial wall thickening			
Upper right quadrant	0	1	2
Upper left quadrant	0	1	2
Lower right quadrant	0	1	2
Lower left quadrant	0	1	2
Diffuse consolidations			
Upper right quadrant	0	1	2
Upper left quadrant	0	1	2
Lower right quadrant	0	1	2
Lower left quadrant	0	1	2
Annular images			
Upper right quadrant	0	1	2
Upper left quadrant	0	1	2
Lower right quadrant	0	1	2
Lower left quadrant	0	1	2
Large opacities			
Upper right quadrant	0	1	2
Upper left quadrant	0	1	2
Lower right quadrant	0	1	2
Lower left quadrant	0	1	2

(Chart organized from the original article by Chrispin & Norman, 1974)

CHART 6

Brasfield score

CATEGORY	DEFINITION	SCORE
Air trapping	generalized lung hyperinflation, bulging sternum	0 = absent 1–4 = depending
Linear lesions	diaphragm flattening, thoracic kyphosis, linear densities due to bronchial preeminence, dense parallel lines or with circular densities interspersed with bronchial wall thickening	on the degree of involvement
Nodular cystic lesions	multiple discrete circular densities,(diameter = 0.5), with radiopaque or radiolucent centers, confluent nodules not classified as extensive lesions	
Extensive lesions	atelectasis or consolidations (lobar or segmented), including acute pneumonia	0 = absent 3 = atelectasis 5 = multiple atelectasis
Generalized involvement	impression of severity based on radiographic findings	0 = absent 1–4 = worsening 5 = complications

(Chart organized from the original article by Brasfield, 1979)

CHART 7

Huang scoring system

CLINICAL EVALUATION (50)	SCORE PRE-TREATMENT					SCORE POST-TREATMENT					TOTAL FINAL PERCENTAGE
weight	5	4	3	2	1	5	4	3	2	1	
activities	5	4	3	2	1	5	4	3	2	1	SUM OF PARTIAL
cough	5	4	3	2	1	5	4	3	2	1	VALUES
appetite	5	4	3	2	1	5	4	3	2	1	
pulmonary auscultation	5	4	3	2	1	5	4	3	2	1	$\frac{AS-DS}{DS} \times 100$
respiratory frequency	5	4	3	2	1	5	4	3	2	1	
fever	5	4	3	2	1	5	4	3	2	1	
blood cell count	5	4	3	2	1	5	4	3	2	1	AS = admission sum
cultures	5	4	3	2	1	5	4	3	2	1	DS = discharge sum
general condition	5	4	3	2	1	5	4	3	2	1	
RADIOGRAPHIC (25)											COMPLICATIONS
Air imprisonment	5	4	3	2	1	5	4	3	2	1	
Peribronchial thickening	5	4	3	2	1	5	4	3	2	1	pneumothorax
Nodules/cysts	5	4	3	2	1	5	4	3	2	1	hemoptysis
Segmental/lobar atelectasis	5	4	3	2	1	5	4	3	2	1	hematemesis
											respiratory failure
General impression	5	4	3	2	1	5	4	3	2	1	heart thickening/
TOTAL											congestive heart failure
PULMONARY FUNCTION (25)											
Vital capacity	5	4	3	2	1	5	4	3	2	1	**Score from 5 to 10**
FEV_1/FVC	5	4	3	2	1	5	4	3	2	1	pre-and post-treatment
MMEFR	5	4	3	2	1	5	4	3	2	1	
RV/TLC	5	4	3	2	1	5	4	3	2	1	5 = mild
Vmax 25% VC	5	4	3	2	1	5	4	3	2	1	10 = severe
TOTAL											

(CHART ORGANIZED FROM THE ORIGINAL ARTICLE BY HUANG, 1981)

FEV_1: forced expiratory volume in one second; FVC: forced vital capacity;RV: residual volume; TLC: total lung capacity; Vmax: maximal respiration rate

CHART 8

Bhalla score

CATEGORY	0	1	2	3
BRONCHIECTASIS SEVERITY	absent	Mild (slightly more light transmission than the adjacent vessel)	Moderate (2 to 3 times more light transmission than the adjacent vessel)	Severe (3 times more light transmission than the adjacent vessel)
PERIBRONCHIAL THICKENING	absent	Mild (wall thickness equal to vessel vessel thickness)	Moderate (wall thickness up to 2x vessel vessel thickness)	Severe (wall thickness 2x greater than vessel thickness)
BRONCHIECTASIS EXTENT (BP segments)	absent	1–5	6–9	>9
EXTENT OF MUCOID IMPACTION (BP segments)	absent	1–5	6–9	>9
ABSCESSES OR VESICULATION (BP segments)	absent	1–5	6–9	>9
GENERAL ASPECTS OF BRONCHIAL ZONE INVOLVED (bronchiectasis/impaction)	absent	above 4th generation	above 5th generation	above 6th generation and distal
NUMBER OF BLISTERS	absent	unilateral (≤4)	bilateral (≤4)	>4
EMPHYSEMA EXTENT (BP segments)	absent	1–5	>5	
COLLAPSE/ CONSOLIDATION	absent	subsegmental	segmental lobar	

(Chart organized from the original article by Bhalla, 1991)
BP: bronchopulmonary

CHART 9

Wisconsin score

HYPERINFLATION	NONE	NORMAL / PRESENT	MODERATE	SEVERE
Diaphragm contour	-	0	0.3	1.0
Retrosternal air pressure	-	0	0.3	1.0
Heart position	0	1.0	-	-
Prominent sternum	0	1.0	-	-
Kyphosis	1.0	-	-	
PERIBRONCHIAL		**CENTRAL**		
THICKENING	**MILD**	**MODERATE**	**SEVERE**	**NONE**
PERIPHERAL				
MILD	0.40	0.50	0.65	-
MODERATE	-	0.75	0.85	-
SEVERE-	-	1.00	-	
NONE	0.10	0.20	0.30	0
BRONCHIECTASIS	**NONE**	**MODERATE**	**SEVERE**	
Score every quadrant	0	0.50	1.0	
NODULAR OR BRANCHING OPACITIES	**NONE**	**MODERATE**	**SEVERE**	
Score every quadrant	0	0.50	1.0	
DEFINITE OPACITIES	**ABSENT**	**PRESENT**		
1 lobe	-	0.20		
2 lobes	-	0.40		
3 lobes	-	0.60		
4 lobes	-	0.80		
5 lobes	-	1.0		
ATELECTASIS	**PARTIAL**	**COMPLETE**		
No lobes affected	0	0		
1 lobe	0.0825	0.333		
2 lobes	0.165	0.667		# = 100 points
3 lobes	0.2475	1.00		
4 lobes	0.333	#		
5 lobes	0.415	#		

(Chart organized from the original article by Weatherly, 1993)

CHART 10

Northern scoring system

CLASSIFICATION	RADIOGRAPHIC ALTERATIONS	SCORE
normal	no evident pulmonary involvement	0
mild	minimal increase in linear signs or nodular cystic lesions larger than 0.5 cm diameter	1
moderate	more pronounced linear signs and more diffuse nodular cystic lesions; prominent increase in linear signs	
severe	profuse nodular cystic lesions extensive collapse/consolidation areas	3
very severe	no or small visible areas of normal lung, dense infiltrate	4

(Chart organized from the original article by Conway, 1994)