A simplified cystic fibrosis scoring system (a preliminary report)

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Summary: The inherent variability in the natural severity of cystic fibrosis (CF) makes each aspect of therapy of this condition difficult to isolate and to evaluate objectively. There is little hope of resolving the problems which arise as a result until a way of measuring the severity of the disease at any one point in time is devised. We present a simplified CF scoring system (analogous to the Apgar scoring system for the newborn) based on five simple measurements.

In 1938, Anderson[1] defined cystic fibrosis (CF) as a distinct clinical entity of unknown cause, which may be congenital and familial. Approximately one out of 1000 newborn babies is afflicted with this as yet incurable, progressive disease.[2] Though the child with cystic fibrosis today has a greater chance of reaching adolescence and even early adulthood,[3] the complete cure is not at hand. CF is a multisystem disease. The prognosis is usually dependent on the degree of respiratory tract involvement. Among any group of untreated patients, and among those receiving "ideal" treatment, there is an inherent variability in the natural severity of the disease. Treatment includes, among other things, pancreatic enzyme, vitamins, appropriate antibiotics, a variety of inhalation and mist treatments, mucolytic agents and chest physiotherapy. The multisystem involvement, the variability in severity and the large number of treatments and medications employed make every form of CF therapy suspect. Mist therapy, for instance, which at one time was considered the mainstay of treatment, has in the last few years come under severe criticism. The arguments on both sides have recently been reviewed.[4] A comprehensive clinical grading would help to achieve objectivity in the understanding and management of this disease.

Other scoring systems

The first grading system was conceived "in an effort to compare one patient with the next; to determine the severity of the disease; and to grade the value of therapy".[5] The Shwachman[6] system is based on an appraisal of the following features (1) general activity, (2) physical findings, (3) nutritional status and (4) chest x-rays. A score of 25 points is allowed for each category to give a possible total of one hundred. Scores above 85 are considered excellent; 85-71 is good; 70-56 is mild; 55-41 is moderate, and below 40 is severe. The Shwachman scoring system is the one most widely used and accepted to date. In 1964 Doershuk *et al.*[7] introduced a slightly modified version of the original system, to permit universal application for patients of all ages. These systems, however, are complex and subjective in the hands of other than very experienced clinicians. We have attempted herein to provide a simple reliable tool for personnel working in CF clinics.

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Simplied cystic fibrosis scoring scale (SCS)

This system as devised by us includes five evaluations: (A) general activity, (B) chest radiograph findings, (C) degree of clubbing, (D) growth and development and (E) complications. In the style of the Apgar[8] system, each dimension is scored 0, 1 or 2. The sum of the five individual scores can be calculated instantly (Table I). A patient in excellent health could theoretically have a total score of 10. Also included in our system is a graph chart (Fig. 1) where the clinical score is plotted. We have found it convenient to do the scoring and plotting at the time of the annual chest radiograph.

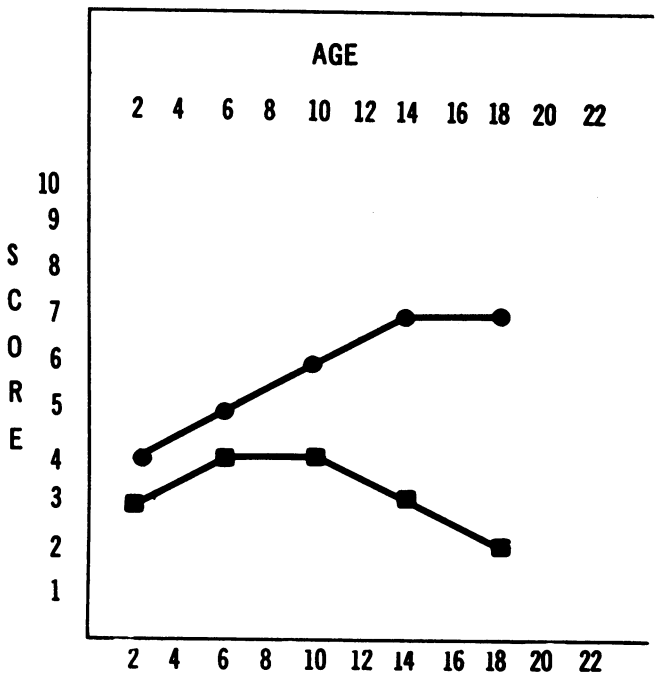

FIG. 1— Graph chart showing the plotted curves of two separate patients over an 18-year follow-up period. The top curve (dots) indicates steady improvement. The bottom curve (squares) indicates a progressive deterioration.

Table I
Simplified CF scoring scale or SCS

	2	1	0
(A) Activity	1. Engages in athletics with normal peers 2. Full activity	Attends regular school with normal peers—misses not more than 2 days per month	
(B) Chest x-ray	Normal	Minimally increased markings and emphysema	
(C) Clubbing	0 to 1+	1+ to 2+ with no cyanosis	2+ and greater
(D) Growth and development	Above 25th percentile for height and weight	Above 3rd percentile for height and weight	Below 3rd percentile for height and weight
(E) Complications	None	Transient	Fixed

Table I

Format used for the clinical scoring with five dimensions for evaluation. Scores of 2, 1 or 0 are assigned to each dimension. Maximal total score of 10 reflects the best prognosis.

Explanation of categories

(A) *Activity*

(i) *School-age children*—All patients having a normal school attendance record and active in sports such as bicycle riding, swimming and hockey would receive a 2 score. An absence of 9.6 days per year has been established[9] as the average absence rate for normal Canadian children. A score of 1 would be given to the child who attends classes with his peers and misses no more than an average of two days per month or 20 days over the entire school year. Engaging in usual activity and moderate exercise, this child keeps up with his peers and at the same time requires no more sleep than they do. Children who miss more than two days of school per month and who do not engage in physical activity with their peers would score 0.

(ii) *Pre-schoolers*—Those required by illness to remain in bed for more than two days per month and those who consistently require more than 17 hours' rest per 24 hours would score 0. (The average Canadian pre-school child sleeps approximately 13 to 15 hours daily.[9])

Those able to keep up with their peers at play and with normal sleep requirements score 2, while children who are occasionally easily fatigued at play and who have an average daily sleep requirement of 15 to 17 hours would score 1.

(B) *Chest radiograph findings*

A score of 2 is given for a normal posteroanterior and lateral chest radiograph. These films are read by a radiologist unattached to the unit. A report of emphysema, linear streaking and nodularities of minimal degree scores 1. Well-defined changes with emphysema, marked streaking and nodularities with or without cystic cavitation score 0.

(C) *Clubbing*

Except in the very young, this phenomenon is one of the most consistent signs of advanced cystic fibrosis and may

Table II

Date	Score					Total	Clinician
	A	B	C	D	E		
Example 1	0	0	0	2	0	2	
Example 2	2	1	2	1	2	8	

Score:	Activity	0
	Chest x-ray	0
	Clubbing	0
	Growth and development	2
	Complications	0
	Score	2

Score:	Activity	2
	Chest x-ray	1
	Clubbing	2
	Growth and development	1
	Complications	2
	Score	8

Table II

Scoring format illustrating the evaluations of two sample cases. A total score of 2 in example 1 indicates a poor prognosis; a score of 8 in example 2 indicates a good prognosis. A place is provided for the signature of the evaluating physican.

be equated with the presence of bronchiectasis. It is a fairly accurate reflection of the severity of pulmonary disease. Clinical estimation of absent to 1+ clubbing scores 2. Definite clubbing with no cyanosis of the nail beds scores 1 and greater than 2 + clubbing scores 0.

(D) *Growth and development*

These are plotted in terms of height and weight using the growth charts of the Boston Children's Hospital. A grading of 25th percentile or better for height and weight scores 2; a grading above the 3rd percentile for height and weight scores 1, and height and/or weight recordings below the 3rd percentile score 0.

(E) *Complications*

Most of the complications of cystic fibrosis, such as rectal prolapse and nasal polyposis, may be troublesome but do not influence prognosis. Meconium ileus, if successfully treated, has no influence on the long-term prognosis. Diabetes mellitus may occur but is not a management problem, nor does it influence prognosis.

Some complications are "fixed" and ominous, and very little good treatment is available. These include cirrhosis of the liver and portal hypertension, cor pulmonale and widespread bronchiectasis with advanced emphysema. The presence of any of the above "big 3" would score 0, as would the presence of persisting widespread rales and crepitations. For scoring purposes, clinically evident minimal to moderate emphysema and/or widespread focal crepitations and rales are considered "transient", and score 1.

Example 1 (Table II)

A 10-year-old boy with known CF presents with portal hypertension. His chest radiograph shows a moderate increase in bronchovascular markings and severe emphysema. Nail beds are 1-2+ clubbed and cyanotic. The boy attends school with his peers and missed eight to 10 days per month in the last year. Height and weight are in the 40th percentile.

Example 2 (Table II)

An 8-year-old girl with recognized cystic fibrosis presents with hemoptysis. The chest x-ray shows minimal increase of bronchovascular markings with no radiological evidence of emphysema. There is no clubbing and the nail beds are free of cyanosis. The child is in grade 2 and attends classes with her peers. In the previous academic year she missed an average of one day per month. She rides a bicycle and keeps up with the other children at play. She had rectal prolapse as an infant, but this cleared after a proper regimen of medication was instituted. She has no other recognized complications. Height and weight are in the 10th percentile.

Discussion

It remains to be seen whether this system of classification will be of value. We hope that it will serve to indicate the merits of various medications and treatments in the management of cystic fibrosis. It must be admitted that the authors' own experience with this system is still limited. It is only during the past year and a half that we have been using it at the Ottawa Civic Hospital CF Clinic. Forty-five patients are periodically examined and evaluated at the clinic. It has been our experience so far that patients scoring 4 or under have a very poor prognosis. On the other hand, those patients scoring 8 and over have an excellent outlook. These are only initial impressions and we hope that the long-term value of the system can be dealt with in a later paper.

There are certain pitfalls that we are aware of. Within any one clinic there is variation in assessment of the degree of clubbing and emphysema; the activity scale of a child under 18 months of age is hard to quantitate. Few serious differences of opinion are evident in actual practice. The scoring scale is weighted towards the status of the lung. This is as it should be, since in our experience and in the experience of others[7] this is the single most important determinant of prognosis. Most workers agree that pulmonary involvement is responsible for most of the morbidity and mortality in cystic fibrosis.

It is our general policy to have annual posteroanterior and lateral chest radiographs taken on all CF patients. It is our impression that this is the best time to assign the score to the patient and plot the graph.

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Résumé

Méthode simplifié de cotation de la mucoviscidose: Rapport préliminaire

Par suite du caractère essentiellement variable de la sévérité de la mucoviscidose, il est difficile d'évaluer de façon objective chaque aspect isolé du traitement. Il y a peu d'espoir de résoudre les problèmes thérapeutiques qui se posent à moins de mettre au point un moyen de mésurer la sévérité de la maladie à un moment quelconque de son évolution. Nous proposons ici une méthode simplifiée de cotation de la mucoviscidose, basée sur cinq mesures simples (méthode analogue à la méthode d'Apgar pour le nouveau-né).