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This Week's Citation Classic®

Shwachman H & Kulczycki L L. Long-term study of one hundred five patients with cystic fibrosis: studies made over a five- to fourteen-year period. *AMA J. Dis. Child.* 96:6-15, 1958.
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The cited report provides a method for scoring the severity of illness in patients with cystic fibrosis. One hundred five patients were studied over a 5- to 14-year period. Their clinical conditions were stated at the beginning and at the end of the study. In addition, the paper introduced the sweat test as a new method of confirming the diagnosis. [The *SCI*® indicates that this paper has been cited in over 295 publications.]

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[Editor's note: H. Shwachman wrote this commentary for *Citation Classics* before his death in 1986. We are publishing it posthumously with comments added by L.L. Kulczycki, his coauthor.]

My interest in cystic fibrosis (CF) was first aroused when I was a house officer at the Boston Children's Hospital in 1938. There I saw my first CF patients even though I had spent over 16 months on the pediatric service at the Johns Hopkins Hospital.

Upon completion of my five years of clinical training in Boston, I was offered a research fellowship to study CF with Sidney Farber. He had made original observations of CF pathophysiology and had subsequently coined the term mucoviscidosis. In 1955 Lucas L. Kulczycki was invited by Farber and Charles Janeway of the Children's Hospital Medical Center in Boston to join me in this research. At that time Kulczycki was working on childhood tuberculosis. (He had trained in Scotland, England, and Canada.) Kulczycki became my close associate at the Children's Hospital in Boston for over 10 years.

Most of our time was devoted to the diagnosis and management of CF patients. We built one of the earliest and largest CF centers in the country, eventually reaching a roster of nearly 700 patients. Patients from many countries and from all over the US came to see us for diagnostic and therapeutic purposes.

In our report we pointed out that one need not have complete pancreatic insufficiency to have CF. This was contrary to accepted knowledge.[1] In 1954 we developed the first practical sweat test,[2] which was modified in 1959 by L.E. Gibson and R.E. Cooke[3] and became the standard diagnostic procedure. But we failed to mention that the majority of adult males with CF were sterile, a fact we reported a few years later when we described the anatomical defect that explains the cause of the azospermia.[4,5] We also failed to mention dental staining, a complication commonly seen following prolonged use of tetracyclines in children under seven years of age.[6]

The main thrust of the paper was to develop a system of clinical scoring that defined the severity of the disease in any one patient at any given time. The score did not require laboratory testing but rather a careful clinical appraisal and a chest roentgenogram of the patient. (Four serious complications that affect the scoring system and the prognosis had not yet been observed at the time of our report. These complications, all affecting the respiratory system, are (1) pneumothorax, (2) massive pulmonary hemorrhage, (3) cor pulmonale, and (4) the permanent colonization of the pulmonary flora with strains of *Pseudomonas* species, particularly mucoid strains that are almost always found only in patients with CF.[7])

The study also provided new information on the natural history of the disease, until then considered a fatal nutritional disease of infants and children. In addition, the report provided hope and encouragement to doctors and to patients and their families by providing a beneficial therapeutic program. We indicated that many patients could lead comfortable lives into adulthood, even if they had been born with meconium ileus, the first clinical manifestation of CF, which occurs in 15 percent of all CF patients.[8]

In March 1978 the Cystic Fibrosis Foundation arranged a special conference in Tucson, Arizona, devoted to "Cystic Fibrosis Patient Evaluations and Scoring Systems." From previous scientific references and from that conference, it became obvious that the Shwachman-Kulczycki (S-K) scoring system is a prevalent system for evaluating CF patients' progress. It is practical and easily applied even in the physician's office. The chest X-ray film scoring is the only parameter requiring laboratory input.

After 25 years of daily experience using the S-K scoring system, it appears that each parameter included in the system (activity, physical examination, nutrition, and X ray) has an arbitrarily assigned value of 25 points and reflects very adequately the CF patient's long-term prognosis.

Several recent sophisticated studies using computerized tomography, ultrasound, lung scanning, and pulmonary function tests complement and verify the S-K scoring system. One recent example is cited by W.J. Warwick and C. Wielinski,[9] who, after running a computerized study of S-K and NIH scoring systems, concluded that S-K scoring would indeed gauge the severity of the complications and that a single number may have substance.

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2. **Shwachman H, Leubner H & Catzel P.** Mucoviscidosis. *Advan. Pediat.* 7:249-323, 1955. (Cited 135 times.)
3. **Gibson L E & Cooke R E.** A test for concentration of electrolytes in sweat in cystic fibrosis of the pancreas utilizing pilocarpine by iontophoresis. *Pediatrics* 23:545-9, 1959. (Cited 440 times.) [See also: **Gibson L E.** Citation Classic. (Barrett J T, ed.) *Contemporary classics in clinical medicine.* Philadelphia: ISI Press, 1986. p. 51.]
4. **Kaplan E, Shwachman H, Perlmutter A D, Rule A, Khaw K T & Holsclaw D S.** Reproductive failure in males with cystic fibrosis. *N. Engl. J. Med.* 279:65-9, 1968. (Cited 105 times.)
5. **Holsclaw D S, Perlmutter A D, Jockin H & Shwachman H.** Genital abnormalities in male patients with cystic fibrosis. *Urology* 106:568-74, 1971.
6. **Shwachman H & Schuster A.** The tetracyclines: applied pharmacology. *Pediat. Clin. N. Amer.* 3:295-303, 1956.
7. **Shwachman H.** Cystic fibrosis. *Curr. Prob. Pediat.* 8:1-72, 1978.
8. **Donnison A B, Shwachman H & Gross R E.** A review of 164 children with meconium ileus seen at the Children's Hospital Medical Center, Boston. *Pediatrics* 37:833-50, 1966. (Cited 85 times.)
9. **Warwick W J & Wielinski C.** A new look at the NIH prognostic score for cystic fibrosis. *Cystic Fibrosis Club Abstr.* 27:170, 1986.

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