Effective improvement of cystic fibrosis care by application of elementary measures: essential lessons not only for developing countries

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Cystic fibrosis is a frequent, pan-ethnic (Figure 1),[1] inborn disease that leads to early death without appropriate treatment. The increasing impact of cystic fibrosis in the developing world has been emphasized recently for further countries.[2]

Continuous improvements in cystic fibrosis care have led to a linear increase of survival with the disease (Figure 2). This positive effect was achieved in the industrialized countries without revolutionary medical innovations. The practice of "best care" alone has been responsible for such a success up to now. "Best care" must not be costly but needs a preferable multidisciplinary, consistent and well-educated team to be delivered.

Morrow et al. report in this issue of the journal the successful improvements in lung function of a pediatric cystic fibrosis population in a developing country.[3] With small changes in management and their longstanding and continued efforts, an improvement of average lung function of their cystic fibrosis population by about 20% was achieved between 1999 and 2006. In accordance, the average severity of lung disease has improved during the 8-year study period.

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Lung function, in particular forced expiratory volume in 1 second (FEV_1) is a surrogate for survival.[4] Although the latter has not been demonstrated directly in this cohort, there is good evidence that it will follow the trend.

Which factors accounted for the success?

It is certainly *not* necessary to use elaborated and expensive diagnostic tools, such as computed tomography (CT) scanning or fancy lung function testing. Therapeutic management of cystic fibrosis may include use of readily available hypertonic saline in place of DNase (Table 1 of the article by Morrow et al.)[3] and the supply with locally available enteric-coated enzyme tablets instead of elaborated enteric-coated spherules.

However, a major factor of cystic fibrosis care is the availability of manpower. This is still a major issue, even in industrialized countries. A team that steadily sticks to "best care" attitudes will be able to attain reduced morbidity and mortality of the patients.

Constant team members including the senior cystic fibrosis physician, cystic fibrosis physiotherapist, nurse specialist, and others build long-term relationships with their patients, simultaneously establishing cystic fibrosis experience and expert knowledge. Care in large cystic fibrosis centers provides a systematic and highly cost-effective way to use available resources. A disadvantage of specialized centers might be travel expenses for the patients and it is a wise approach to help the patients with these in order to achieve regular appointments.

Cystic fibrosis center care as well as inpatient hospital care must segregate all patients according to their microbiological status. This can be achieved at almost no costs, simply by organizational means.[5] Ultimate hygiene procedures during lung function testing, accurate weight and height measurements, as well as microbial cultures of respiratory secretions at scheduled follow-up visits, best every 1-3 months, will lead to the optimal assessment of the disease condition and prompt adequate treatments.

Removal of secretions by appropriate physiotherapeutic techniques must be taught and each patient and the family need instruction and reinstruction on a continuous base. It may be difficult to guarantee access to this very important part of care due to long distances to the center, socioeconomic problems, and the lack of qualified staff outside the center. Thus, teaching the patient and the family at every visit to the center is crucial.

Access to care should be guaranteed irrespective of socioeconomic status. Difficulties referring to this issue might be

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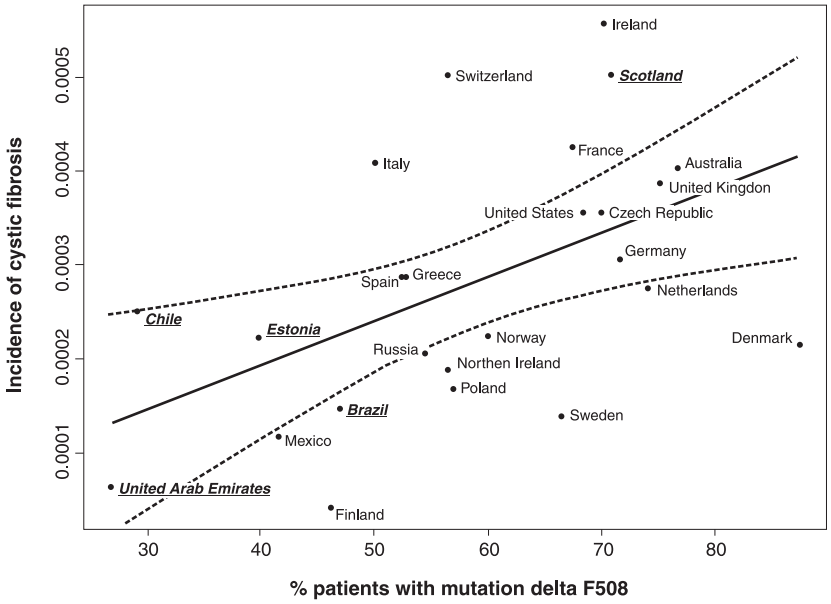

Figure 1 - Percentage of patients with the most frequent cystic fibrosis mutation dF508 and the incidence of cystic fibrosis. The line represents linear regression analysis with 95% confidence interval[1]

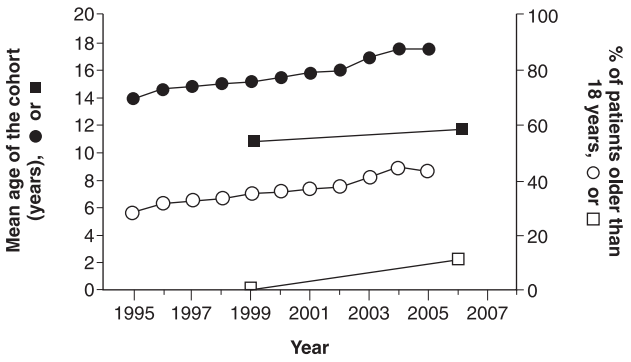

Figure 2 - Mean age (left axis, filled symbols) and percentage of patients above 18 years (right axis, open symbols) of the Cape Town cystic fibrosis center (squares) and of the German CF cohort from the German Cystic Fibrosis Quality Assessment project 1995-2006 (circles). Note that from the Cape Town center only patients (Table 1 in Morrow et al.)[3] who were old enough to be able to perform lung function tests were included. Thus, median age may be over estimated from the lack of inclusion of younger children

reflected by the underrepresentation of black Africans in the patient cohort (5%) reported from South Africa. Although the exact prevalence of cystic fibrosis in Black Africans is unknown, studies on the migrant African American population in the USA estimated a similar cystic fibrosis mutation frequency in this population.[1]

Chronic infection of the respiratory tract is the major cause of death in cystic fibrosis. In their study Morrow et al. reported that the high percentage of chronic patients infected with *Pseudomonas aeruginosa* was essentially unchanged in 2006.[3] In addition to hygiene measures to prevent cross infection with cystic fibrosis pathogens, early treatment strategies towards eradication of these bacteria are essential to reduce this burden.[6] As expected, the prevalence of *Staphylococcus aureus* was also very high in their population. Antimicrobial treatments using bactericidal antibiotics such as oxacillin or cephalexin may be instituted early on and may lead to eradication of these bacteria.[7] Similarly, the introduction of azithromycin as performed by Morrow et al.[3] may contribute to a reduction of the bacterial load. However, using azithromycin, bacterial resistance of *Staphylococcus aureus* often develops and, if so, this antibiotic does not contribute to an anti-staphylococcal effect.[8] Morrow et al. also reported on the introduction of prophylactic care, including regular seasonal influenza vaccinations.[3]

The database collected by the authors may be the starting point of a patient registry (Figure 2) that may further help to assess and subsequently also to improve the quality of care.

We have to congratulate the authors of this paper for their longstanding interest and fundamental care for this demanding group of patients.[9-11] They demonstrated that the introduction of the traditional tools to improve care of patients with cystic fibrosis even under very difficult conditions will achieve substantial health improvements regardless of which part in the world you live in.

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