

Editorial

Standards of Care for Cystic Fibrosis ten years later



In 2004 the European Cystic Fibrosis Society (ECFS) convened a Consensus Conference on standards of care for cystic fibrosis (CF). CF professionals from many European countries, and some from outside Europe, produced a document that was later published in the *Journal of Cystic Fibrosis* [1]. This consensus on standards of care proved to be very useful for CF centres negotiating with the funding agencies in their countries and set the scene for better CF care in Europe.

Ten years later CF care worldwide and specifically in Europe has evolved in many ways:

- Early diagnosis through neonatal screening has proliferated across Europe [2].
- The adult population of patients has been steadily increasing and in some countries has now numerically passed the paediatric population [3].
- New treatments are available, including mutation-specific therapies that might have the potential to change the natural history of the disease [4].
- More emphasis is given to clinical research and a network of European CF centres has been established with the aim of facilitating high quality clinical trials to translate new therapies into clinical care [5].
- Epidemiological data from many European countries have become available through the EuroCareCF project [6] and national and international registries [3]. Evidence of disparities in allocation of resources and in clinical outcomes has emerged.

These momentous changes prompted the ECFS to plan an updated version of the 2004 document. A broad spectrum of CF professionals from Europe, North America and Australia and various representatives of patient support groups were involved in pre-meeting consultations and in the drafting of preliminary documents. These were discussed during a dedicated Consensus Conference that took place in Verona, Italy, on April 12–13, 2013, attended by 39 experts and stakeholders. The three articles in this supplementary issue of the *Journal of Cystic Fibrosis* are the results of this process.

Each one of the three papers has been developed by a team of professionals/experts directed by a group coordinator and addresses a specific field of the standards of care. These three

areas cover: a) the required framework of the CF Centre; b) best clinical practice; and c) quality management in CF care.

The *Centre Framework* document emphasizes the unquestionable role of dedicated CF centres for achieving optimal outcomes. The paper highlights the importance of the multidisciplinary team, describes the general structure of paediatric and adult CF centres and stresses the need for their cooperation and for a transition programme. The roles of professionals involved in the care of CF are described, including the clinical director, specialist doctors, specialist nurses, physiotherapists, dietitians, microbiologists, pharmacologists, psychologists, clinical geneticists and social workers. All disciplines are strongly encouraged to be members of their national and international specialist groups, to update themselves by attending meetings, and to participate in audit and research whenever possible. The challenges related to the development of dedicated health services in low income countries and the perspectives of people with CF mediated by patients' organisations are also discussed.

The *Best Practice* document describes the services centres should provide using a question and answer model. The paper is extensive, and difficult to condense in a few sentences. A synopsis of the main messages includes:

- Early and accurate detection through neonatal screening and expertise to undertake biochemical and genetic diagnostic testing are vital and enable access to specialist care from early in life.
- Prompt recognition of pulmonary infection and deterioration permits intervention with timely treatments which are proven to reduce the impact of CF on the lungs.
- Nutritional and metabolic complications require regular monitoring and timely and effective intervention.
- CF professionals need to have experience and expertise in all common complications and to have developed referral pathways with other disciplines to support more complex situations.
- Appropriate management and communication are particularly important at transplantation referral and end of life.
- Despite excellent improvement in clinical outcomes and survival for people with CF, psychological complications are common for the patient and their family and require early recognition, assessment and management with

the active involvement of experienced psychological health care professions.

In consideration of the magnitude of the topics, specific treatments and procedures are not described in detail. For example, the necessity of having protocols for *Pseudomonas aeruginosa* eradication is clearly expressed, but single protocols are not discussed.

The *Quality Management* document reviews management of quality of care for individuals with CF at several levels: patient, centre, regional, national and international. Improvement issues are examined with particular reference to annual assessment, patient quality management charts, CF team sessions, therapy goals, certification, peer review, public reporting, quality groups, ranking, learning from best practice, interaction with registries, benchmarking, and cooperation with national organisations.

A lesson learned after the publication of the previous standards of care document [1] was that although awareness of it and its dissemination in Europe were good, its implementation was quite dishomogeneous [7,8]. The heterogeneity of care organisations in Europe and the absence in some countries of dedicated CF centres plus inadequate resources made it difficult, if not impossible, to universally apply the suggested standards.

The authors of the present documents are aware of the barriers that some countries, in which CF services are absent or minimal, may encounter in implementing the new standards of care. Their accomplishment is made challenging by deficiencies in political prioritization, inappropriate funding and a lack of staff recruitment and training. However, we strongly believe that all European nations should strive to achieve a model of CF care in accordance with the ECFS recommendations. Inequalities in levels of assistance and the extreme variation in the survival of people with CF across European countries [6] are not acceptable, and every effort must be made to deliver high standards. Optimal care may not be an easily achievable target and in some less advantaged countries a stepwise approach may be the only deliverable approach, but the present standards should remain as a goal.

ECFS considers the publication of these documents as an important step in the provision of the best possible care to people with CF, and plans to make it not a one-off action, but a component of a wider project which will include further initiatives. The concepts in the articles will be integrated in the papers produced by the Joint Task Force on CF Adult Care assembled by the European Respiratory Society and the ECFS. Additional developments under consideration are the periodical review and update of the Best Practice section, the production of training tools for quality improvement and the analysis of potential international quality indicators in collaboration with the ECFS Registry.

Conflict of interest

C. Castellani: consultancy for Vertex and Gilead, lectures for Chiesi and Novartis, outside the submitted work. A.R.

Smyth: personal fees from Gilead, MPEX, Pharmaxis and Vertex, grants from Forest Labs, outside the submitted work. J.S. Elborn: President of ECFS. Institutional payments for consultancy and clinical trials from Vertex, Gilead, Novartis, outside the submitted work. S. Conway and M. Stern have no conflicts of interest to report.

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