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Editorial

European cystic fibrosis society standards of care: A road map to improve CF outcome

The phenomenal improvement in survival of children and adults with cystic fibrosis (CF) is one of the unsung success stories of modern medicine. A lethal disease with no current cure which previously was considered to be associated with early death in childhood is now one of adult survival. The two senior statesman of CF who have written this editorial can well remember the medical carnage and sustained mortality of children with CF that occurred in the late 60s and early 70s. Facilities and therapy for the care of paediatric and adult patients with CF in Europe were negligible. Gradually this dismal scenario has been transformed and eroded whereby the morbidity and almost all of the mortality associated with CF has shifted to adults. Adults with CF have become older than members of their healthcare team and survival curves have improved decade on decade; median survival is now into the fifth decade of life in parts of the world. This survival success for CF as a lethal disease far outstrips the survival figures for many common cancers and other lethal diseases.

How has this success story been achieved? Clearly there have been significant therapeutic advances which have controlled and modulated disease progression but most important has been the recognition that CF is a complex multisystem disease which requires long term, centralized and multidisciplinary team (MDT) care with designated facilities. An MDT will consist of many different disciplines such as physiotherapists, doctors, dieticians, nurses, social workers and clinical psychologists.

The first step to improve quality is to measure outcomes. The advent of CF registries has examined outcomes in CF centers for prime markers which influence disease progression such as lung function, nutritional status and microbiological prevalence of infecting (and even cross infecting) organisms. These registries have illustrated variation between centers within and between countries [1–3]. Often these variations are understandable and related to poor funding and resourcing of CF care, however, they also point to different practices and treatment approaches. Bench marking comparisons revealed differences in disease severity of patients between the centers. Further studies revealed that centers that had better results were characterized by maintaining standards of care and adherence to guidelines [4–6].

The document Standards of Care of Children and Adults with Cystic Fibrosis in the UK was updated in 2011 and has been a crucial tool for the continuing development of CF centers in the UK [7]. Published in this special supplement of the Journal are the updated European Cystic Fibrosis Society Standards of Care. They undoubtedly break new ground in their concept. Under the title heading of Standards of Care they are three separate papers;

1) A Framework for the Cystic Fibrosis Center [8] which describes the recognised requirements of care in a CF center
2) Best Practice Guidelines [9] which details questions and answers to the most common clinical problems in CF care
3) Quality Management in Cystic Fibrosis [10] (vide infra).

Each of the monographs is multi-authored by world- wide experts in CF and is clearly the distillation of considerable hard work and expertise. Each of the papers is well written and essential reference material for benchmarking CF care. The best practice document is an important tool to standardize the daily care of patients with CF. It provides guidelines many of which are based on experts' opinions and not on evidence based on clinical trials. Therefore these guidelines need to be reviewed regularly and physicians should keep updated with the medical literature for updates or changes.

However, the paper addressing Quality management in Cystic Fibrosis is the most interesting. It is essential reading for all leads of the MDT in CF centers. It describes the process of evaluating quality care delivered at patient, center and national level. As a consequence of accumulated data from registries national and international comparisons can be made for CF care. The need for patient involvement is emphasized.

The question that needs to be asked is whether these European Cystic Fibrosis Society Standards of Care have the power to influence and improve European care of CF children and adults. The drive to provide high level CF care will be different for economically advantaged and disadvantaged countries. There are several steps needed in order to modify traditional practices and implement changes in the care of patients. It needs leadership by the CF Center Directors and their teams since many of these guidelines do not require extra resources. In addition, these documents should be a road map for health policy makers to plan the development of their

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institutions or communities care. By standardization of care and comparison of outcomes through registries it will be possible to identify areas that need to be augmented in order to improve outcome. Peer Review has made considerable differences to improving CF within the UK over the last decade and the topic of peer review and quality accreditation programmes is carefully discussed in quality management in cystic fibrosis.

These documents present the current optimal standards of care; a goal that needs to be adopted by all centers in all the countries. We suggest that each of the centers will review their own practice and point the areas that need to be improved.

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