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Review

European Cystic Fibrosis Society Standards of Care: Quality Management in cystic fibrosis

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Abstract

Since the earliest days of cystic fibrosis (CF) treatment, patient data have been recorded and reviewed in order to identify the factors that lead to more favourable outcomes. Large data repositories, such as the US Cystic Fibrosis Registry, which was established in the 1960s, enabled successful treatments and patient outcomes to be recognized and improvement programmes to be implemented in specialist CF centres. Over the past decades, the greater volumes of data becoming available through Centre databases and patient registries led to the possibility of making comparisons between different therapies, approaches to care and indeed data recording. The quality of care for individuals with CF has become a focus at several levels: patient, centre, regional, national and international. This paper reviews the quality management and improvement issues at each of these levels with particular reference to indicators of health, the role of CF Centres, regional networks, national health policy, and international data registration and comparisons.

Keywords: CF registries; Standards; Models of care; Quality management; Outcomes in CF

Contents

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1. Introduction

Since the earliest days of cystic fibrosis (CF) treatment, detailed summaries of large clinical groups have been employed to determine and describe the best approach to treatment, based on improved outcomes [1,2]. A comprehensive approach to therapy, routine monitoring, and attention to individual profiles and prognostic subgroups were highlighted in these early papers on CF management. The early years also included a cautionary tale, when mist tent therapy was cited as the key component responsible for remarkable survival in one large clinic, as documented in the newly established US CF Registry [3,4]. Over the following decade, however, it became clear that the scientific evidence of a beneficial effect was lacking for mist tent therapy [5]. Nevertheless, the improved outcomes were real, and emphasis was eventually, and more appropriately, placed on the comprehensive management package, including early diagnosis, patient and parent education, frequency of patient visits, daily physical therapy and aggressive antibiotic therapy. It was also during this period that a new focus on growth and nutrition was evolving. Again, it began with reports from a large clinic where greatly improved outcomes were observed in patients with CF who were prescribed a high-fat diet in place of the historical low-fat diet [6,7]. But it was only when the CF registry data for two large, university-based clinics with similar demographics and approaches to other aspects of treatment were compared that the possibility of a normal diet and the goal of normal growth in patients with CF were widely embraced [8].

Although the benefits of specific treatments must be supported by evidence from well-controlled studies, there is great value in compiling and comparing outcomes in large clinical populations in order to document changes over time and to identify patterns and practices that may be associated with benefit or concern. Of particular importance are national registries that account for all, or a large and well-defined proportion of, CF patients in a region. National, annually updated CF registries in the USA since 1966 [9] and Canada since 1970 [10] were instituted primarily to describe population patterns of diagnosis, demographics and mortality. Over the years additional information was added to track