ELSEVIER

Journal of Cystic Fibrosis 13 (2014) S23 – S42

Journal of Cystic Fibrosis

www.elsevier.com/locate/jcf

Review

European Cystic Fibrosis Society Standards of Care: Best Practice guidelines

Alan R. Smyth [a,*], Scott C. Bell [b,c], Snezana Bojcin [d,v], Mandy Bryon [e], Alistair Duff [f], Patrick Flume [g], Nataliya Kashirskaya [h], Anne Munck [i,j], Felix Ratjen [k,l], Sarah Jane Schwarzenberg [m], Isabelle Sermet-Gaudelus [n,o,p], Kevin W. Southern [q], Giovanni Taccetti [r,s], Gerald Ullrich [t], Sue Wolfe [u]

[a] *Division of Child Health, Obstetrics & Gynaecology (COG), School of Medicine, University of Nottingham, UK*
[b] *Department of Thoracic Medicine, The Prince Charles Hospital, Australia*
[c] *Queensland Children's Medical Research Institute, Brisbane, Australia*
[d] *Cystic Fibrosis Europe, Denmark*
[e] *Cystic Fibrosis Unit, Great Ormond Street Hospital for Children, London, UK*
[f] *Regional Paediatric CF Unit, The Leeds Children's Hospital, Belmont Grove, Leeds LS2 9NS, UK*
[g] *Medical University of South Carolina, Charleston, SC, USA*
[h] *Department of Cystic Fibrosis, Research Centre for Medical Genetics, RAMS, Moscow, Russia*
[i] *Assistance publique-Hôpitaux de Paris, Hôpital Robert Debré, Paediatric Gastroenterology and Respiratory Department, CF Centre, Université Paris 7, 75019, Paris, France*
[j] *Association française pour le dépistage et la prévention des handicaps de l'enfant (AFDPHE), France*
[k] *Division of Respiratory Medicine, Department of Paediatrics, The Hospital for Sick Children, University of Toronto, Canada*
[l] *Physiology and Experimental Medicine, Research Institute, The Hospital for Sick Children, University of Toronto, Canada*
[m] *Pediatric Gastroenterology, Hepatology, and Nutrition, University of Minnesota, Amplatz Children's Hospital, Minneapolis, MN, USA*
[n] *INSERM U1151, France*
[o] *Université René Descartes Paris 5, France*
[p] *Unité fonctionnelle de Mucoviscidose, Service de Pneumo-Pédiatrie, Hôpital Necker-Enfants Malades, 149 rue de Sèvres, 75743, Paris, France*
[q] *Department of Women's and Children's Health, University of Liverpool, UK*
[r] *Institute of Child Health, Alder Hey Children's Hospital, Eaton Road, Liverpool L12 2AP, UK*
[s] *Cystic Fibrosis Centre, Department of Paediatric Medicine, Anna Meyer Children's University Hospital, Florence, Italy*
[t] *Reutzstr. 1, 19055, Schwerin, Germany*
[u] *Paediatric Cystic Fibrosis, Regional Paediatric CF Unit, The Leeds Children's Hospital, Belmont Grove, Leeds LS2 9NS, UK*
[v] *Macedonian Cystic Fibrosis Association, Misko Mihajlovski 15, 1000 Skopje, Republic of Macedonia*

Abstract

Specialised CF care has led to a dramatic improvement in survival in CF: in the last four decades, well above what was seen in the general population over the same period. With the implementation of newborn screening in many European countries, centres are increasingly caring for a cohort of patients who have minimal lung disease at diagnosis and therefore have the potential to enjoy an excellent quality of life and an even greater life expectancy than was seen previously. To allow high quality care to be delivered throughout Europe, a landmark document was published in 2005 that sets standards of care. Our current document builds on this work, setting standards for best practice in key aspects of CF care. The objective of our document is to give a broad overview of the standards expected for screening, diagnosis, pre-emptive treatment of lung disease, nutrition, complications, transplant/end of life care and psychological support. For comprehensive details of clinical care of CF, references to the most up to date European Consensus Statements, Guidelines or Position Papers are provided in Table 1. We hope that this best practice document will be useful to clinical teams both in countries where CF care is developing and those with established CF centres.

Keywords: Cystic fibrosis; Standards of care; Multidisciplinary management

* Corresponding author.
E-mail address: Alan.Smyth@nottingham.ac.uk (A.R. Smyth).

http://dx.doi.org/10.1016/j.jcf.2014.03.010
1569-1993/

Contents