# Table of Contents

<table>
<thead>
<tr>
<th>Section</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>Executive Summary</td>
<td>4</td>
</tr>
<tr>
<td>Foreword</td>
<td>5</td>
</tr>
<tr>
<td>Purpose and Scope</td>
<td>5</td>
</tr>
<tr>
<td>Consultation Process</td>
<td>6</td>
</tr>
<tr>
<td>Review Process</td>
<td>7</td>
</tr>
<tr>
<td>Steering Committee</td>
<td>7</td>
</tr>
<tr>
<td>Disclosure Statement</td>
<td>8</td>
</tr>
<tr>
<td>Glossary of Terms</td>
<td>9</td>
</tr>
<tr>
<td>Methods - Search Strategies and Levels of Evidence</td>
<td>10</td>
</tr>
<tr>
<td>Introduction – Facilities, Staffing and Services</td>
<td>11</td>
</tr>
<tr>
<td>Newly Diagnosed Children with Cystic Fibrosis</td>
<td>21</td>
</tr>
<tr>
<td>Newly Diagnosed Adolescent and Adult with Cystic Fibrosis</td>
<td>25</td>
</tr>
<tr>
<td>Outpatient Care</td>
<td>31</td>
</tr>
<tr>
<td>Inpatient Care</td>
<td>37</td>
</tr>
<tr>
<td>Home Therapy</td>
<td>43</td>
</tr>
<tr>
<td>Cystic Fibrosis: Transition from Paediatric to Adult Care</td>
<td>47</td>
</tr>
<tr>
<td>Outreach Services and Clinics</td>
<td>51</td>
</tr>
<tr>
<td>End of Life Care and Transplantation</td>
<td>55</td>
</tr>
<tr>
<td>Australian Cystic Fibrosis Data Registry</td>
<td>63</td>
</tr>
<tr>
<td>Role of Cystic Fibrosis Organisations in Australia</td>
<td>65</td>
</tr>
<tr>
<td>Appendices</td>
<td>67</td>
</tr>
</tbody>
</table>
The Cystic Fibrosis (CF) Standards of Care project was proposed by the Cystic Fibrosis Centre Directors Committee, and supported by Cystic Fibrosis Australia and the Thoracic Society of Australia and New Zealand (TSANZ). This document represents the first comprehensive analysis of the requirements for health care and its delivery for people with CF in Australia. Cystic fibrosis is the most common lethal genetic condition affecting Australians with currently ~3000 people living with CF in Australia.

The project scope included assessment of need for CF guideline development, particularly focussing on the requirements for facilities, staffing and services to provide CF care with specific reference to health care delivery and systems, population demographics and climate in Australia. A multidisciplinary Steering Committee which included representatives from clinical care teams, lay organisations and adults with CF from Australia was established to oversee the overall development of the project, including review of published guidelines for CF. Regular teleconferences, meetings and extensive consultation with health care professionals involved in CF care delivery in Australia were undertaken. The Standards of Care have been submitted to the Royal Australasian College of Physicians, the TSANZ and Cystic Fibrosis Australia who have each endorsed the document.

Executive Summary

Specific areas covered by the Standards include:

i  Facilities, staffing and services required at CF centres and CF clinics;
ii Newly-diagnosed children with CF;
iii Newly-diagnosed adolescent and adult with CF;
iv Outpatient care;
v Inpatient care;
vi Home therapy;
vii Transition Care;
viii Outreach Services and Clinics;
ix End of Life Care and Transplantation;
x Role of Cystic Fibrosis Organisations.

Each chapter provides a comprehensive literature review and outlines guidelines for clinical care by taking into account requirements for facilities, staffing and services. This document is based on current evidence and best practice, and outlines the ideal situation for the management of people with CF in Australia. Whilst CF centres may not achieve all standards at present, CF services should aspire to, and ideally, have resources available, to support this care.
The writing of Cystic Fibrosis (CF) Standards of Care was proposed by the Australian Cystic Fibrosis Centre Directors Committee, and supported by Cystic Fibrosis Australia and the Cystic Fibrosis Special Interest Group of the Thoracic Society of Australia and New Zealand (TSANZ). Following the Sixth Australian Cystic Fibrosis Conference held in Adelaide in August 2006, a multidisciplinary Steering Committee was established to coordinate the overall development of the Standards of Cystic Fibrosis Care Project, including review of current published guidelines for CF. Terms of reference were agreed upon and members of the Steering Committee have worked with other interested parties and stakeholders throughout the development of these guidelines.

The scope of this project included:

1. **Assessment of areas of need for CF guideline development and included:-**
   a. What constitutes a cystic fibrosis centre and cystic fibrosis clinic;
   b. Assessment of the requirements for facilities, staffing and services to provide CF care;
   c. Assessment of the requirements for outpatient care, including home intravenous therapy, outreach services, regional centre care and access to allied health specialist services;
   d. Assessment of the requirements for the delivery of inpatient care;
   e. Assessment of the requirements for the assessment and education of newly diagnosed patients with CF.

2. **Guidelines were to be written with specific reference to health care delivery and systems, population demographics and climate in Australia.**

The membership of the Steering Committee included representatives from clinical care teams, lay organisations and adults with CF from Australia (details of committee membership provided below). Each section includes the Section Leaders and all Steering Committee members had input into each of the Section Drafts throughout the project.

Writing groups led by Medical and Nursing specialists in the following areas were established:
   i. Introduction, including facilities, staffing and services required at CF centres and CF clinics;
   ii. Inpatient care;
   iii. Outpatient care;
   iv. Newly-diagnosed child with CF;
   v. Newly-diagnosed adolescent and adult with CF;
   vi. Transition;
   vii. Outreach/Shared Care;
   viii. End of Life (including Transplantation);
   ix. Home therapies;
   x. Role of Cystic Fibrosis Organisations.
Consultation Process

The initial meeting of the Steering Committee was held by Teleconference in February 2006. Monthly teleconferences were held throughout 2006, and a face-to-face meeting of Steering Committee members, facilitated by Professor Craig Mellis from the University of Sydney, was held in November 2006. During this one-day meeting, Draft Guidelines for each of the sections were reviewed and modifications of the sections were made accordingly.

In April 2007, Dr Dominic Fitzgerald from the Children’s Hospital at Westmead was invited to act as Medical Editor of the Draft Guidelines. Written feedback and suggestions for modifications to the Draft Guidelines was provided, and writing groups further modified the chapters. A further review of the Guidelines was performed by Dr Fitzgerald in July 2007.

Copies of the Draft Guidelines were provided to the CF community, including CF centres and clinics, Lung Transplant Units, CF Organisations, the TSANZ and the Royal Australasian College of Physicians. During Australian Cystic Fibrosis Conference in August 2007, there was an opportunity for conference delegates from the CF community to provide comment to Steering Committee members. A presentation regarding the progress of the Standards of Cystic Fibrosis Care Project was given at the Australian CF conference. Final comments were received following the conference and appropriate modifications made on the basis of the feedback in October 2007. A final review of the Guidelines was performed by Dr Fitzgerald in November 2007. The final document was have been reviewed by Cystic Fibrosis Australia, the TSANZ and the Royal Australasian College of Physicians for endorsement.

This project, along with the recently published Australasian Clinical Practice Guidelines for Nutrition in Cystic Fibrosis, the Infection Control Guidelines and Australian Clinical Practice Guidelines for Physiotherapy in Cystic Fibrosis represent the first comprehensive descriptions of CF care guidelines to be prepared in Australia. The Steering committee believe these projects constitute a beginning and further Guidelines should be considered, including the Management of specific CF related complications (eg. Management of Pulmonary Disease in CF and Management of Endocrine Complications in CF) and Treatment strategies for health maintenance of people with CF.

The following guidelines represent the ideal characteristics (staffing, facilities and services) for the management of people with CF in Australia. Whilst CF centres may not reach all standards at present, CF services should aspire to, and ideally have resources available, to support this care.

This document is based on current best practice.
An evaluation of the guidelines will be undertaken across all CF centres in Australia. Prior to 2012, this document will be reviewed and updated. Updates of the guidelines will incorporate the findings of the evaluation as well as new evidence from the scientific literature.

We are grateful for the expertise and enthusiasm from all the health care professionals involved in CF care delivery in Australia who have participated in the Project.

Review Process

An evaluation of the guidelines will be undertaken across all CF centres in Australia. Prior to 2012, this document will be reviewed and updated. Updates of the guidelines will incorporate the findings of the evaluation as well as new evidence from the scientific literature.

We are grateful for the expertise and enthusiasm from all the health care professionals involved in CF care delivery in Australia who have participated in the Project.

Steering Committee

The membership of the Steering Committee included:

Scott Bell, The Prince Charles Hospital, Brisbane (Co-Chair) - Physician
Philip Robinson, Royal Children's Hospital, Melbourne (Co-Chair) - Paediatrician
Brenda Button, The Alfred Hospital, Melbourne - Physiotherapist
Peter Bye, Royal Prince Alfred Hospital, Sydney - Physician
Clare Collins, John Hunter Hospital, Newcastle - Dietitian
Conrad Guerra, Cystic Fibrosis Australia, Adelaide - CF organisation representative
Colleen Jackson, The Alfred Hospital, Melbourne - Dietitian
Lisa Martin, CF Adult, New South Wales- Adult with CF
Kerry Mordaunt*, Cystic Fibrosis WA, Perth - CF organisation representative
Carmel Moriarty, Royal Prince Alfred Hospital, Sydney - Registered Nurse
Christopher O'Connor*, Cystic Fibrosis SA, Adelaide - CF organisation representative
David Reid, Royal Hobart Hospital, Hobart - Physician
Pamela Rowell, Royal Hobart Hospital, Hobart - Registered Nurse
Gerard Ryan, Sir Charles Gairdner Hospital, Perth - Physician
Esta-Lee Tannenbaum, Royal Children's Hospital, Melbourne - Physiotherapist
Claire Wainwright, Royal Children's Hospital, Brisbane - Paediatrician
Bruce Whitehead, John Hunter Children's Hospital, Newcastle - Paediatrician

*During 2007, participation in the Steering Committee continued despite leaving organisation
The committee acknowledges the expertise of Associate Professor Peter Middleton, Westmead Hospital, Sydney; Dr Maxine Braithwaite, The Alfred Hospital, Melbourne and Professor Susan Sawyer, Centre for Adolescent Health, Royal Children's Hospital, Melbourne for their input in the writing of these guidelines. Many other CF Specialist Health Care Professionals have provided feedback and suggestions through the drafting of these guidelines. The Steering Committee is very grateful to each of them for sharing their expertise.
Disclosure Statement

Development of the guidelines was funded with financial support from Cystic Fibrosis Australia. This included travel and accommodation to the Steering Committee meeting held in Sydney in November 2006 and regular teleconferences held in 2006 and 2007. A grant from Cystic Fibrosis Australia supported the editorial role of Dr Dominic Fitzgerald. Potential conflicts of interest by membership of the Steering Committee are listed in Appendix 2.

Scott BELL  Philip ROBINSON